The BRAIN in IDIOCY. An Anatomical Study.

being a Thesis

Presented to the Senate of Glasgow University

for the degree of Doctor of Medicine.

By

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County Asylum,

Rainhill,

Liverpool.

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INTRODUCTION.

The material made use of in this study consists of fourteen Brains, taken from subjects, which during life, suffered from Amentia of a gross type, all of which brains show some gross lesion of one kind or another.

The specimens are at present in the possession of the Pathological Museum of the County Asylum, Rainhill; they are the product of a collection taken from over 2000 post-mortem examinations held in a Mental hospital of over 2000 beds, and with an average annual admission of about 300. These numbers show the great difficulty experienced in obtaining Idiot brains in which macroscopic lesions can be demonstrated to account for the Amentia, for although the total number of Idiot brains (all of which have been placed at my disposal) preserved here are greatly in excess of these numbers, yet those excluded have not shown gross lesions sufficient to allow of their fair inclusion within the scope of this inquiry.

This thesis deals for the most part with the Morbid Anatomy of the Brains in Idiocy; the Etiology, Microscopy, etc., have only in a few cases been touched upon when they, in my opinion, throw light either in support of the theories of other observers, or in placing before your notice

your notice

some new ideas which I have not previously seen put forward.

It is my intention to give a faithful description of a series of brains; some of these are possibly unique in their character, others will perhaps only add to the number of previously described cases of the same condition, but some it is to be hoped will throw more light on the position and causation of the lesion in the particular condition.

In the majority of cases it will be seen that the Idiocy has been of Uterine origin, and more or less probably due to some partial or complete arterial occlusion. Within the limits of this thesis, however, it has not been found possible to enter to any extent into the embryological side of many of the questions raised, but I would hope that the specialist in that field of work might derive some benefit from the records which I have obtained of many pathological changes which pertain to foetal life, more particularly with regard to the arrest of circulation of the embryonic brain.

CLASSIFICATION.

As regards the classification of the following cases, it must be clearly understood that it is based entirely on the morbid anatomical appearances.

It is to be regretted that Amentia as a whole cannot be classified in much the same way, for when tabulated on any other grounds, such as, for instance, on the clinical variations, the classification becomes extremely unsatisfactory, as so much depends on the personal equation of the observer.

It seems best to preface my remarks on classification by an allusion to that of Bournville, who has done the most work on this subject. His classification is as follows:-

Idiocy due to Chronic Meningitis.

" " " Meningo-encephalitis.

" " " interference of development of the brain without malformation.

" " " Hypertrophic or Tuberose Sclerosis.

" " " Atrophic Sclerosis.

" " " Internal or External Hydrocephalus.

" " " Myxodema.

" " " interference of development of brain along with congenital malformation, as Porencephaly, etc.

Idiocy due to Microcephalia.

to Microcephalia.

Idiocy with unilateral or bilateral paralysis.(due to softening, haemorrhage, etc.).

While there is much to be said for this method of classification, I have felt myself compelled to modify it in certain respects; for instance, I am forced to separate Hypertrophic from Tuberose Sclerosis; again to subdivide Atrophic Sclerosis into two, namely, Localised Atrophic Sclerosis and Microgyria. Idiocy with unilateral or bilateral paralysis I reject as a clinical distinction with which I do not deal. I have not had the opportunity of dealing with any case of Myxoedematous Idiocy or of any case in which there was any definitely conclusive Meningitic affection. I have added Macrocephalia, a condition akin to Bournville's Hypertrophic Sclerosis to the classification.

My classification is as follows:-

Idiocy due to Porencephaly,including Pseudo-porencephaly.

" " " Hydrocephalus.

" " " Localised Sclerosis. (Atrophic).
" " " Tuberose Sclerosis.

" " " Microgyria.

" " " Microcephalia.

" " " Macrocephalia.

" " " Cerebral Asymmetry or Disparity without any definite deformity.

definite deformity.

Under each of these headings I propose to give illustrated cases. Previously to describing the morbid anatomical appearances I shall give a resume of the literature and views of other observers, and, where possible, place new views and proofs forward regarding these states, following each case with a summary of the more important points in the morbid anatomy and more particularly those bearing on the causation and pathology of the condition.

P O R E N C E P H A L Y .

By the term Porencephaly is meant, a defective formation in the foetal brain, caused by destructive processes, which as regards their nature and cause are not very different from those occurring in the developed brain as a result of haemorrhage, thrombosis or embolism. The use of the term has in past years been much extended, and it may now be said to include cases which in the original terminology would have been excluded.

True or complete Porencephaly is extremely rare, by it is understood a "hole in the brain substance." In this condition the Subdural space is separated from the ventricles by simple Pia-Arachnoid, everything else (White and Grey matter) being obliterated; it must nevertheless be granted that that form of true porencephaly which may be termed "Incomplete"(Scarred) is not excluded, as here the same process has taken place without, however, penetrating to such a depth.

A remark may be interpolated concerning another condition which has been included by most observers in the category of Porencephaly, under the term of "Acquired Porencephaly;" by this Kundrat and others define Porencephalic-like defects of extra-uterine occurrence. If we accept

If we accept this definition, it is perfectly clear that every case of healing of a traumatic or haemorrhagic lesion in which a cyst or scar on the surface of the brain is left, must be termed Porencephaly, and thus every case of haemorrhagic cyst must be brought under this term, a state of matters which is absurd.

Passing now to the morbid anatomy and pathology of Porencephaly, we find that very divergent views are held by different observers. Curveilhier holds that it is a direct Encephalitis, while Roger states that it is "congenital, acquired, or direct, and is the result of healing of an idiopathic or traumatic inflammatory process". Klebs publishes three "Congenital" cases, and states that the lesion is entirely due to a vascular obliteration, but to this Kundrat does not agree, looking upon the vascular obliteration as one of the results of the process, not the cause.

The writer to whom we are indebted for the most valuable literature on Porencephaly is, without doubt, Kundrat, who has minutely classified and described twelve of his own cases. He found that the amount of destruction varied greatly; in the case most affected (a child of a few months old) there was an absence of the whole of the cerebral hemispheres excepting the remains of the Corpora

of the Corpora

Striata, Optic Thalami and the Basal surface of the Right Occipital Lobe; in the case least affected (a man whose power of intelligence was not in any way impaired) there was only destruction of a small part of the convexity of the Left Frontal Lobe.

Let us now examine the True variety more in detail. We find that in this condition the underlying and most important fact is that the development of the brain is interfered with, and it is this interference with development which causes the curious anomalous bizarre arrangement of the convolutions to which all observers have drawn attention. Examining this distribution of convolutions we find that they are arranged in a peculiar radiate or fan-like manner, the radiations being directed from the middle point of the lesion; this may be said to be a diagnostic point of Porencephaly. If the lesion occurs before the primary sulci are laid down, it necessarily follows that they will be incapable of definition, and it is by a study of the sulci and convolutions that the embryologist is able to state definitely at what period of uterine life the lesion has taken place.

Regarding the "Poros" or hole, we find its lower limit to be bounded by the ependyma and that it is lined by arachnoid and pia.

pia

The condition is in all probability brought about by an occlusion of an artery, often the middle cerebral or a branch of the same. We must here note certain differences between vascular occlusion affecting the developing brain and the same thing occurring in the fully developed organ; in both, of course, we get softening, but the amount of this during foetal life can be much greater than in the adult without causing the death of the foetus, as the brain is not then an essential organ; again a more important point in this question is that if we have occlusion of a vessel occurring early in the development of the organ, we have destruction of certain parts and this is followed by distortion of others, if the occlusion occurs after the brain is fully developed we get destruction but no distortion of other parts.

In some cases the vessels may be seen remaining and the surrounding nervous tissue completely absent; this seems due to the partial occlusion of a vessel such as occurs in Senility, Syphilis, etc., and in the same way leading to necrosis, although the vessel may be to a certain extent patent.

The meninges and ependyma have a double blood supply and are in only a few recorded cases entirely

entirely

obliterated, in these few there is usually some concomitant pathological condition present, such as Hydrocephalus.

As regards the position of the lesion, this always follows the distribution of the cerebral arteries, and is always confined to the mantle of the brain; (the basal ganglia never being affected in pure porencephaly; they may, however, be impinged upon if this condition is complicated with hydrocephaly); the Insula seems to be an extremely vulnerable area, the artery to this part being specially liable to occlusion during foetal life.

In some cases signs of an injury remain, the altered remains of the meninges, ependyma, rough walls, pigment, etc., all pointing to a lesion previously.

PORENCEPHALIC IDIOCY.

CASE No.1313.

M.B.(female), aet.24, admitted March 12th,1890.

Died, May 16th,1899.

Cause of death:- Phthisis.

Family History:-

Nothing of importance.

Condition previous to admission:-

She was always a backward child, she was long in speaking and walking. At the age of three she had a fit of an epileptiform character, and these occurred at three monthly intervals until she was five, but she had none after that age. In August 1899 she had four attacks of stupor. Her mind generally deteriorated, and she was admitted to Rainhill Asylum in the following state.

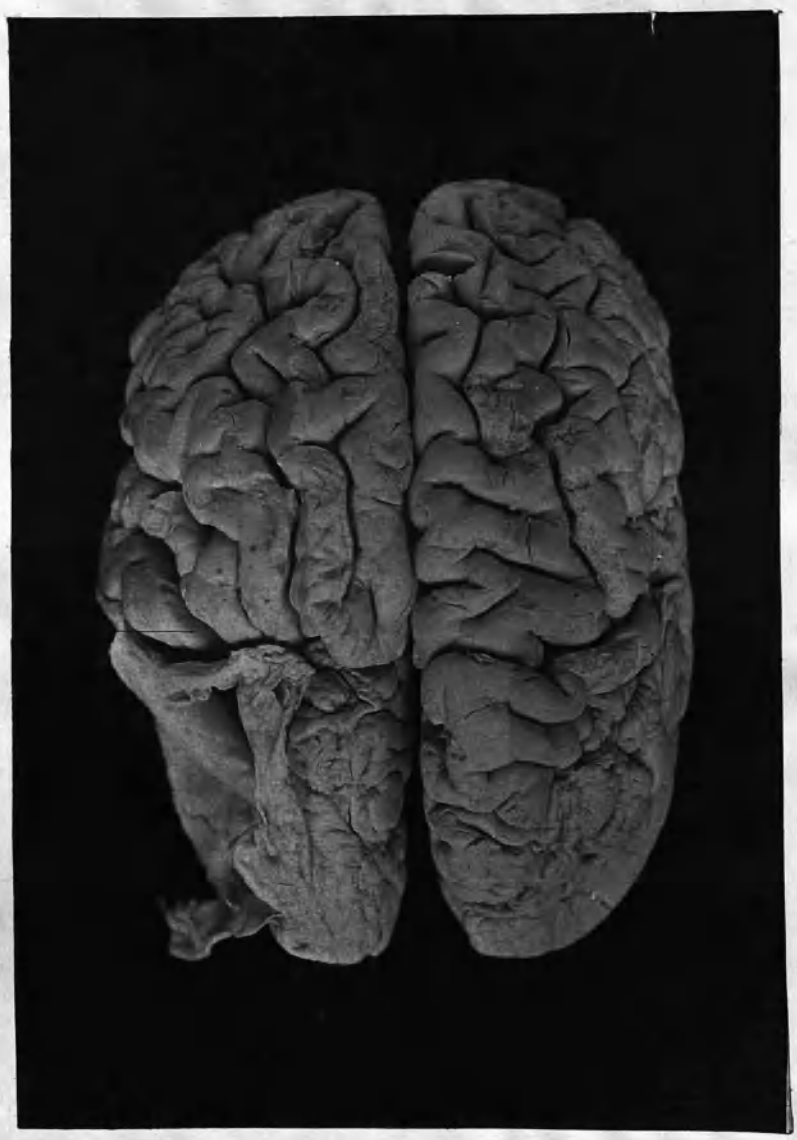
Physically:- She was fairly healthy. Right side somewhat paretic. Double Strabismus which only from time to time occurred and pointed in various directions.

Mentally:- She was an idiot. Her attention was in abeyance; she could not converse. Chattered a good deal to herself, but no sense could be made out of her articulatory efforts. She was wet and dirty in her habits.

Later:-

Her mental state did not improve; she died an

Case No. 1313.

Left.  Right.

View of Both Hemispheres from above.
Note:- The position of the Porencephalic defect. The peculiar manner in which the posterior Frontal convolutions radiate from the seat of the lesion.

died an

absolute idiot.

Post Mortem Examination:-

"Patient is a woman of average muscular development and no deformities are to be noted:-

Skull:-

In the left parieto-temporal region there is an unusual extensive indentation (viewed from within) of the bone, corresponding with a porencephalic defect in the left cerebral hemisphere. The bone in this situation is somewhat thinned and the grooves for the meningeal artery are much shallower than those on the right side. The right temporal and occipital fossae are larger than those of the left. No abnormality of the cranial sutures is discoverable. The Dura Mater is of normal thickness excepting along the sutures where it is thickened; it is bulged in the left parieto-occipital region corresponding with the lesion.

Upon reflecting the dura, what looks like a large cyst, is present in the left parieto-occipital region. The enclosing wall of this "cyst" is extremely thin (and was accidently torn in the removal of the brain), it is composed of an opaque layer of (1). pia-arachnoid in which attenuated veins and arteries run, and (2), what seems to be the remains of the ventricular ependyma; this layer is observed to be

to be

delicately nodulated.

The "cyst" is in direct connection with the lateral ventricles."

RIGHT HEMISPHERE:-

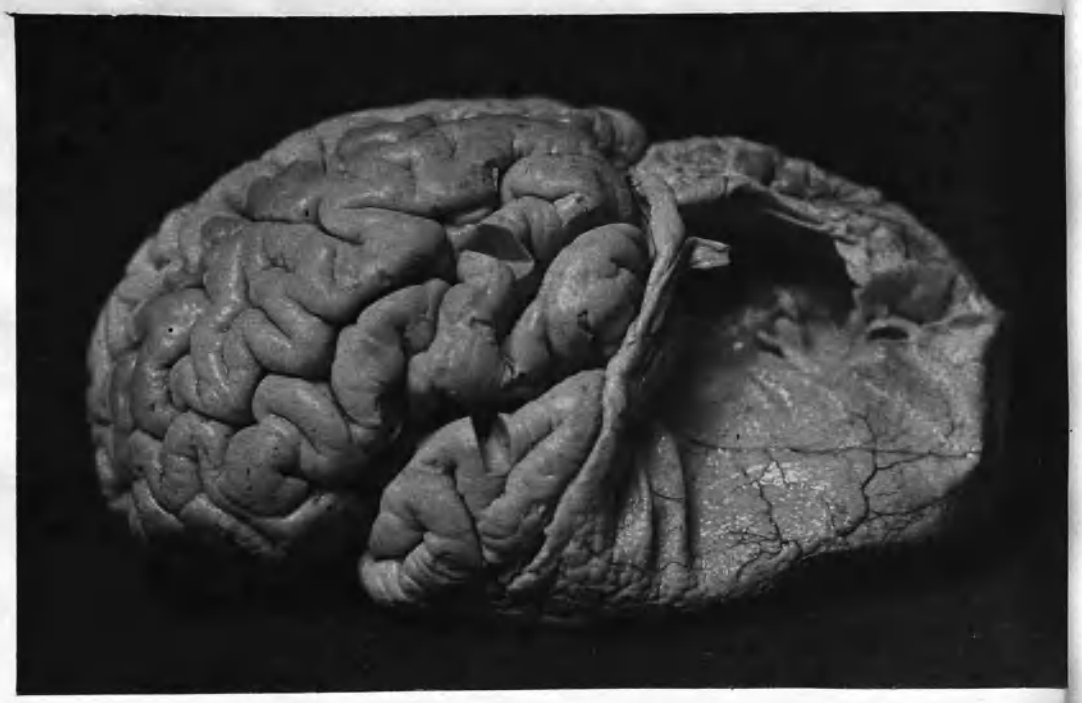
The fissure of Rolando is easily determined. The first Frontal sulcus is also easily made out, but its posterior extremity is abnormal in that it arches downwards instead of curving upwards; this, it will be noticed, gives undue width to the posterior third of the First Frontal convolution, and in this part of the convolution are situated two sulci about 2 1/2 ctm. long, running parallel to one another, and forming an angle of about 45 degrees with the upper half of the fissure of Rolando. These sulci are to be specially noted, as they appear to be radiating towards the main lesion, and they remind one of the convolutional arrangements met with in intra-uterine porencephaly.

The Second Frontal sulcus is also easily defined but the Second Frontal convolution tapers posteriorly.

The Third Frontal convolution is of normal size, but there is a very peculiar Pars Triangularis, owing to the wide separation of the anterior ascending and horizontal limbs of the fissure of Sylvius.

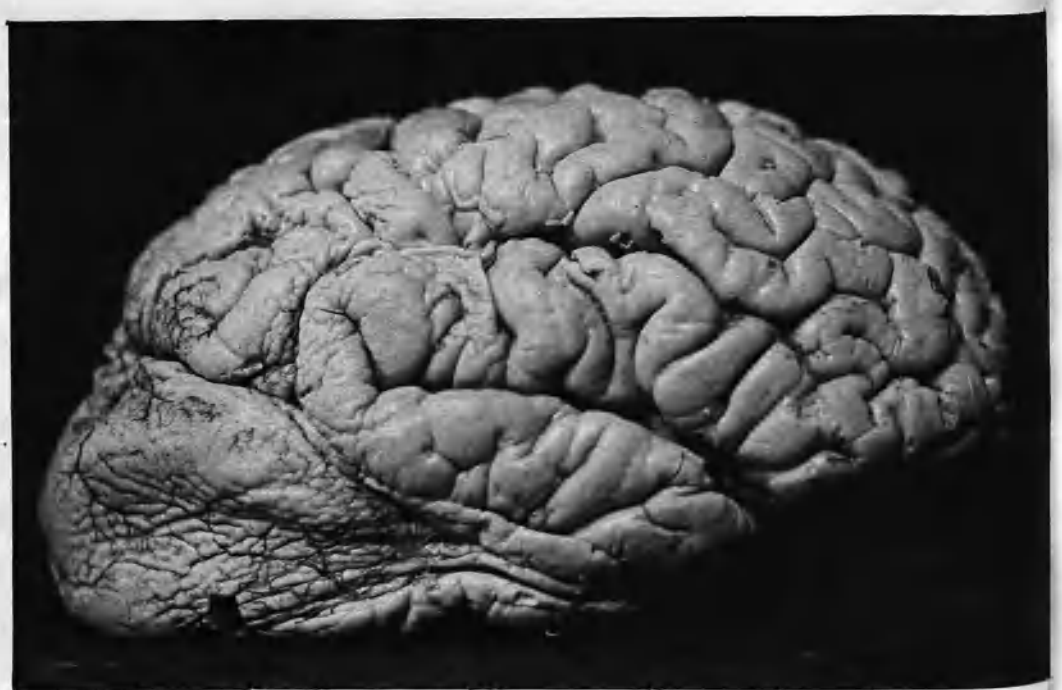
All the gyri above mentioned are of a rather small

Case No. 1313.



Left Hemisphere. Outer Surface.

Note:- The huge"cyst-like" formation occupying t
Parieto-Occipital region. The vertical fissur
cutting through the fissure of Rolando, and in
front of this the comparative normal appearance
of the Frontal and Antero-Temporal regions.



Right Hemisphere. Outer Surface.

Note:- The wrinkled leathery appearance of the
Occipito-Temporal region, with in the Occipital
Lobe expecially, the complete absence of convol-
utional indication

small

size, but otherwise normal in appearance.

The ascending Frontal convolution is abnormal; at the junction of its lower and middle thirds ~~x~~ a shrunken puckered patch 1 1/2 ctm. long is to be seen. The lower third of the Ascending Parietal convolution is in a similar state.

In the Parietal region, there is very much distortion, owing to the shrinking and puckering of a number of convolutions, and this has occurred to such a degree as to make it impossible to define the various sulci. The Superior Parietal convolution is the only one that is not wasted, but it will readily be noticed that its individual sulci radiate towards the lesion below.

In the external Occipito-temporal area is a large tongue shaped area, tapering anteriorly, where not a single sulcus nor convolution can be defined, (this is well brought out in the photograph of this area); and here the ventricle is only separated from the surface by a layer of substance about 3 m.m. in thickness. The upper boundary of this area corresponds with an imaginary line drawn backwards from the Sylvian fissure, posteriorly it reaches to and extends over the tip of the Occipital lobe, below, it in a like manner reaches over the inferior surface of the Occipital

Occipital

lobe, and anteriorly its boundaries converge and meet at the centre of the Temporal lobe.

The first Temporal convolution is of good size but it tends to become puckered posteriorly.

The Second Temporal convolution is involved in the lesion posteriorly, while the Third Temporal convolution is practically intact.

The absolutely wasted area above noted has a wrinkled leathery aspect.

Inner Surface:-

Excepting the Quadrate lobe and the Internal Occipito-Temporal convolutions, the inner surface has a normal aspect.

In the Quadrate lobe there is no lesion but the sulci have the radiating arrangement previously mentioned, there are three sulci from 2 to 4 ctm. long, equidistant from one another, and all running parallel to the Parieto-Occipital fissure, which later is normal in position.

The entire Cuneus and all the convolutions which lie immediately below the same are withered and thinned like those in the main external lesion.

As regards the Corpus Callosum it is noticed that the posterior 2/5ths is markedly thinner than the remainder.

remainder.

Lower Surface:-

The Orbital Surface is normal but all the convolutions of the under surface of the temporal lobe are either absolutely wasted or puckered as in the posterior portion, or else partially wasted and puckered as in the anterior portion. In the anterior portion the sulci are extremely shallow, and for the most part radiate towards the lesion on the outer surface of the hemisphere. The layer of cerebral substance between the ventricle and the surface is only 2 or 3 mm. thicker here than it is in the absolutely wasted portions.

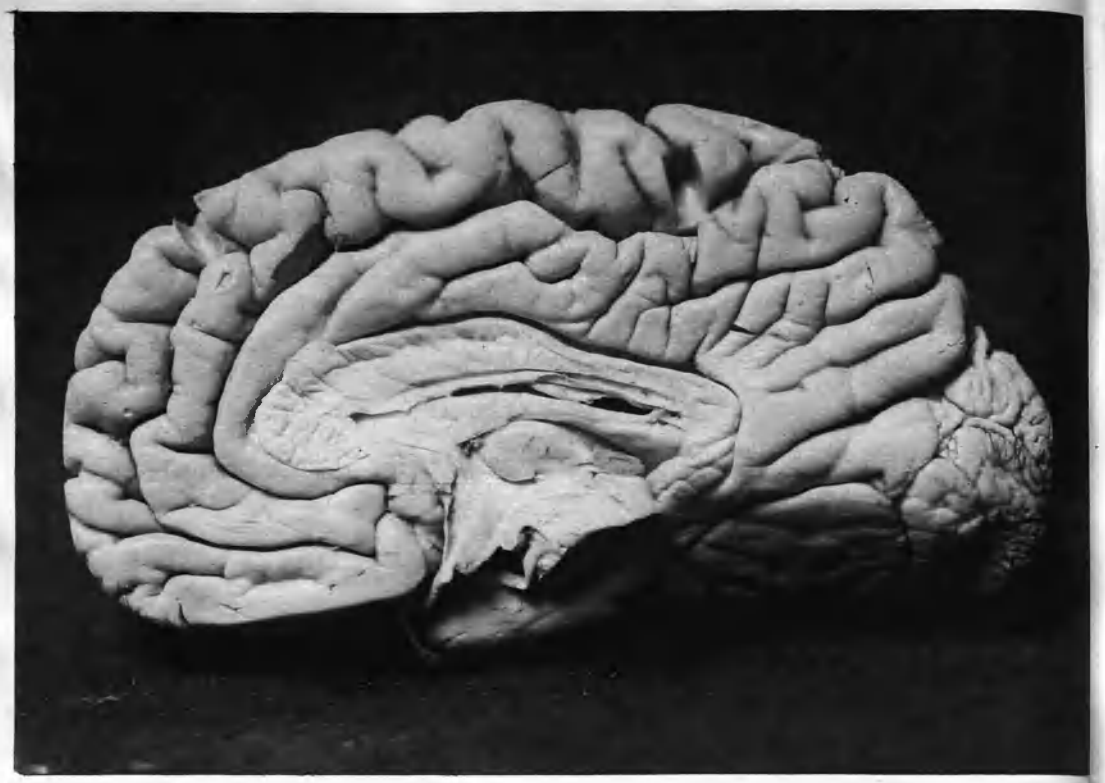
LEFT HEMISPHERE:-

In the left hemisphere the porencephalic defect is much more extensive, and the wall of the cavity is much thinner than that of the right, but on the whole it is of similar character.

Outer Surface:-

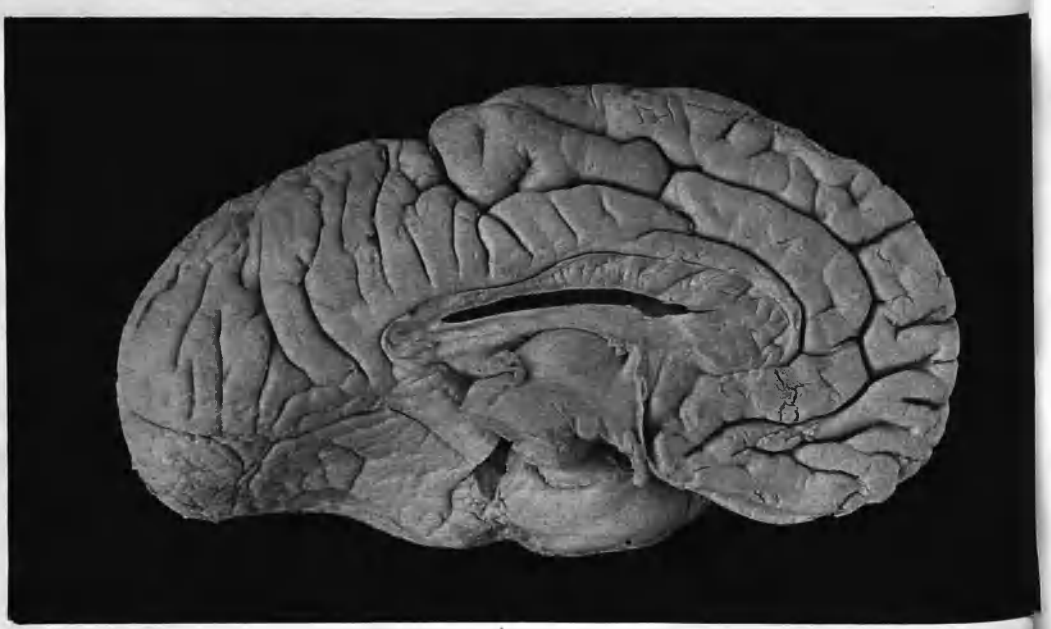
At about the centre of the outer surface a vertical furrow or sulcus extends from the upper internal margin of the hemisphere well down into the Temporal lobe, where it arches forwards. It commences, above, somewhat anterior to the fissure of Rolando and does not have such an

Case No. 1313.



Right Hemisphere. Inner Surface.

Note:- The shrunken leathery appearance of the posterior pole of the hemisphere. The distort of the convolutions in the Cuneus and Precuneus The narrowing of the posterior two-fifths of th Corpus Callosum. The normal appearance of the anterior two-thirds of the hemisphere.



Left Hemisphere. Inner Surface.

Note:- The large vertical fissure which almost bisects th hemisphere. The curious Quadrate and Cuneus, and the mal formation of the Calcarine region. The thinning of the Corpus Callosum.

such an

oblique course as that fissure, it crosses the fissure of Rolando at its middle point and the fissure of Sylvius posteriorly.

Roughly speaking all the convolutions in front of the above fissure have a more or less healthy aspect, while those behind it are replaced by an immense "cyst".

The first and Second Frontal sulci are easily definable and the various gyri, with the exception of being a trifle small, look normal; but the posterior portions tend to be arranged in super-imposed layers radiating towards the main lesion. For this reason, only the lower half of the precentral sulcus is definable.

The anterior ascending and transverse branches of the Sylvian fissure are again widely separated and the Pars Triangularis is of U-form.

The lower two thirds of the Ascending Frontal convolution are normal in position and are of average size, but the superior third is involved in the lesion and cannot be defined.

The lower third of the Ascending Parietal convolution is all that remains of that gyrus.

The Insula is slightly exposed, its anterior convolutions appear normal, but one or two of the poaterior

posterior

are involved in the lesion.

Of the Temporal lobe, about the anterior half of the First and the anterior third of the Second Temporal gyri are all that remain healthy.

There is a strip of Superior Parietal convolution bordering the Superior Longitudinal fissure which is not quite destroyed but is represented by a collection of attenuated and puckered irregularly arranged convolutions.

The entire remainder of the outer surface of the hemisphere is represented by the wall of the "cyst-like" cavity.

Inner Surface.

On the inner the destruction is not great but the arrangement of the convolutions is markedly peculiar.

The great vertical sulcus described on the outer surface is now seen coming over the crest of the hemisphere and in this situation it is of great depth, it bisects the Paracentral lobule and continues downwards almost to the Corpus Callosum. All the convolutions in front of this sulcus are normally disposed; an off-shoot of the calloso-marginal sulcus, however, courses backwards through the Paracentral lobule in an unusual manner and it is undoubtedly radiating towards the lesion.

lesion.

The Quadrate lobe is remarkable, and the Parieto-Occipital fissure extremely difficult to define; count-back from the above mentioned vertical fissure there are seven rather small convolutions of varying length, arranged parallel to that fissure, and the sulcus behind the seventh I take to be the Parieto-Occipital.

The Cuneus is equally difficult to define, it seems to me to be of large size and made up of several flat convolutions separated from one another by shallow sulci radiating towards the lesion. Below the Cuneus is a large horizontally-directed sulcus which is probably the Calcarine fissure, but the anterior portion of this fissure is difficult to define.

The Collateral fissure defies definition, and all the Occipito-Temporal convolutions, with the exception of the tip of the Temporal lobe, are flattened out and thin, and all the sulci are extremely shallow.

The Cerebral substance of the convolution below the Calcarine fissure is only 3 to 4 m.m. thick, while that of the Cuneus and Quadrate lobes varies between 5 and 10 mm.

The posterior half of the Corpus Callosum is very thin.

thin.

The Orbital Surface is normal.

The Cerebellum is of good size and the hemispheres equal and normal in every detail.

The Pons and Medulla appear normal, except that the left pyramid is smaller than the right.

SUMMARY.

Commenting on the above case the following are the chief points of interest:-

Bilaterally-situated Porencephalic defect is of great rarity, as is also the occurrence of this lesion in the Occipital lobe, although not perhaps to such a degree.

The radiating arrangement of the sulci, so characteristic of Porencephaly is extremely well seen.

The Posterior Cerebral artery must, without doubt, have been that occluded, and although the distribution of the affection does not exactly coincide with the parts supplied by that artery in the fully developed brain yet it is quite within the bounds of possibility that in foetal life the actual area implicated is that then supplied by the same. On no other assumption can the distribution of the lesion be explained.

Considering the large area of cortex interferred with, it is very remarkable that so little clinical

clinical

manifestation should have been present; with so much of the motor area on the left side destroyed as we have here, we should naturally have expected considerable paralysis, contracture and atrophy, but there was only a very slight degree of right hemi-paresis. With so much implication of the occipital lobe, visuo-sensory and visuo-psychic disturbances were to be looked for; the first were not present; as regards the second, the patient's mental condition precluded examination.

The peculiarity of this affection in having definite areas destroyed without correspondingly exact physical manifestation has been alluded to before by various observers; and the question arises as to how this can be accounted for. It is perhaps possible for another portion of the cortex to assume a vicarious functional activity, but I think that in this case at least, the less damaged hemisphere has taken upon itself to a large extent, the work of the other.

Another point of interest is, that in spite of the great amount of tissue destroyed on the left side, there is no distinct "poros", probably this may have been due to the lesion having taken place before the formation

formation

of the pia-arachnoid.

Regarding the patient's mental state, it is a well-known fact, and one which will be likewise emphasised in many other cases to be here described, that if any gross lesion of the brain occurs, no matter in what position, a condition of Amentia results.

In this case no less than three of the great association centres of Flechsig are interferred with, a condition which assuredly is only compatible with profound Idiocy.

CASE.No.1433.

J.B. (male), aet.36, adm.Aug 2nd.1899.

Died:- Feb.5th.1900.

Cause of Death:- Phthisis.

Idiocy.

History previous to admission:-

The paralysis came on at the age of three. He could not be kept at school on account of his dulness and mischievous tendencies, and it was quite impossible to teach him anything.

On admission:-

There was right-sided hemiparesis due to an attack of Infantile Paralysis; this was accompanied by much deformity and shortening of both extremities.

He was able to speak, and moaned and cried a great deal.

He could not tell the day of the week or even the month.

His attention was much impaired and he was incoherent.

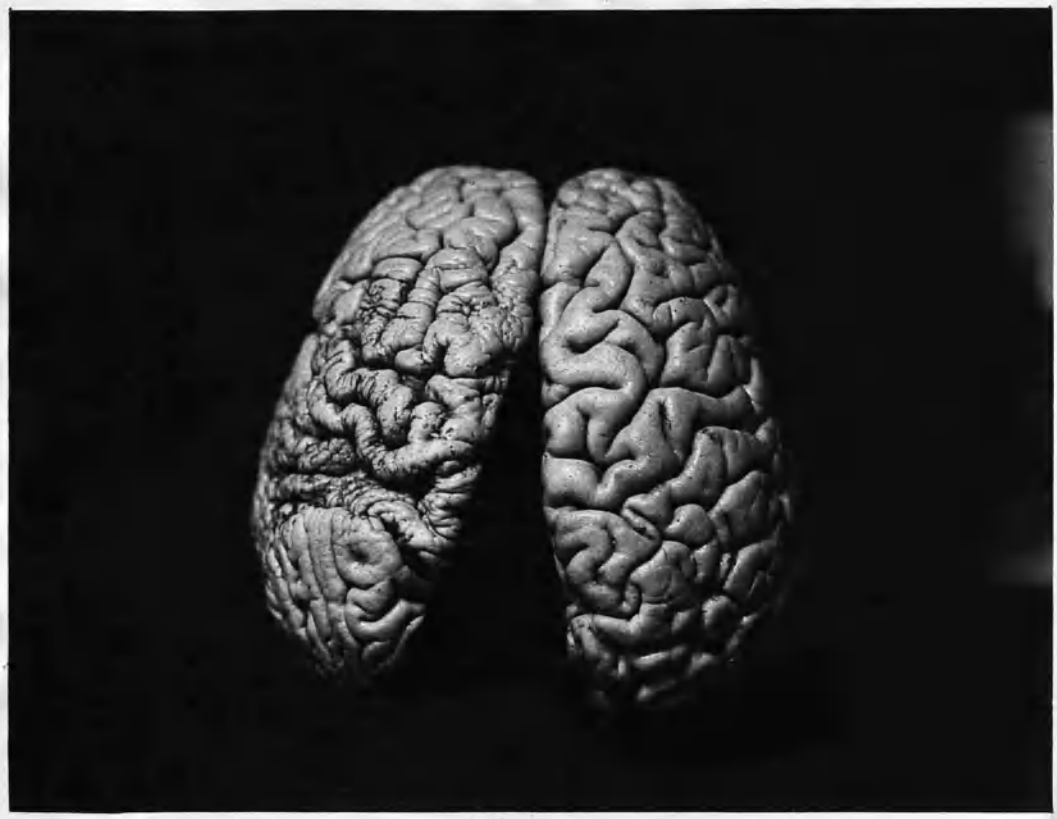
Later:-

He became more degraded and often had to be tube-fed. He had to have all his wants attended to, and was wet and dirty in his habits.

Post-Mortem Examination:-

"There is marked atrophy or non-development of both extremities of right side; shortening and thinning

Case No.1433.



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View of Upper Surface of both Hemispheres.

Note:- The difference in size between the two hemispheres. The distortion of the Rolandic and Parietal areas are well-shown.

thinning

of the bones is not so marked as the poorness of muscular development. There is pronounced talipes equinus of right foot."

CEREBRUM:-

Left Hemisphere:-

The left cerebral hemisphere is markedly smaller than the right, and its arrangement of convolutions curiously deformed, owing to a lesion in the Parietal region.

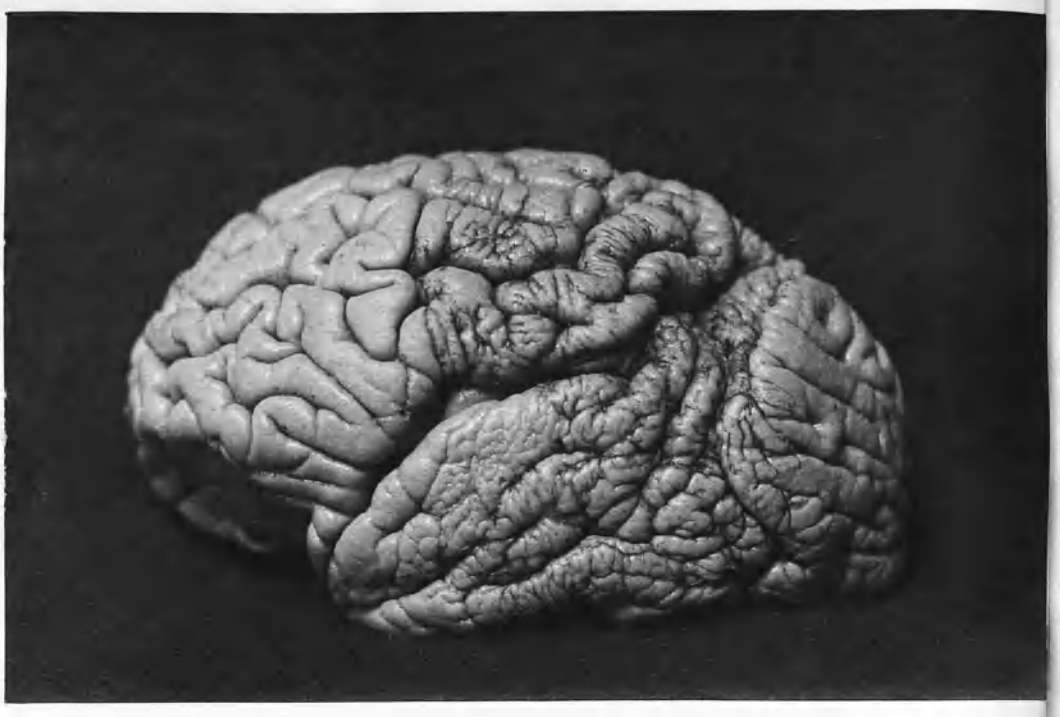
On the outer surface, the Frontal segment is of good size, the Parietal, Temporal and Occipital areas are small, in addition, in the Parietal and Temporal regions there are seen many puckered wasted convolutions and shallow sulci.

The fissure of Rolando is difficult to define, but taking the upturned end (hinder) of the Calloso-marginal sulcus as a guide, it seems to be the second sulcus in front of the "gap" in the Parietal region, (to be presently described.).

The fissure of Sylvius is 6 1/2 ctm. in length, and ends without deflection or bifurcation.

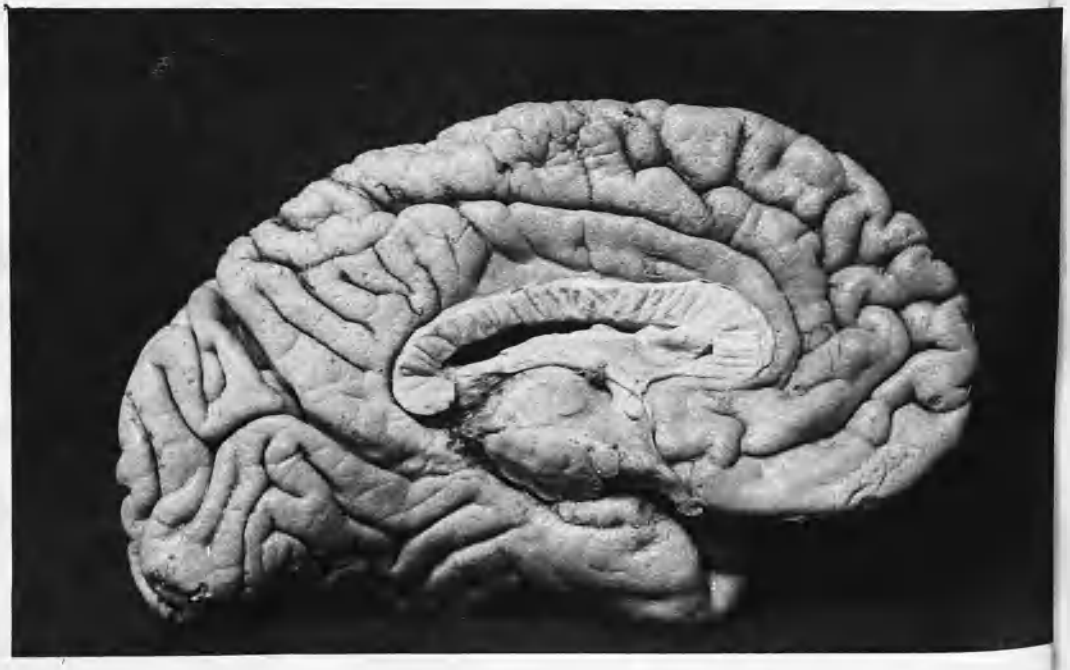
The Intraparietal fissure is not definable. Although the Frontal region is of good size, its arrangement

Case No. 1433.



Left Hemisphere. Outer Surface.

Note:- The puckering in the Postero-frontal, Parietal and Temporal regions. The appearance of the fissure of Rolando & Sylvius. The great disturbance of the Intraparietal region.



Left Hemisphere. Inner Surface.

Note:- The appearance of the Paracentral Lobule, Cuneus, and the Gyrus Fornicatus.

The Calcarine fissure

arrangement

of gyri and sulci differs considerably from normal.

All those gyri which lie within an area bounded behind by the fissure of Rolando, above by the inner margin of the hemisphere, below by the fissure of Sylvius, and in front by a line dropped vertically through the Sulcus Diagonalis, are rather small and crossed by puckers and shallow furrows; they also tend to arrange themselves in tiers one above the other, and an ascending Frontal convolution of normal shape cannot be made out. Instead of this area there are four horizontal sulci above one another and no vertical ones; but there is a normally-placed Sulcus Diagonalis which, however, does not join the fissure of Sylvius.

The arrangement of the External Occipital convolutions is simple, they consist of 6 convolutions and 5 sulci, all arranged parallel one above the other and extending from the "gap" to the hinder border of the hemisphere.

The Insula is covered, but the fissure of Sylvius is shallow anteriorly and deep posteriorly.

Inner Surface:-

The Corpus Callosum is short but of good depth. There is a well-marked Calloso-Marginal sulcus without branches; the Gyrus Fornicatus tapers considerably anteriorly.

anteriorly.

The inner Frontal convolutions appear normal. The Paracentral lobule is small and puckered. The Precuneus is reduced in area and divided into two portions by a shallow oblique sulcus, directed backwards and upwards, which is evidently a branch of the Calloso-Marginal sulcus. The upper portion is small and puckered, the lower portion down to the Corpus Callosum is composed of seven small convolutions lying parallel to one another and separated by six shallow sulci, all directed at right angles to the Corpus Callosum.

The Parieto-Occipital fissure is normally placed but short.

The Cuneus is small, and the Calcarine forks into two long branches, 1.5.ctm. behind the Parieto-Occipital fissure.

The Orbital surface is tilted upwards, its convolutions are small but normally disposed.

Anterior and Posterior Commissures, Fornix, Velum, etc., are normal.

Right Hemisphere:-

The Right Hemisphere is of fair size, its shape is good, all its segments are proportionately represented.

represented.

The main sulci are normally distributed, and the convolutions are on the whole small and rather complex.

SUMMARY.

Summarising this case, we find that it represents extremely well that form of Porencephaly which has been mentioned in the introduction to this condition under the designation Incomplete, and it is also an example of the Scarred Type of the same.

That it is a case of Porencephaly, and not one of ordinary cerebral softening, there can be little doubt, as the deformity of the main sulci and convolutions are characteristic of the occurrence of the lesion in the brain during the embryonic period.

It is of interest on account of the large amount of brain substance affected; the artery which has been occluded is probably some branch of the middle cerebral.

The arrangement of the convolutions, radiating from the "poros" is well marked.

CASE No.805.

J.B.(male) aet.28, adm.Sept.1891. (2nd adm.)

Died 25th.Feb.1896.

Cause of Death:- Status Epilepticus.

History previous to admission:-

He was a backward child; he could not walk until the age of four; epileptic fits commenced at the age of 15, and he gradually became dull, although at times after fits he was liable to attacks of acute excitement during which he was quite unmanageable. No history of any injury.

On admission:-

There was no definite paralysis, although there was slight weakness of right hand; his speech was of a stammering character; he was very simple in conversation and actions; he showed much impairment of memory. At this time his mental state was one of moderate imbecility.

Later:-

The fits increased in number up to 239 per annum. He became duller, and at death his mental state bordered on Idiocy.

Post Mortem Examination.:-

"Nothing of importance to be noticed on external examination of the body, no localised atrophy or facial asymmetry.

asymmetry.

Skull:-

Of average size and somewhat Brachycephalic, perfectly symmetrical.

Encephalon:-

Weight 1372 grms.

Scattered over the surface of the brain are numerous small bodies the size of a pea, and in stripping the pia these come away with it.

The right hemisphere is of good size, but the left is smaller from non-development of certain convolutions."

CEREBRUM:-

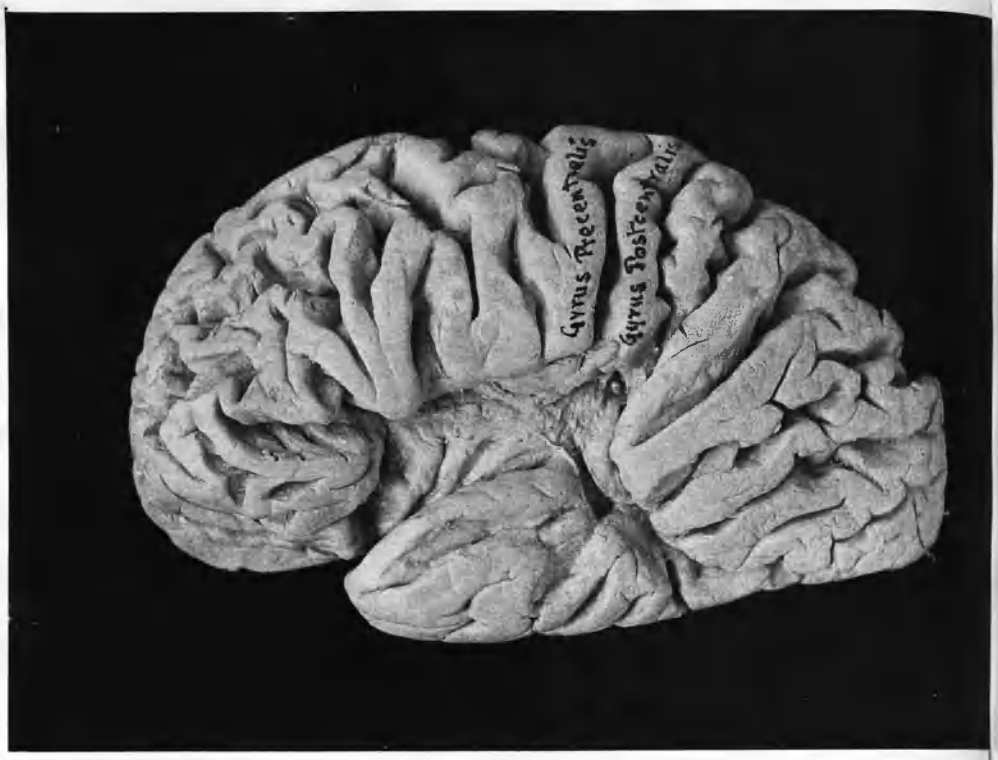
Left Hemisphere:-

In the left hemisphere a curious condition, which has resulted in the disappearance of a considerable amount of cerebral substance and an unusual arrangement of the convolutions, is visible, the condition being more suggestive of arrested Porencephaly than anything else.

Parts Destroyed:-

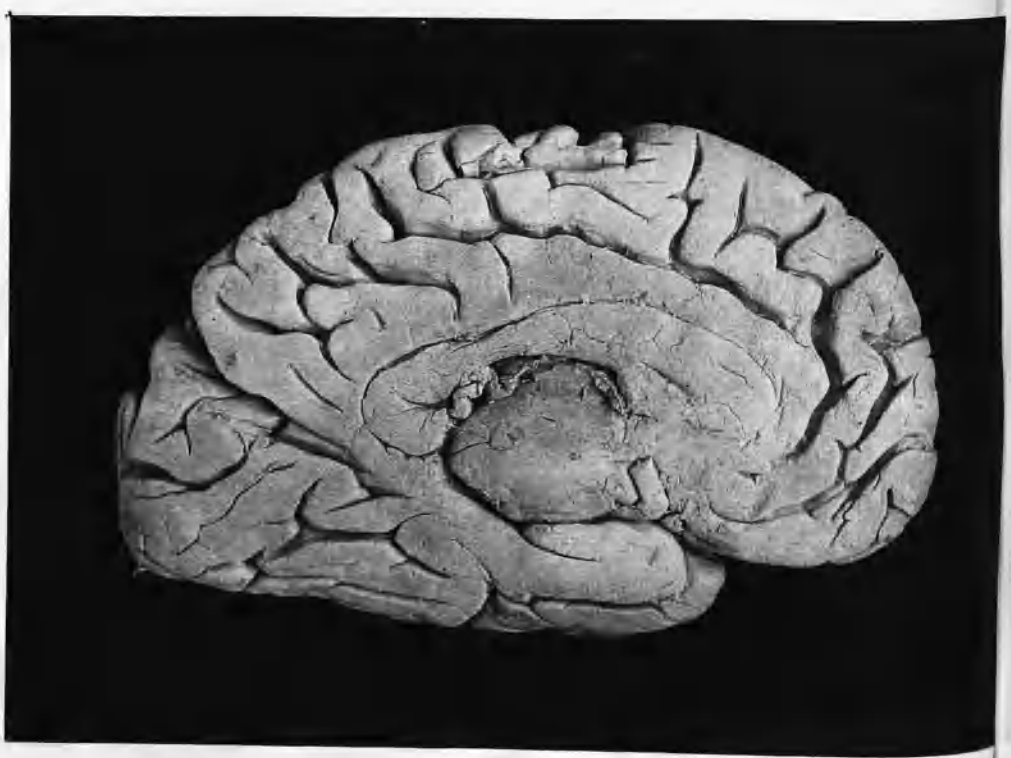
Taking first that portion of the cortex in which the convolutions are not represented, these are the Opercular parts of the Third Frontal (posterior part), Ascending Frontal, Ascending Parietal and Inferior Parietal

Case No. 805.



Left Hemisphere. Outer Surface.

Note:- The exposure of the Insula, as a result
the disappearance of the Fronto-parietal Operc
The deep vertical fissure cutting through the
poral Lobe. The fan-like disposition of the c
volutions and sulci from the seat of defect.



Left Hemisphere. Inner Surface.

Parietal or

Supra-marginal convolutions. The remnants of these parts appear as a sclerosed mass of substance bulging into the Fossa of Sylvii over the Insula.

Next, the Temporal segment is divided at about the junction of its middle and posterior thirds by a deep wide fissure running from the Sylvian fossa downwards and slightly backwards to the Third Temporal convolution. This fissure has been obviously formed by a destructive process. An indication of its continuation upwards over the Insula is given by a thin cord-like line on the Insula, and at this situation the long convolutions of the posterior portion of the Insula have undergone destruction.

Arrangement of Convolutions:-

Compared with the Parietal particularly, but also with the Occipital and Temporal, the Frontal segment is of large size and complex arrangement, this is mainly due to a curious arrangement of the Precentral parts; a well-marked Precentral sulcus interrupted in the middle, and joining the fissure of Rolando), then in front of this a vertical convolution running from the inner surface to the fissure of Sylvius, still more anteriorly is another vertical sulcus, not reaching the inner margin, nor joining the fissure

fissure of

Sylvius, then slightly more anteriorly a sulcus, one inch long, is found joining the fissure of Sylvius (this seems to be the Sulcus Diagonalis).

The First and Second Frontal sulci are considerably interrupted.

The fissure of Rolando is shortened at its lower end and runs more vertically than normal.

The Ascending Parietal convolution is thin, it is bounded posteriorly by a parallel Postcentral sulcus joining the fissure of Sylvius, behind the Postcentral sulcus is another vertical but shorter convolution, behind this again is another sulcus continuous with that of Sylvius which runs up to the upper margin of the hemisphere. More posteriorly is another convolution directly obliquely forward and downwards, immediately behind this is a parallel fissure, apparently the prolongation upwards of the First Temporal, and it extends to the upper margin of the hemisphere; it is only separated from the Parieto-Occipital by a thin bridge of tissue.

Behind and below this sulcus the Second Temporal sulcus is continued backwards towards the Occipital region for about two inches. Likewise a very short distance

short distance

below this, the Third Temporal sulcus is prolonged backwards almost to the posterior end of the Occipital segment.

The sulci on the inner surface of the hemisphere are normally arranged and the convolutions are large and simple.

Right Hemisphere:-

This is to all practical purposes normal, both in distribution of sulci and arrangement of convolutions.

SUMMARY.

This case is interesting in the fact that it is a perfect example of Incomplete Porencephaly, the lesion not having extended sufficiently deep to affect the ventricles.

The one diagnostic point of this condition, and the one to which all observers draw attention, namely, the radiating fan-like deposition of the sulci, all spreading from the point of defect, is extremely well exhibited.

CASE.No.869.

J.Y.(male), aet.21. adm.Jan.16th.1891.

Died July 21st,1896.

Cause of Death:- Phthisis pulmonalis.

Idiocy with Epilepsy.

On admission:-

His left arm was not developed and was paralysed, it was drawn across his chest by the flexor muscles, he also had talipes equino-varus of the left foot. Practically no trace of memory or intelligence could be gauged; he muttered unintelligibly when spoken to; he did not pay any attantion when his name was called, and he ate all kinds of rubbish if he were not closely watched. His habits were wet and dirty.

Later:-

He became, if anything, more fatuous and dirty. The fits increased in number, averaging shortly before death 20.23 per mensem.

Post Mortem Examination:-

"The skull is oval in shape, temporal diameter 4 1/2 inches, anterior posterior measurement 6 1/2 inches; the bones of the vault of the right side are smaller than

than

those of the left. The Frontal, Temporal and Occipital fossae of the right side are clearly smaller than those of the left; the Crista Galli is pushed over towards the right side.

The lines of the middle meningeal arteries are clear on both sides but the remifications are more numerous on the left side.

Encephalon:-

Weight 820 gms. (with ventricular fluid)."

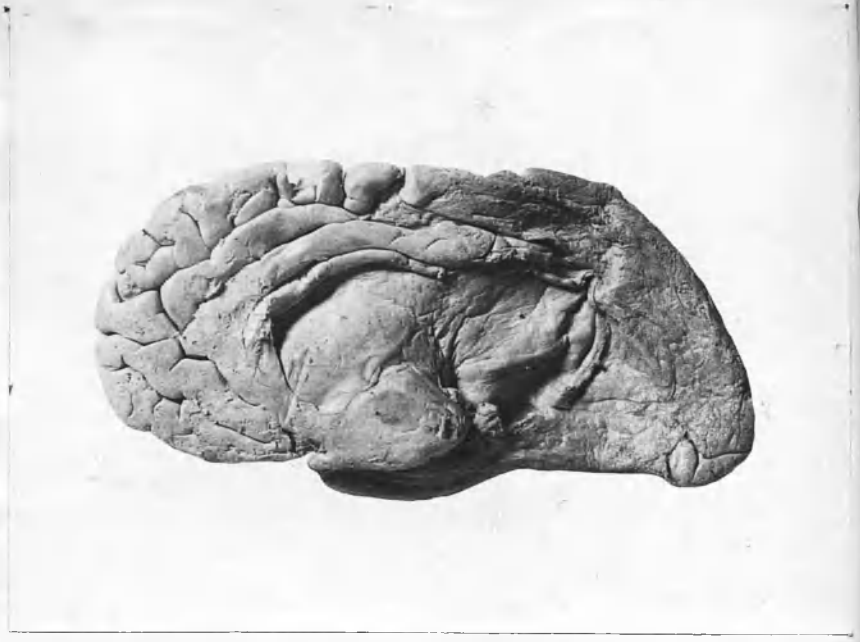
Right Hemisphere:-

Weight 102 grms.

The right cerebral hemisphere shows an extraordinary condition, as the result of some infantile lesion; it is in great part destroyed, certain parts to be mentioned presently in detail remaining, and in general appearance it resembles a bag of membranes more than a cerebral hemisphere.

At the Post Mortem Examination, it was noticed that the arachnoid did not dip down between the two hemispheres in the frontal region, the two sides being adherent to one another, although the adhesions were easily broken down. The Arachnoid and Pia covering the right hemisphere were markedly thickened, opaque and oedematous. A small anterior cerebral artery could be made out but one could only

Case No. 869.



Right Hemisphere. Inner Surface.

Note:- The Pregenual cortex is approximately h
thy. The posterior portion of the Ventricle i
greatly dilated. The convolutional arrangemen
the Paracentral, Cuneal and Occipital areas (w
the exception of the extreme tip) is obliterat

Right Hemisphere . Outer Surface.

Note:- The entire disappearance of the convolu
and sulci from the posterior two-thirds of the
hemisphere. The bulging in the lower central
tion is due to the persistence of the Basal Nu

could only find traces of a middle and posterior cerebral; the carotid on this side (right) was smaller than on the other. The meningeal arteries and veins were practically normal in size and distribution.

On the outer surface of this hemisphere it is seen that portions of the first, second and third Frontal convolutions, almost the whole of the Orbital convolutions and also the tip of the Occipital segment remain.

On the inner surface, part of the First Frontal convolution, the Marginal convolution and the Gyrus Fornicatus, and again the tip of the Occipital lobe remain. In these situations the convolutions are all very small, and the cortex is unstriated; the sulci are wide and shallow.

In the remaining parts of the hemisphere, a thin almost translucent layer is revealed which represents the sole remains of the cortex and white matter.

It is thus seen that the cortex and subjacent white matter right down to the ventricular ependyma, of the entire Tempero-sphenoidal lobe, all the Parietal convolutions, the central convolutions and part of the Frontal and Occipital lobes are destroyed.

It must here be mentioned that the translucent layer above mentioned is somewhat thicker in some places than

than

in others, owing to this fact, it is just possible to localise the original position of the main fissures.

Lateral Ventricle (right):-

Owing to the persistence of some cerebral tissue frontally, the anterior horn of the lateral ventricle is not much dilated but the posterior and descending horns are very much enlarged, hence the "cystic" appearance of the hemisphere.

Basal Nuclei:-

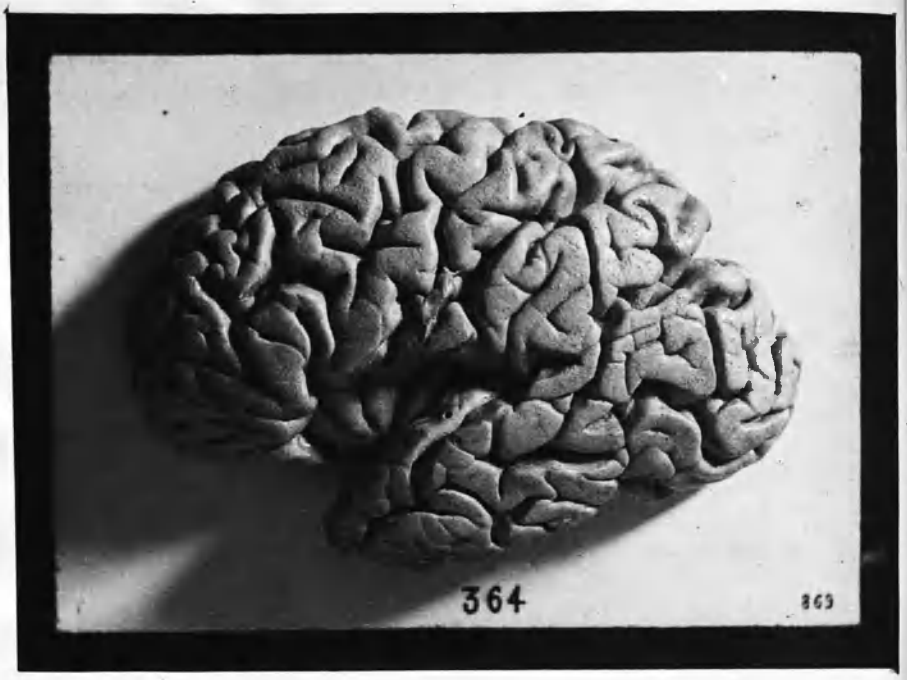
When the hemisphere is looked at from the side, the inner surface being downwards, a pyramidal bulging is seen about the position of the Island of Reil, this is caused by the persistence of the Basal Nuclei.

A Corpus Callosum of good shape but of small size can be plainly seen, also the Optic Thalamus; the latter seems to be even more reduced in size and more fibrosed than the former. Further down, the Red Nucleus is distinctly visible, likewise the Pes and Crus and Substantia Nigra Soemerringii, all being of very small size compared with those of the opposite size, but of good shape.

Corpus Callosum:-

The condition of the Corpus Callosum is interesting, In its anterior half it is about one third of

Case No. 869.



Left Hemisphere. Outer Surface.

Note:- There is no conspicuous abnormality, w
the single exception of the lower portion of
Ascending Parietal convolution.

Left Hemisphere. Inner Surface.

Note:- This is practically normal.

third of

the normal size, it tapers gradually from before backwards, until, in its posterior half, it is represented by the thinnest of films, in which fibres crossing from one side to the other can just be recognised. The continuity of the film is not broken at any part.

The middle and anterior commissures are on the whole fairly normal.

Fornix etc.,:-

A small fornix cann be seen on the right side and a normal one on the left.

The other structures are practically normal.

Left Hemisphere:-

Weight 455 gms.

The left hemisphere is of practically normal size, and the arrangement of the convolutions is moderately complex; secondary sulci are present in fair number, being visible in all segments.

The only flaw in this hemisphere is that the lower extremity of the Ascending Parietal convolution is withered and practically devoid of cortex and subjacent white matter.

Cerebellum:-

The left half of the cerebellum is appreciably

appreciably

smaller than the right, the diminution in size being confined to no particular region.

The Pons and Medulla show no external peculiarity.

Microscopic Examination:-

A rough microscopic examination of a part of the wasted cortex of the right hemisphere, examined and stained for nerve cells by the methos of Nissl, shows no trace of nerve cells, but large numbers of thickened blood vessels.

A portion of the First Frontal convolution, where there is some cerebral tissue, shows a number of small nerve cells, round or oval in shape, with short processes, nucleus and nucleolus, and granular protoplasm.

SUMMARY.

On considering the more important points of this case, the first that arises is, whether or not this case should be grouped under the heading of Porencephaly, in the true sense of the term? Strictly speaking the answer must be in the negative.

My reasons for placing it under this heading is, that according to Kundrat, it would come under the classification of Acquired Porencephaly, by which is under-

it is under-

stood, a porencephalic defect occurring after birth; but as I have emphasised in the introduction to this chapter, the term should only apply to lesions of this nature occurring in utero, it necessarily follows that this case does not come under the heading of Porencephaly, in the true sense of the term.

The grounds for my statement that the lesion at present under consideration is of extra-uterine are:- firstly, that all lesions occurring in the developing brain which produce destruction of tissue also produce distortion of the remaining parts; in this case the former is present, while the latter is almost entirely absent, as even in the smoothest parts of the hemisphere the course of the main fissures can be traced; secondly, the great diagnostic point of a foetal lesion, namely, the bizzarre arrangement of the remaining sulci and convolutions, is not present.

It has been pointed out by Dr. Wiglesworth in a description of the case that the seat of the lesion occurs just at a point where the pressure of the forceps at birth would be applied, and he is inclined to assign the cause to severe crushing of the hemisphere with effusion of blood, followed by a slow absorption of the injured parts which

parts which

eventually produced the appearance found.

Another point of some interest is in the large amount of clinical manifestation that was present in this case (paralysis and non-development of the left arm and the talipes of the left foot) compared with the first case M.B., in which there was quite as much destruction of the motor area with practically no clinical manifestations, the ~~latter~~ first case being one of a distinct foetal lesion.

HYDROCEPHALIC IDIOCY.

In bringing Hydrocephalus under your notice it must be clearly understood that it is entirely of the chronic form that I am about to speak; the acute form due to Meningitis, Pressure on the veins of Galen etc., is not included in any of the following conditions.

The disease now known under the name of Hydrocephalus was early recognised among the ancient physicians. Hippocrates, in speaking of Epilepsy, states "For when the disease has lasted long it is curable no longer, the brain being eaten away by fluid and melts; the part thus melted is converted into water which surrounds the brain and bathes it causing the attacks to become more frequent and more easy."

There may be said to be two theories to account for the pathology of this condition; the first and by far the more ancient being that it is due to some obstruction to the flow of cerebro-spinal fluid through the foramen of Majendie, and to the gradually increased intracranial pressure so produced; there are, however, many facts which may be said to place this theory out of consideration, It has been pointed out by Ruffer, in his paper on Hydrocephalus, that the foramen of Majendie is not the only communication between the ventricles and the subarachnoid

subarachnoid

space, the others being the foramen of Bochdalek and the aperturae ventriculi quarti; owing to the differences in their position it is almost impossible to imagine a condition blocking all these communications. Again by far the most convincing proof against this theory is that in quite a number of cases the foramen of Majendie has been perfectly patent, in a few, it has been dilated; in others this foramen has been found completely occluded without any condition of Hydrocephalus existing.

The other theory, and in all probability the correct one is that it is due to too large a quantity of fluid being poured out by the ependyma for the tissues to absorb in the usual way, this excess gradually causing increased pressure and distention of the ventricles. The granulations, etc., on the ependyma go a long way to prove that this is brought about by some inflammatory state. Further, another point in the chain of evidence supporting this theory is, that although the other openings may be sufficient to carry off the normal excess of fluid, the foramen of Majendie being blocked, it is questionable whether the fluid which, say in an inflammatory condition, would be secreted in a large quantity, could pass off even in the event of the patency of the foramen of Majendie; other serious

cavities are found unable at times to absorb the products of inflammatory states; why therefore should the ventricles be able. Turning now to the composition of the cerebro-spinal fluid found in cases of Hydrocephalus, we find that it contains more albumen and salts than that found in normal fluid; it may also contain pus corpuscles and other products of inflammatory changes.

Hydrocephalus has been divided into two clauses, namely, Congenital and Acquired; the latter being by far the more common. West in 103 cases found only 9 that he could call congenital. It has been pointed out by Meynert that it is a comparatively simple matter to differentiate between the two forms macroscopically; he states that in the congenital it is found that the ventricles are dilated in their long diameter, their posterior horns being pushed back in such a way that they come within a few lines of the surface; while in the acquired form they are dilated in their vertical and cross diameters.

As regards the disturbances of intellect, liability to fits (which is often the first sympton of this condition) etc., these are in all probability due to the changes which exist in the cortex of the brain, the result of intracranial pressure.

HYDROCEPHALIC IDIOCY.

Case No.389.

J.H.(male) aet.36. admitted May 28th.1888.

Died June 20th.1893.

Cause of Death:- Colitis, Hydrocephalus internus.

Idiocy with Epilepsy.

On admission:-

He was a quiet inoffensive man. He sat by himself and refused to speak; at times however, he gave a rambling account of his past life. The epileptic fits commenced as far back as he could remember. He never at any time did anything for a livelihood, being kept by his mother.

Later:-

He became an absolute idiot. He was fatuous and unable to take care of himself or take any interest in his surroundings. He was wet and dirty in his habits. The epileptic fits became more frequent.

Post Mortem:-

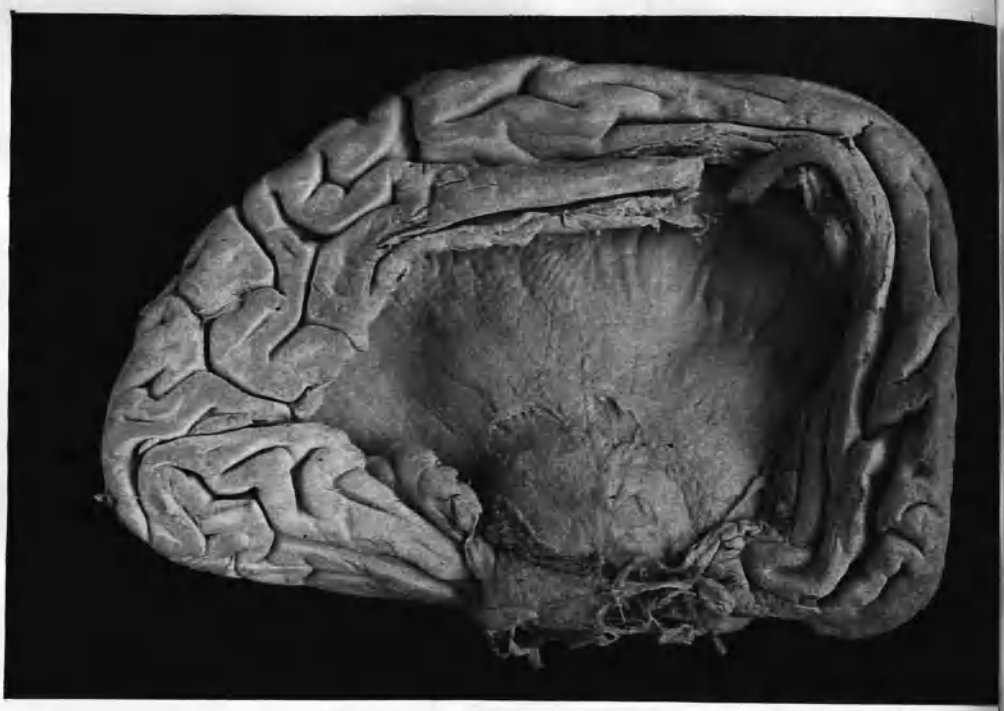
Skull was of large size and symmetrical in shape. The sutures were close.

Encephalon:-

Weight 1590 grammes (with ventricular fluid).

1362 " (without ventricular fluid).

Case No. 389.



Left Hemisphere. Inner Surface.

Note:- The great cavity produced by the enormou
dilatation of the Ventricles, with the alterati
of the entire relationship of the structures.

Right Hemisphere. Inner Surface.

The appearance is similar to that of the above.

ventricular fluid).

Cerebrum:-

The convolutions are all large and everywhere flattened, this is most marked in the anterior half. The convolutions are not to be noted for the complexity, as rather the reverse obtains.

The great superior longitudinal fissure is very shallow. The main sulci are normal in distribution, but they are of a wide gaping character.

The white matter is much reduced in amount, and this is most marked over the anterior horn of the left lateral ventricle, where it is much thinned.

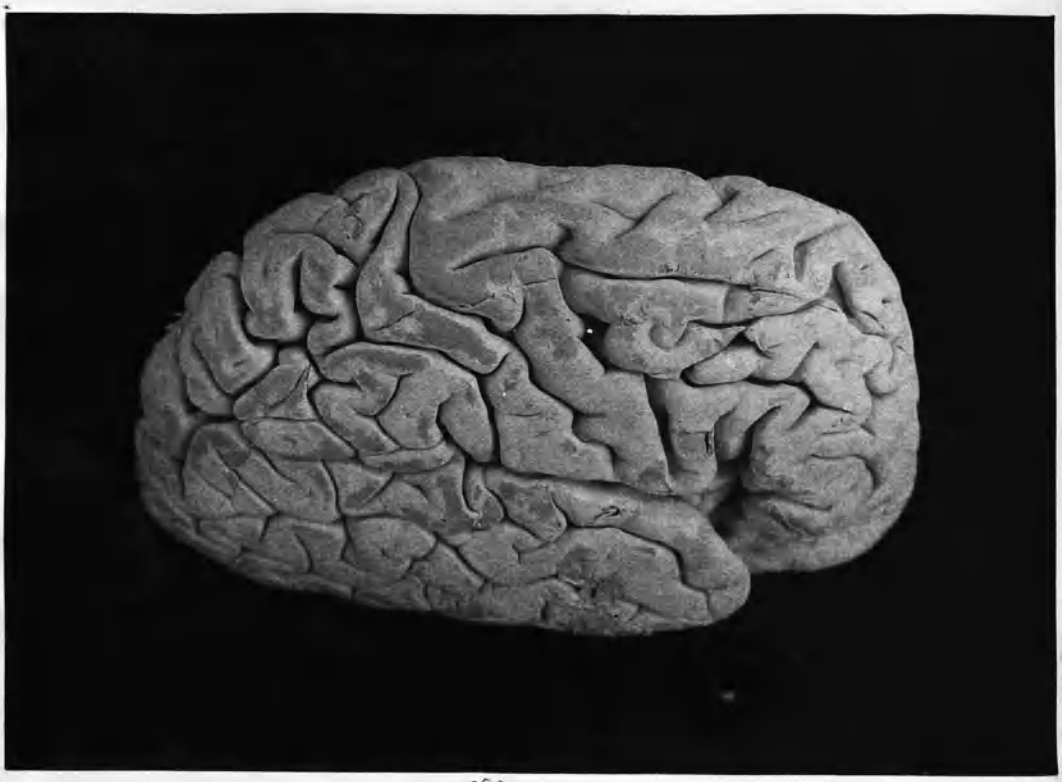
Lateral Ventricles:-

These are enormously enlarged in all directions, the left being considerably larger than the right. The foramen of Monro is annihilated. The ependyma are faintly granular, and numerous whitish annular elevations are seen upon them.

The fornix and corpus callosum are pushed far upwards, the latter being very much thinned and stretched, the former is obliterated apparently at its centre in the vault. The septum lucidum with the attached pillars of the fornix is driven forward and flattened.

The anterior commissure is expanded anteriorly. The Basal

Case No. 389.



389

Right Hemisphere. Outer Surface.

Note:- The greatly flattened appearance of the convolutions. The general simplicity of the convolutional arrangement.

The Basal

Ganglia seem large and flattened.

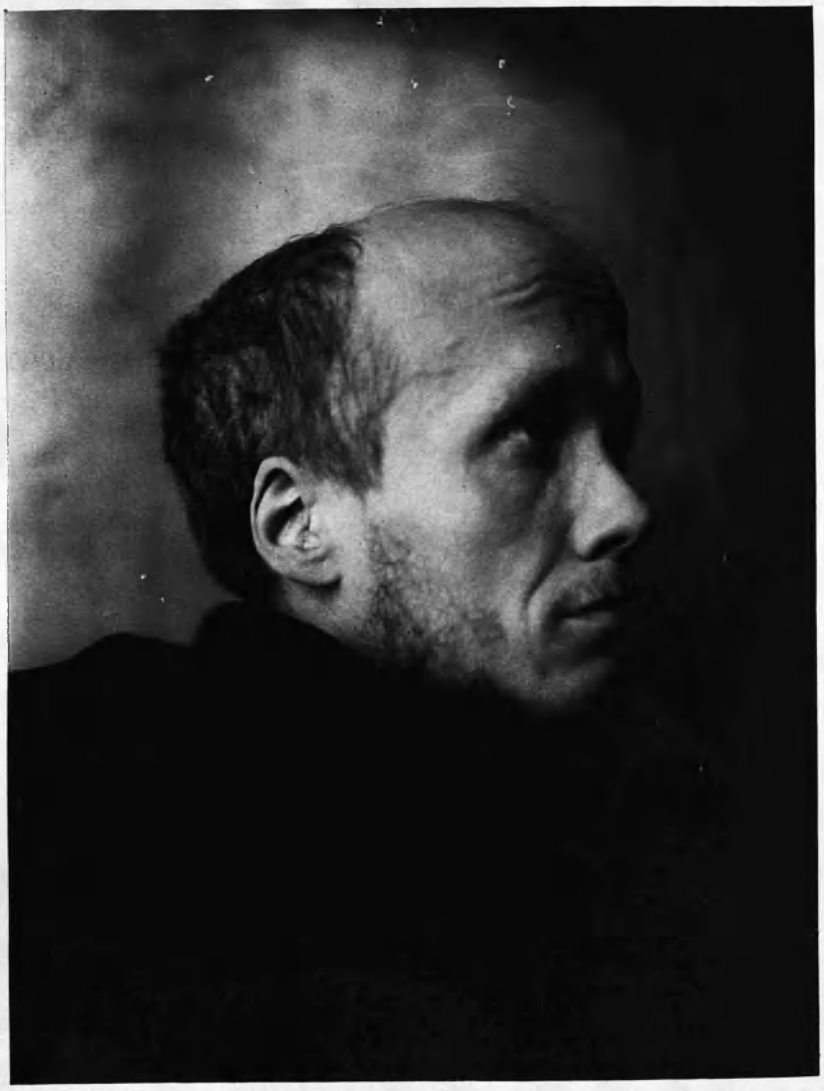
The veins of Galen are very distinct on the floor of the cavity but they are thin and cord-like.

Medulla Oblongata:-

The fourth ventricle appears to be broader than normal and its ependyma are faintly granular.

The Aqueduct of Sylvius is much dilated being almost as large as a goose quill.

Case No. 1526.



D.B. taken a year previous to death.

Note:- The general shape of the skull; the high frontal eminences,etc. are typical of Hydrocephaly.

CASE.No.1526.

D.B.(male) aet.48. adm.Sept.15th.1888.

Died June 29th.1900.

Cause of Death:- Ch.Hydrocephalus and Retro-pharyngeal Abscess.

Idiocy.

History previous to admission:-

He was always irritable and fretful, He never walked. At the age of 18 months he had an attack of convulsions which were followed by fits; however, they ceased at the age of twelve.

On admission:-

He was practically devoid of intelligence and he seemed to understand little that was said to him; although he could articulate one or two simple words.

Later:-

He sat all day in the same position, he could not walk or even stand. He was unable to attend to the wants of nature. He showed no signs of memory or intelligence.

Post Mortem Examination:-

Skull:-

This was typically hydrocephalic.

hydrocephalic.

Greatest Transverse diameter:- 17.5.ctm.

" Antero-posterior diameter:- 21.4.ctm.

The frontal eminences were high and prominent; the sutures close and the lines of the meningeal vessels were easily seen."

Encephalon:-

Weight:- 2479 gms. without fluid 1280 gms.

Cerebrum:-

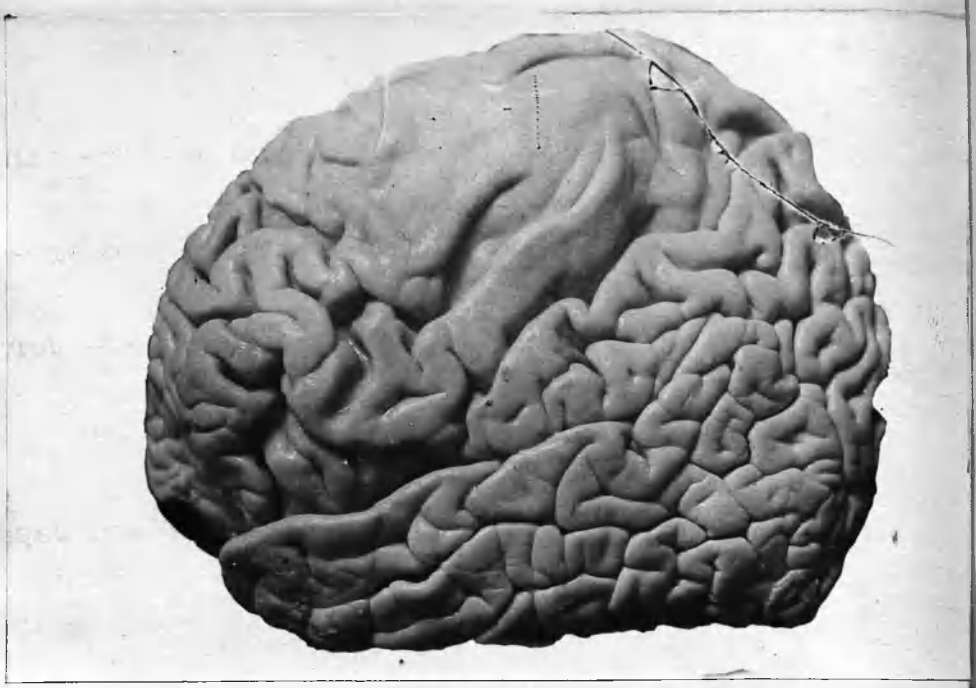
As both hemispheres show practically the same changes, it makes it necessary for them to be described separately.

The cerebrum as a whole is much enlarged and of a rotund character, owing to the great ventricular dilatation; the development appears to be good, and the hemispheres are equal in size and weight.

The arrangement of the convolutions is of average complexity but they are all of a more or less flattened character, this is most marked in the Pre-Rolandic area, where it is almost impossible to map out the continuous length of any of the more important ones; it may almost be said that extending forwards from the Rolandic fissure is an area of about 3 inches square which except for

rudiments

Case No. 1526.



Outer Surface. Left Hemisphere.

Note:- The large"smoothed-out" area anterior to the fissure of Rolando. The dome-like shape of the whole hemisphere.

Inner Surface. Left Hemisphere.

Note:- The enormous size of the Ventricle, with flattening of the Basal Nuclei and thinning of the Corpus Callosum.

rudiments

of the great fissures, such as the precentral, is practically smooth, and in some points, looked at from the ventricular side, translucent.

The sulci are broad and shallow, this again being most marked in the above mentioned area.

The cortex and white matter are very much thinned.

The ventricles are enormous and capable of containing about 1030 cc. pf fluid.

The corpus callosum is much thinned and stretched laterally, this being most marked at a point corresponding to the flattened pre-rolandic area above described. The foramen of Monro is much dilated.

The ependyma are slightly granular.

The cerebellum, Pons and Medulla are rather small but of normal shape.

The fourth ventricle is not dilated but it is markedly granular.

SUMMARY.

These cases present many points of similarity and some of these may be again alluded to.

Taking into consideration Meynert's theory

theory

with regards to the above condition, namely, that when the ventricles are dilated in their vertical diameter the lesion is of extra-uterine occurrence; it will at once be granted that the cases will be classed as ~~congenital~~ acquired.

The enormous dilatation of the ventricles and consequent thinning out of the cortex, with the obliteration of convolutions and sulci, the alteration of all topographical relationship of the parts are characteristic of Hydrocephaly.

Both cases were epileptic, although in that of **B.B.** the fits ceased at the age of twelve.

In both cases granularity of the ventricular ependyma is present, and respecting this, I again call attention to my previous statements on the possible inflammatory cause of this condition.

LOCALISED ATROPHIC SCLEROSIS.

This condition is, on the whole, a fairly common one; in fact, Tauffer, an authority on the subject, states that it and the other conditions akin to the above are the commonest found pathologically in cases of Amentia.

The pathology of the state is fairly simple, and the authorities appear to be for once in a way wonderfully in line with their theories; they are emphatic on the point that the resulting state (i.e. localised sclerosis) is the effect of a number of different processes or changes, of which the causes may be very varied, the commoner, however, being some chronic inflammatory change involving the arterial wall, such as Endarteritis Syphilitica; among others may can be mentioned general Encephalitis, Meningitis, Embolus, etc.

Some of the authorities, notably Jendrassik and Marie, believe that the process commences in the neighbourhood of the smaller arteries and thus causes a number of small areas to be affected which ultimately run together and form the sclerosed portion. On the other hand, others, notably Joseph, Klinke, and Freud, believe and have described cases in which the lesion was due to changes in the state of one of the large Cerebral arteries notably the Anterior and

Anterior and

Middle Cerebrals.

Another common cause is trauma which sets up an Encephalitis as a primary condition which leaves secondarily the Localised Sclerosis; trauma, the results of instruments at birth, has been stated to be a common cause.

We find microscopically that the condition is practically the same as is found in sclerotic states in other tissues, namely a proliferation of the fibrous ground tissue, and, in the case of sclerosis of the cerebral tissue, it is of course the neuroglia which abnormally proliferates; the cells also are affected, being of abnormal shape and showing degenerative changes.

As regards the localisation of this condition, we find that it is by no means constant, any part of the brain may be affected, but by far the commoner seats are the Frontal and Occipital lobes; it is a common concomitant of other lesions, notably around Porencephalic defects and near Softened areas.

CASE.No.1111.

M.G.(female). aet.25. adm.5th.April.1888.

Died May 4th.1898.

Cause of Death:- Abcess of Liver.

Idiocy with Epilepsy.

On admission:-

She was unable to converse, she knew little except that her name was "Mary Gill". She was wet and dirty in her habits.

Previous to Death:-

The fits averaged about 40 per mensem and increased up to 114 per mensem. She gradually became more degraded and was quite unable to do anything for herself.

Post Mortem Examination.

Cerebrum:-

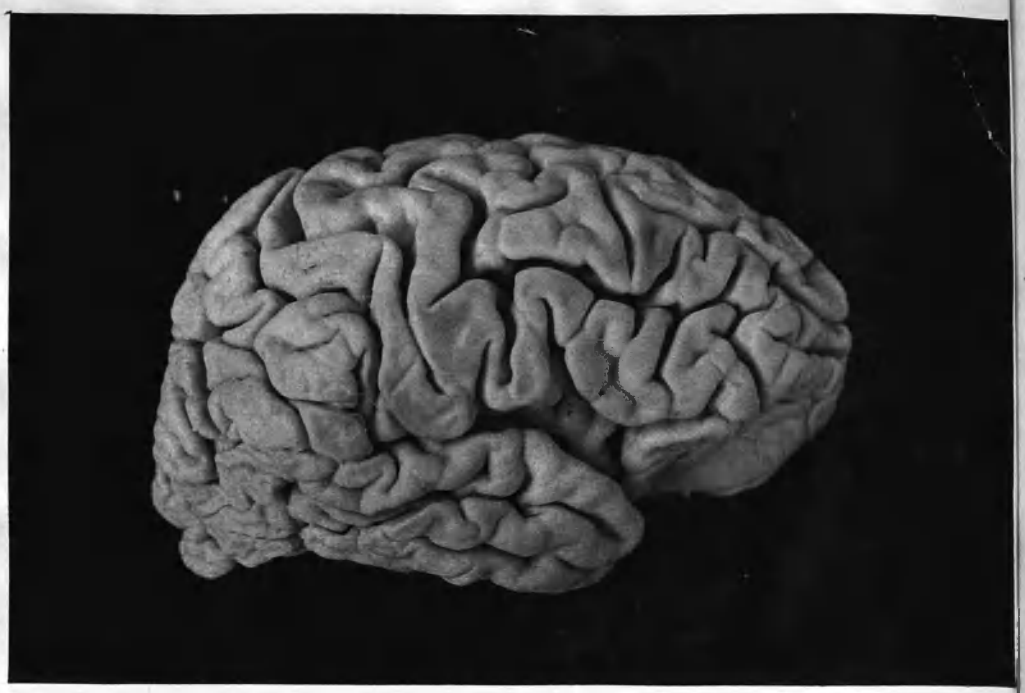
Right Hemisphere:-

Weight:- 315 gms. stripped 296 gms.

Owing to an old standing lesion of the Occipital region of this side, there is pronounced Cerebral asymmetry, the left hemisphere being markedly larger, and the disparity is of course more obvious in the posterior segment.

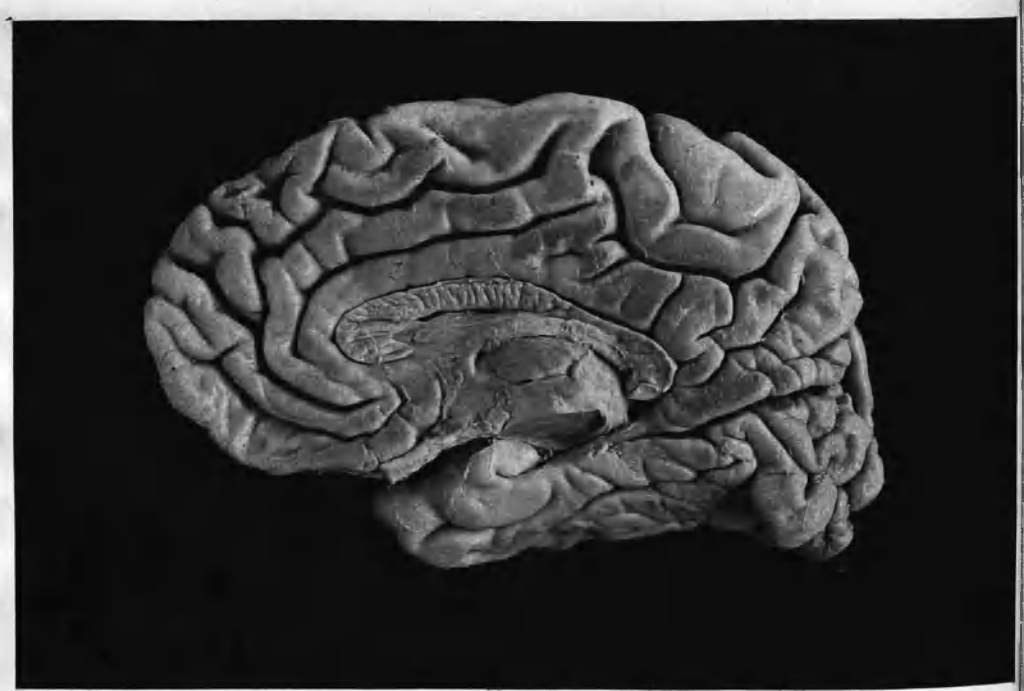
In this hemisphere, viewed from the external aspect,

Case No. 1111.



Right Hemisphere. Outer Surface.

Note:- The atrophied condition of the Occipital and the posterior portions of the Parietal and Temporal regions.



Right Hemisphere. Inner Surface.

Note:- The condition of portions of the Cuneus, Precuneus and the Calcarine areas.

external aspect,

a collection of convolutions embraced in an area composed of the External or Lateral Occipital, the Posterior Parietal and the Posterior Temporal regions is atrophied to half or less than half its normal size, it is shrunk considerably below the surface level and is of a firm leathery consistence. The shape of the convolutions and the arrangement of the dividing sulci is withal preserved, that is to say, there is no complete disappearance of substance.

Analysed more closely, the affected area is bounded in front by the upturned end of the First Temporal sulcus and by a line projected downwards from the bend of that sulcus; above by the Posterior limb of the Intraparietal sulcus and the Upper part of the Anterior Occipital sulcus; behind by the Posterior margin of the Occipital lobe; and below by the Lower margin of the Posterior portion of the Temporal convolutions.

On account of this distortion following shrinkage, and on account of a certain curious arrangement of the Posterior limb of the Intraparietal sulcus, which looks like a sort of reduplication, it is difficult to accurately define the individual affected gyri, but the following appear to have suffered, the Posterior half of the Angular gyrus, the entire Posterior Parietal gyrus, the hinder end of the Second

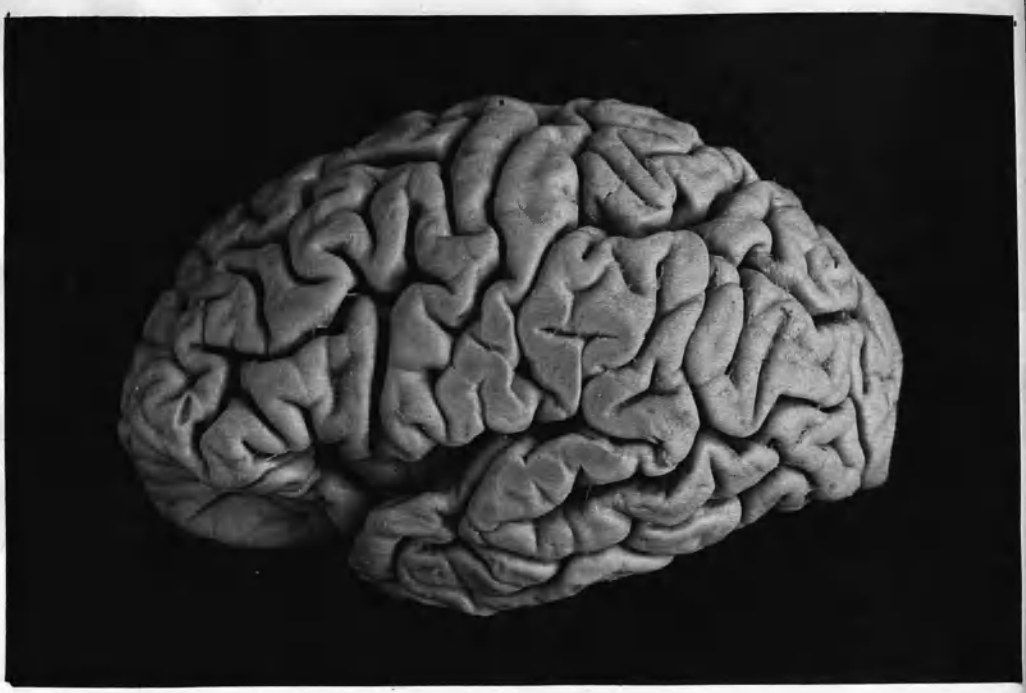
the Second

and Third Temporal gyri, the Superior Parieto-occipital Annectant gyrus (joining the Superior Parietal,) the Inferior Parieto-occipital Annectant gyrus (joining the Posterior Parietal) and the Lateral Occipito-temporal Annectant gyrus joining the Third Temporal, and all the occipital gyri on the outer aspect of the hemisphere.

Viewed from the inner surface the Occipital lobe appears small and blunt, but the only convolution sclerosed and wasted in the same way as those above described is the upper and hinder end of the Cuneus adjoining the upper end of the Parieto-occipital sulcus, but at the same time the remaining convolutions of the Cuneus, and the convolutions bounding the Calcarine fissure below, and that part of the Precuneus bounding the Parieto-occipital fissure are smaller than they should be. It must be further mentioned that along the upper margin of the hemisphere, at about the middle of the First Frontal gyrus, rather on its inner aspect, is a small atrophied and sclerosed focal lesion about 1.5 ctm. in diameter.

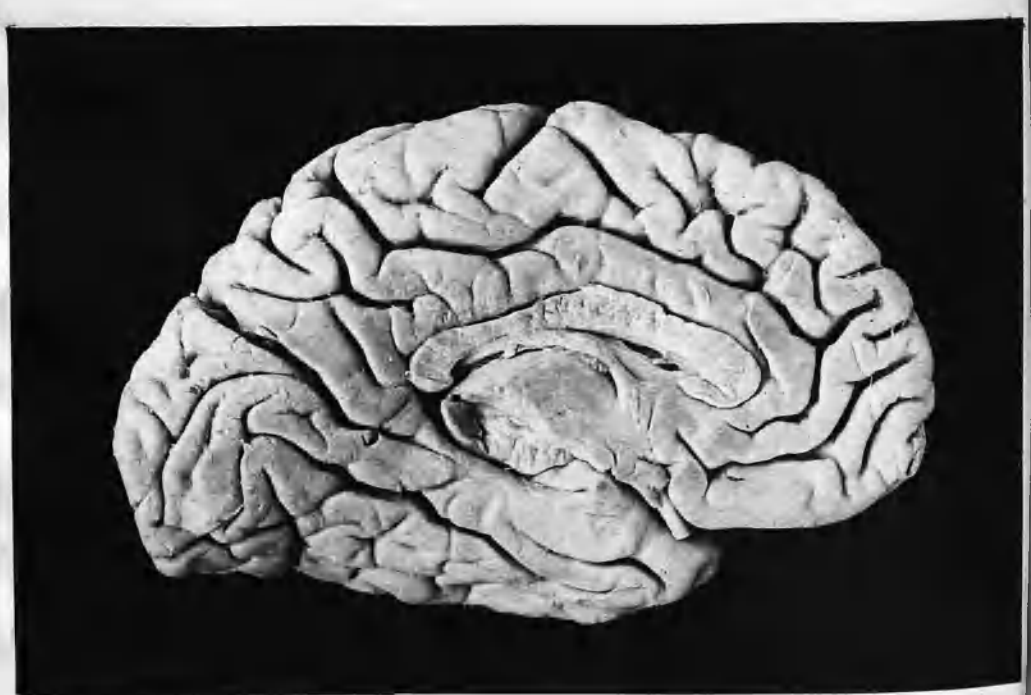
It also remains to be stated that all the remaining unmentioned convolutions of the Temporal lobe, including the Cornu Ammonis and Hippocampus, are markedly firmer in consistence than other healthy parts, although they are not

Case No. 1111.



Left Hemisphere. Outer Surface.

Note:- Some wasting of the anterior third of the Temporal region.



Left Hemisphere. Inner Surface.

Note:- The Precuneus is small.

are not

markedly wasted.

Other noteworthy points concerning the general arrangement of the convolutions are, that the Sylvian fissure is rather wide anteriorly and the anterior convolutions of the Insula are exposed; the Anterior Horizontal and the Anetrior Vertical branches of the Sylvian fissure are indefinitely separated from one another, in broad U-form, making the Pars-Triangularis of the Third Frontal unusually expansive.

Left Hemisphere:-

This hemisphere is free from focal lesions like those described in the right, and its convolutions are likewise richly arranged and small.

The external part of the Parieto-occipital sulcus is curious; in the first place, it is forked, and sends one small vertical projection downwards for 2 or 3 ctm. (an attempt at the "Affensphalte"), and another anteriorly, which runs obliquely forwards through the Superior Parietal gyrus to join the horizontal limb of the Intraparietal.

The Sylvian fissure tends to gape, but otherwise the hemisphere exhibits no noteworthy morphological curiosity.

The Cerebellum is small but there is no noteworthy hemispher-

hemispher-

ical disparity.

The Pons and Medulla are likewise small, but there is no change from normal to be seen externally, and all the Cranial nerves including the Optic and Olfactory are normally represented.

SUMMARY:-

This case is of interest as demonstrating the typical points of Localised Sclerosis; here the area affected is slightly larger than that usually found.

The causation of this state has without doubt been some affection of the Posterior Cerebral Artery, as the area affected corresponds with that supplied by this artery down to the minutest detail.

It is of course quite impossible for us to tell if the process commenced in the minutest twigs of this artery as it is said to do by Jendrassik, etc,. or whether the main trunk of the artery was primary affected, my own opinion being that the latter condition is the more probable.

TUBEROSE IDIOCY.

As regards this form of idiocy, we find that the literature is extremely scanty, and so far as I have been able to gather only a very few observers describe cases, the most prominent being Bournville, Sailer and Gavazzeni.

All the observers, however, appear to be of the opinion that it is a tuberose sclerosis, although Sailer terms it a "hypertrophic nodular gliosis."

As regards the etiology of this state, we find that the question of it being of syphilitic origin arises, and this seems to be the opinion of Bournville, as in several of the cases described by him he places great emphasis on a distinct syphilitic taint, and also in the improvement that was seen in one case after the use of antisyphilitic remedies. Sailer on the other hand looks upon it as due to an hypertrophy of the neuroglia occurring "in utero", and he places no weight on the influence of syphilis.

As regards the morbid anatomy it is much as described in the following case. Numerous whitish nodular bodies are found in the surface of the brain, these consist for the most part of neuroglia; nerve fibres being practically absent. This is practically the finding of all the above mentioned observers; although Sailer lays

lays

stress on the fact that the perivascular lymph channels of the blood vessels in these bodies are much dilated.

The tendency of tumour masses to occur in other organs is also noted, in most of the cases mentioned we find that they occurred in the same organ as in the case I have described, namely, in the kidney.

I would here like to draw attention to the occurrence in my case of the skin eruption known as Ademona Sebaceum; several similar cases to my knowledge have been noted.

Another point is, that in practically all the cases described, we find that epilepsy is a constant clinical manifestation; Sailer examined 28 cases and found them all epileptic.

CASE.No.1442. (male), aet.17.

Adm.Aug.23rd.1889.

Died.Feb.10th.1900.

Cause of Death:- Exhaustion following Epilepsy.

Idiocy with Epilepsy.

On admission:- He could not speak more than a few words, such as "yes" and "no", and then only one at a time. He showed no trace of intelligence or memory. He could not answer to his own name. He was wet and dirty in his habits.

Later:- The fits increased in number, and he became variable in his behaviour, at one time playing with the people in the ward and suddenly without any discernible cause crying and shouting loudly. He became more degraded in his habits than he was on admission.

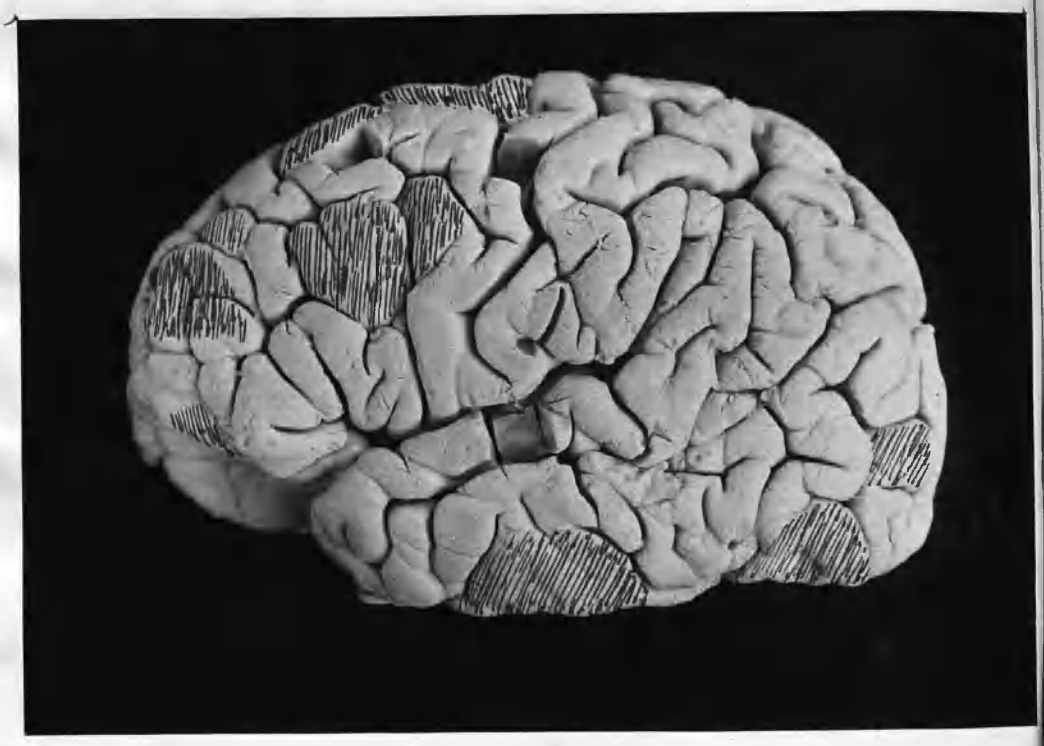
Post Mortem Examination.

CEREBRUM:-

The size and form of the cerebrum is on the whole rather smaller than usual, but of fairly normal shape, hemispheres equal and the segments normally represented.

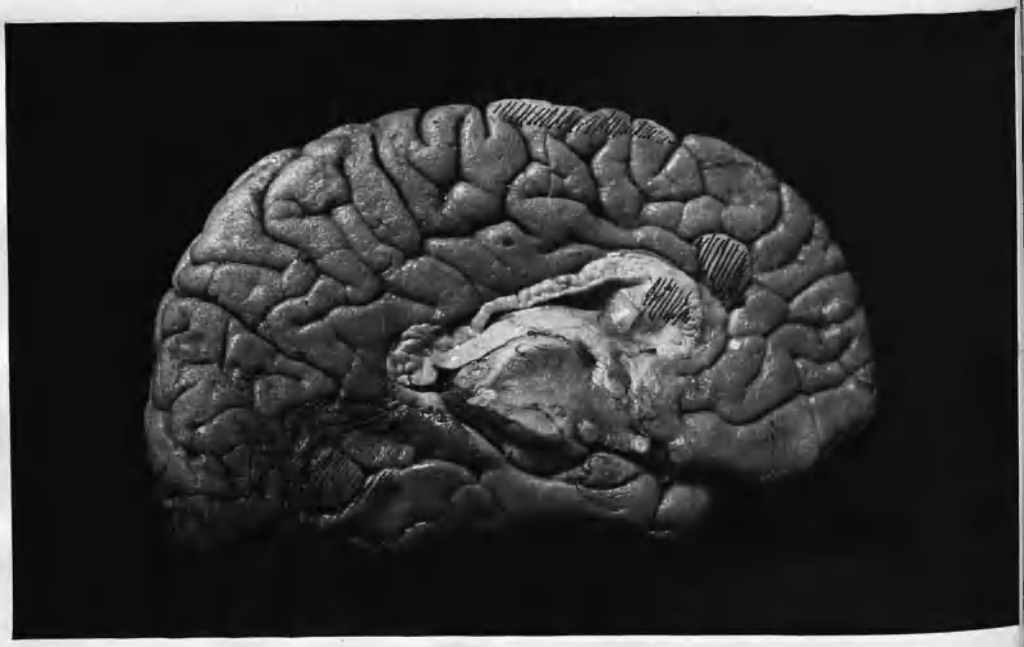
There is nothing of abnormal appearance on first sight, but on touching the surface one finds numerous areas where the substance is of a firm and almost cartilag-

Case No. 1442.



Left Hemisphere. Outer Surface.

Note:- The Sclerosed areas are shaded in red.
(The above shading applies to all the photographs of this case).



Left Hemisphere. Inner Surface.

almost cartilag-

inous consistence, the areas are slightly raised above the surface of the convolutions, and are of a whiter colour than the surrounding cerebral substance.

The areas are sharply defined and the edges bordering the sulci are perpendicular. There are parts where the consistence varies a good deal, between hard and soft, these may be areas of commencing sclerosis.

As regards the exact position of these areas this is best seen from the photographs.

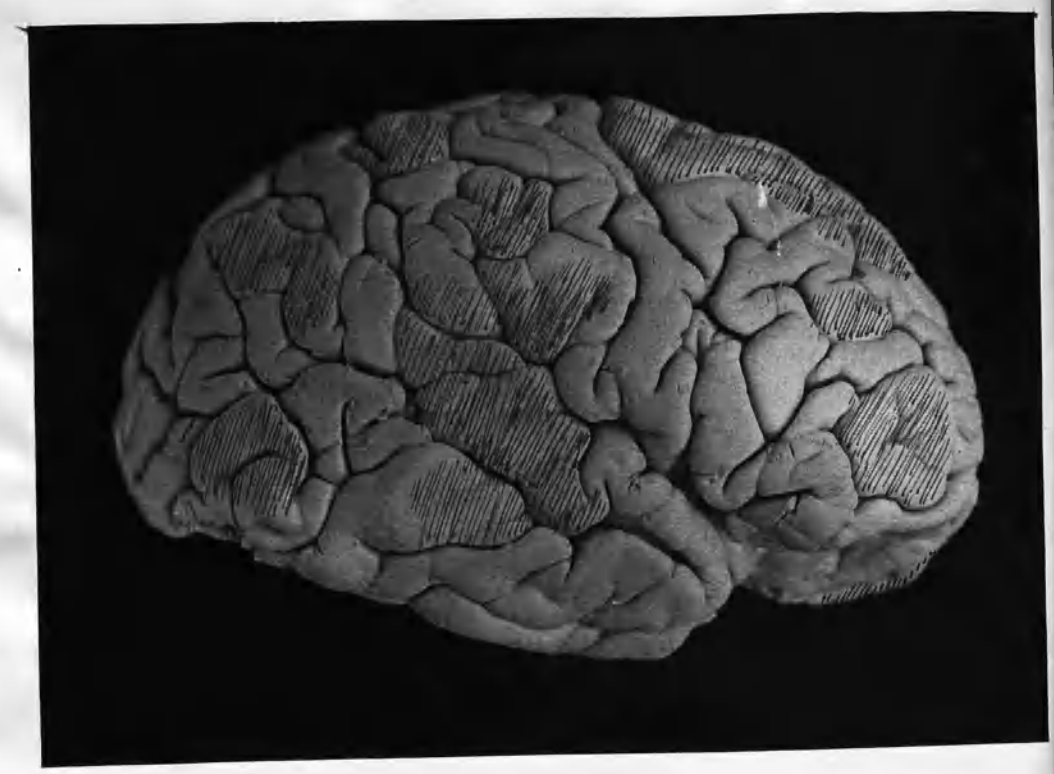
Microscopic Examination of the Cortex:-

Pieces taken from the centre of sclerosed area in upper precentral convolution of left hemisphere, hardened in Formol and stained for nerve fibres.

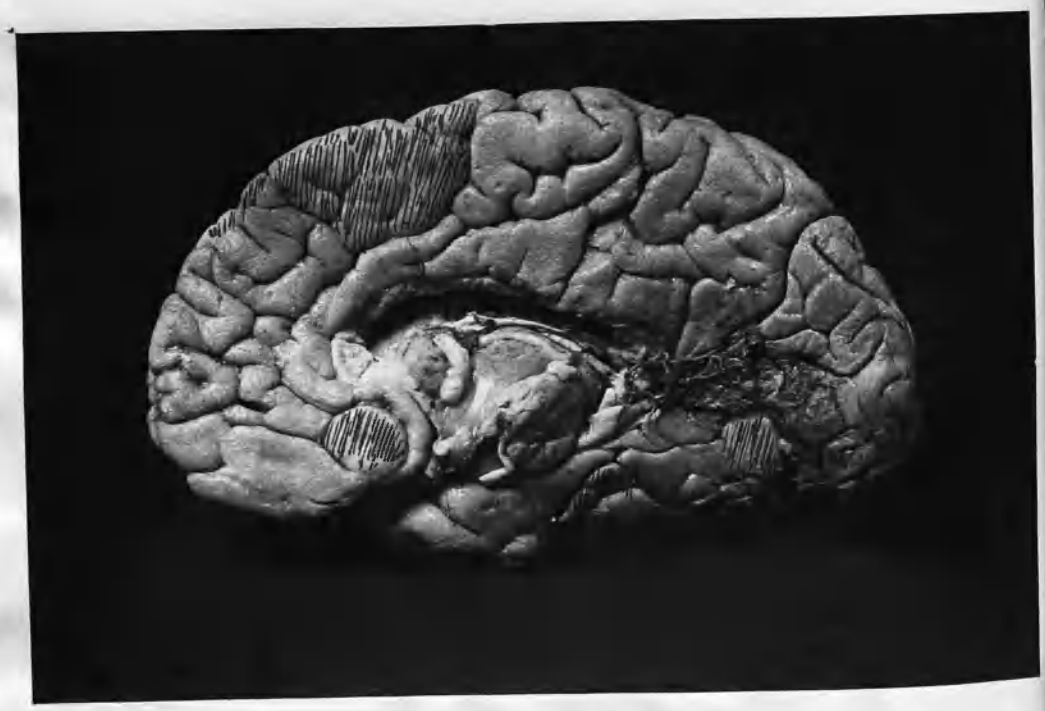
There is a wholesale disappearance of fibres. No zonal layer and no supraradiary fibres are at all recognisable. The line of Baillarger is practically undefinable. Just a few atrophied fibres remain to mark the position of the radiations of Meynert. The association system of fibres is entirely wanting. It may be noted that this is the position where the fibre wealth of the cortex should normally be very great.

In the white substance it is important to observe that the only fibres which remain are the coarse

Case No. 1442.



Right Hemisphere. Outer Surface.



Right Hemisphere. Inner Surface.

coarse

and fine varicose ones, the large medullated ones having entirely disappeared.

It is interesting to note that small fibrous tumours of about the size of a pea were also found in the kidneys of this case, also that the patient was at the time of death suffering from Ademona Sebaceum, the following being the distribtuion of the eruption:-"a large number of small sebaceous tumours were seen on the face extending over the bridge of the nose and the adjoining parts of the cheeks, down almost to the angle of the mouth; they were hard and firm to the touch and felt like minute grains of shot."

MICROGYRIA.

The name Microgyria was given to the condition by Heschl in the year 1878.

Of this interesting condition, we find the views expressed widely divergent; no two observers holding similar views regarding the pathology and causation.

Turning to the views of the more recent observers, we find two great opposing theories expressed, one to the effect that trauma is the essential factor, the other, that the condition is brought about by some form of developmental arrest.

Among the adherents of the first theory may be mentioned Oppenheim, who expresses himself in a most decided manner; he states that the condition is due to a gumming together of the gyri, along with the thickening of the pia, thus concluding that the process originates on the external surface, and that it is due to trauma, possibly occurring at birth. This view is ably supported by Koepen; he likewise gives trauma at birth as a possible cause, and avers that the changes originate in the meninges and spread to the cortex; Bresler is also of the same opinion, holding that the condition is due to an exudative meningitis, caused by pressure on the pia; he also mentions the condition as secondary to purulent meningitis.

meningitis.

The views of the opposing school may be briefly summarised:- Meirzejewski holds that it is directly due to developmental arrest, and he bases his opinion on the grounds that the Insula remains open. (This point, as will be noticed later, I have been unable to corroborate). Otto following on the same lines, states, that the condition is caused by local developmental changes in the foetal white substance and cortex; Binswanger avers, that the grey matter although abundantly present remains in a lower plane of development. Others believe the condition to be brought about by a richer fissure formation of the cortex with a non-development of grey matter.

We find from the following that a great many observers place great stress on the fact that the pia is thickened over the area affected, they also mention that the cortex to the naked eye appears delicately fissured; from my observations of the following cases I am unable to substantiate either of these statements.

In the cases here described, we find some valuable light thrown on the subject; the outstanding point of interest in them is that definite symmetrical areas have been affected, and on this account there can be no doubt that they are properly placed under the heading of Microgyria, which

which

is not to be in any way confounded with the condition known as Localised Sclerosis; this latter we must admit may be due to trauma or even to a meningitis affecting the external grey matter, and it is probably to this condition that the observers allude when they place so much stress on the trauma theory, the same remark applies to the theory that some affection of the meninges is the cause.

Referring again to the view that trauma is the essential factor, and keeping the distribution of the affection in the cases here described well in view, we find that it can be dismissed in a few words; in all the cases one could not imagine any form of traumatic lesion which could affect such symmetrical and wide spread areas as are here touched; and another point which is also well demonstrated here is that in none of the cases did the membranes adhere to the surface of the brain, showing that the meninges had not been affected, at least to any degree.

My own opinion tends towards the developmental theory; I hold that it is either a direct example of agenesia, and case No 1600 is a good illustration of this, showing a failure of development of the neopallium which is the last to appear in the phylogenetic process which marks the superiority of the human brain and harbours the higher as-

higher as-

sociation centres for intellectual faculties; or it may be due, as Porencephaly, to some foetal arterial occlusion, and case No 18. which presents tha best grounds for this theory, has only one weak point in this connection, that being, that the Paracentral Lobules have escaped, while all the rest of the frontal lobes supplied by the Anterior Cerebral Arteries are the subject of this condition; it is, however, quite within the bounds of possibility that these parts have during foetal life some richer supply than the more anterior gyri, and that that supply has saved tham in this case. (Case No. 461. also presents good grounds in support of this latter view.).

As regards the localisation of this condition, we find that most observers state that any part of the cortex may be affected, In my own investigations, while admitting that Localised Sclerosis may be found in any part,I must state that I have never seen nor heard of any case of Micro-gyria affecting the Central convolutions. (Ascending Frontal and Ascending Parietal.).

On the subject of localisation of this condition, the evidence gathered from the succeeding cases points most clearly to the fact that only the later-developed portions of the brain (Higher Association Centres) are affected by

affected by

this condition, and that the Central Convolutions which are laid down earlier escape. So far as I am aware no writer has previously called attention to this fact, and it seems to me of great importance as a link in the chain of evidence in support of the developmental theory of the condition.

As regards the varieties of Microgyria, we find that Otto differentiates three classes:-

1. In which the general development of the convolutions is on the whole good, but on the surface are numerous holes or depressions.

2. In which the convolutions are not developed but simply form little elevations on the white substance.

3. In which the white substance is not developed and the convolutions in section appear like a "stag's antlers."

CASE No. 1600.

E.M.(female), aet.39. adm.Nov.26th.1878.

Died:- Jan.14th.1901.

Cause of Death:- Status Epilepticus.

Idiocy with Epilepsy.

History previous to admission:-

She was healthy and intelligent up to the age of sixteen.(she could then converse).At this age she fell from a step-ladder and struck her head on the fender; she was not at any time unconscious, but next day she could not speak. She shortly after this became restless and destructive, ~~she~~ and had on this account to be placed under restraint. She had her first fit a fortnight previous to admission.

On admission:-

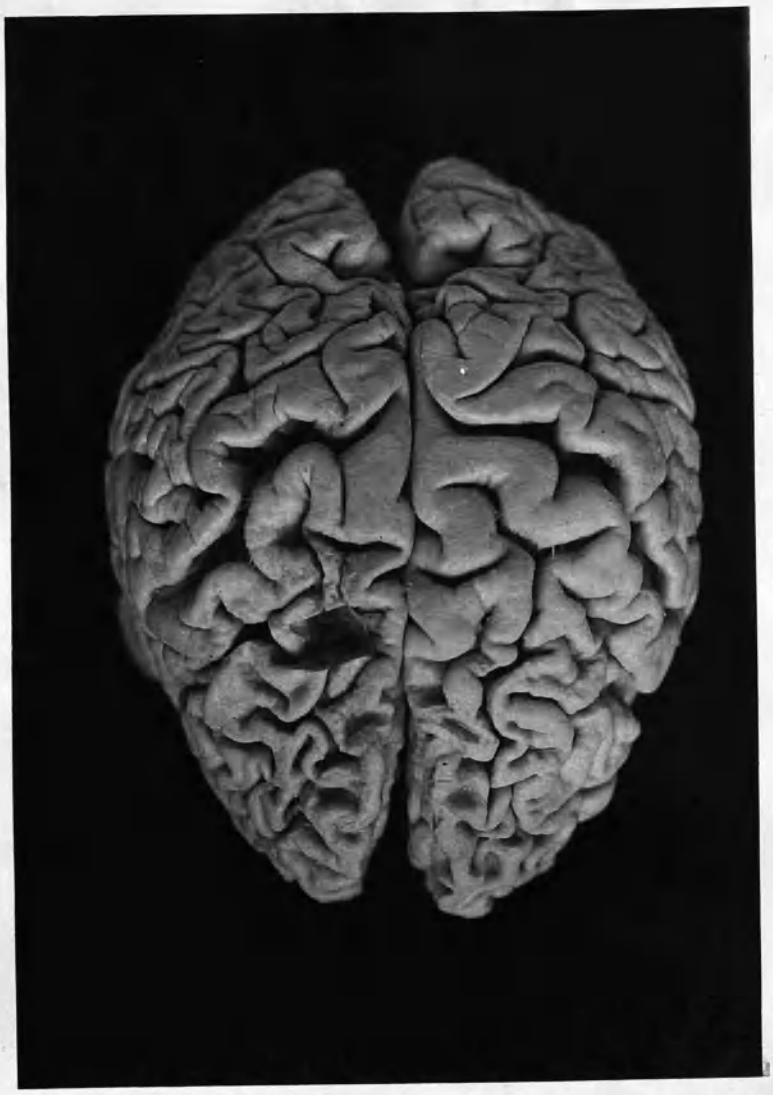
It was noticed that her left arm and leg were smaller than the right. There was also a distinct loss of power in the same.

She was practically devoid of intelligence; she could not speak; she was continually cramming all kinds of rubbish in her mouth, and she was very destructive. She was wet and dirty in her habits.

Later:-

She became more degraded, kicking and biting all

Case No. 1600.



1600

View of both Hemispheres from above.

Note:- The Central convolutions are the only ones which are practically normal in size, elsewhere the condition of Microgyria is obvious.

biting all

within reach.

The fits increased in number and averaged shortly before death 41 per mensem.

Encephalon:-

Weight 920 gms.

The Cerebrum exhibits remarkable general alterations, of which the most important lies in the Frontal region. All the convolutions of the Frontal lobes on both sides with the exception of the Ascending Frontal convolutions are greatly reduced in size. Posteriorly both hemispheres are pointed in form, due to a microgyrous condition of all the Occipital and Parietal convolutions, save the Ascending Parietal. A special defect in the position of the Superior Parietal convolution and Precuneus becomes evident when the hemispheres are viewed from above and behind. In other words, when viewed from the sides, the Central convolutions and the Temporal lobes, although (being) of more or less normal size, are prominent, whereas the rest of the convolutions on the lateral surface are defective in size. Viewed from within, the Marginal gyri and Precunei are defective, while seen from below the changes are not striking.

Right Hemisphere:-

Right Hemisphere:-

The Main fissures may be said to follow a fairly normal course and to occupy a normal position, although, that of Sylvius tends to gape anteriorly owing to the small size of the Frontal Operculum.

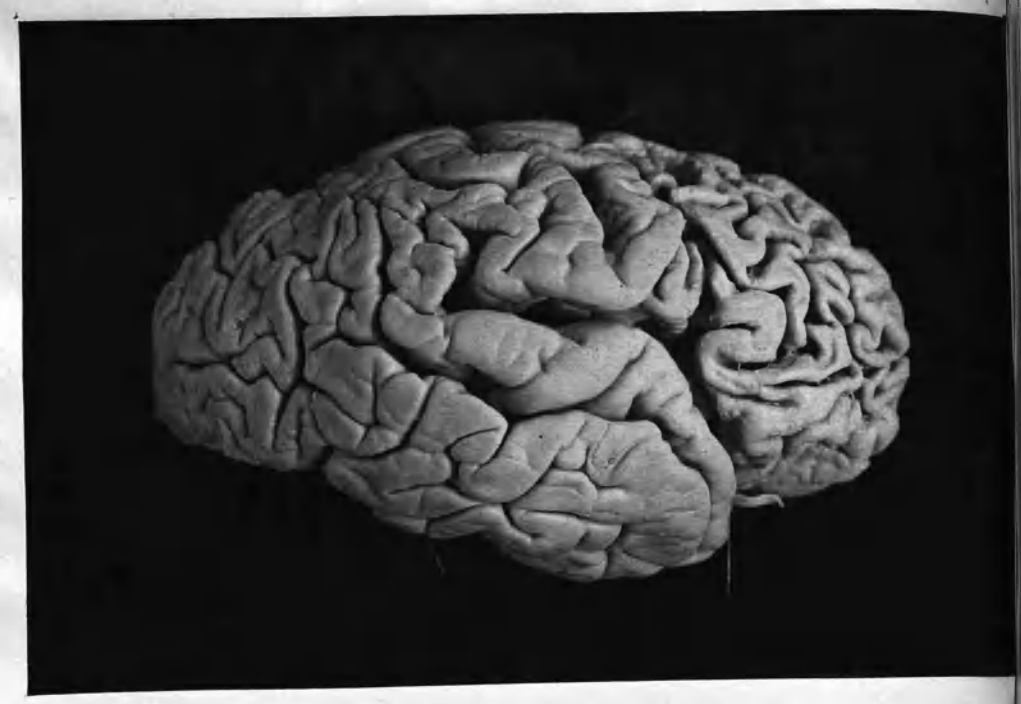
Frontal Lobe:-

In the Frontal lobe as seen from the side, there is hardly a single gyrus, excepting the Ascending Frontal, which is not in an extremely microgyrous state, although some are more affected than others. The gyri may be described as worm-like; they are considerably firmer than normal, and the sulci all gape and are shallow.

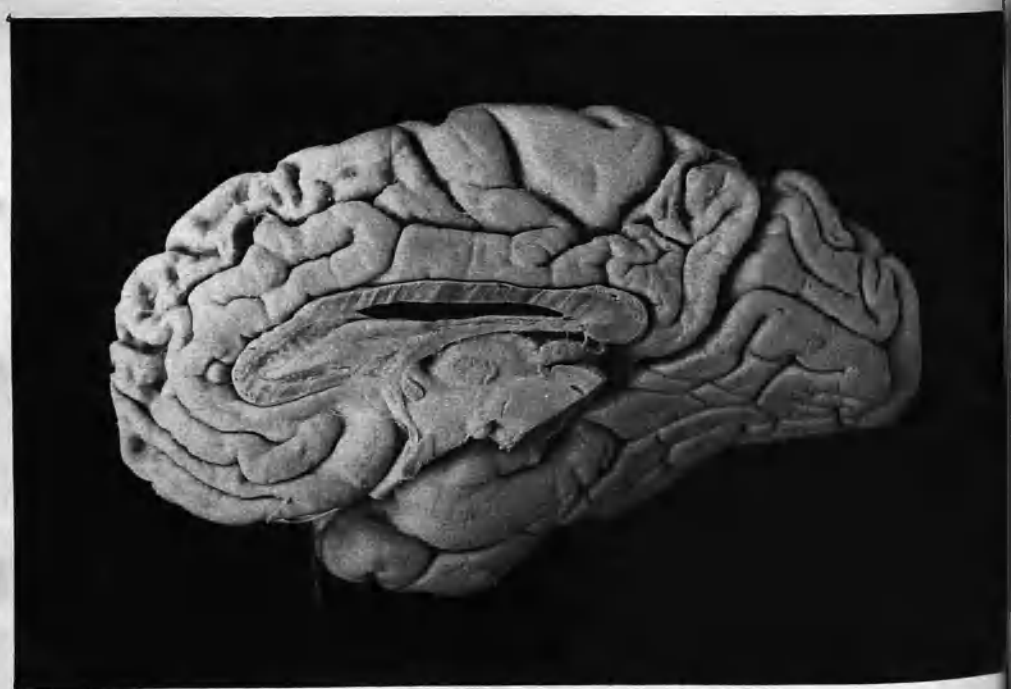
The first or Superior Frontal Convolution:- On the lateral aspect the entire convolution is withered, but that part which lies along the upper margin of the hemisphere is less affected than that which borders the First or Superior Frontal sulcus. In its posterior third it is divided horizontally into two portions by an attempt at a mesial sulcus, but in front of this, as many as six sulci cut it transversely; three of these are isolated, and three join the First Frontal sulcus.

The Second or Middle Frontal Convolution:- The entire convolution is again withered, but that part bordering the Second or Inferior Frontal sulcus is less affected than the

Case No 1600.



Right Hemisphere. Outer Surface.
Practically the whole of the Frontal and Occi
ital Lobes are in a condition of Microgyria. T
Parietal Lobe is similarly affected but to a
less degree.



Right Hemisphere. Inner Surface.
Note:- The very marked shrinking of the Frontal
and Precuneate convolutions, with the subjacen
portions of the Limbic Lobe.

than the

upper part.

The Third or Inferior Frontal Convolution:- This also joins in the atrophy, but the Orbital Operculum is less wasted than the "partes triangularis et basilaris," and immediately anteriorly to the horizontal Sylvian limb is a portion of convolution which is in a fairly healthy condition.

The Ascending Frontal Convolution:- Although much larger than the other described above and of normal consistence, it is considerably below the normal size.

On the Mesial Surface, the entire Marginal gyrus is moderately withered, like that portion of the First Frontal convolution bordering the upper margin of the hemisphere. The Paracentral Lobule, although perhaps smaller than normal, stands out as being in a more or less healthy condition.

Parietal Lobe:-

This, saving the Ascending Parietal convolution, which,although considerably below the normal size, is like the Ascending Frontal of more or less healthy appearance, is of reduced size and its component parts are microgyrous,but to a less extent than the Frontal Convolutions, and here again certain parts have suffered more than others. That part of the Superior Parietal Lobule which joins the Precuneus is severely affected, while the rest is less involved.

less involved.

The Supra-Marginal gyrus is on the whole relatively little affected, but its convolutions are smaller than the Ascending Parietal.

The Quadrate Lobe is in its entirety as completely withered as any portion of the Frontal Lobe; it measures only 20 x 30 millimetres. The sulcus dividing it from the Post-limbic lobe is not well-represented. This atrophied Pre-cuneus, coming as it does between a more or less healthy Paracentral lobule and Cuneus, forms a striking feature when the brain is viewed from within, and as the subjacent portion of the Limbic lobe is similarly affected, this feature is much accentuated.

Occipital Lobe:-

While all the convolutions of the Occipital lobe are of much reduced size, it is only those on the outer surface which are specially affected like those of the greater part of the Parietal lobe.

The convolutions composing the Cuneus are of good consistence and approximately healthy.

Temporal Lobe:-

This stands out as being almost free from Micro-gyri.

The First and Second Temporal convolutions are

convolutions are

practically normal; the Third is also more or less normal in its anterior two thirds, but in its posterior third it is unduly firm and atrophied like the adjoining External Occipital convolution. The Fourth and Fifth Temporal convolutions take no share in the microgyrous state.

Insula:-

This so far as can be seen is normal.

Left Hemisphere:-

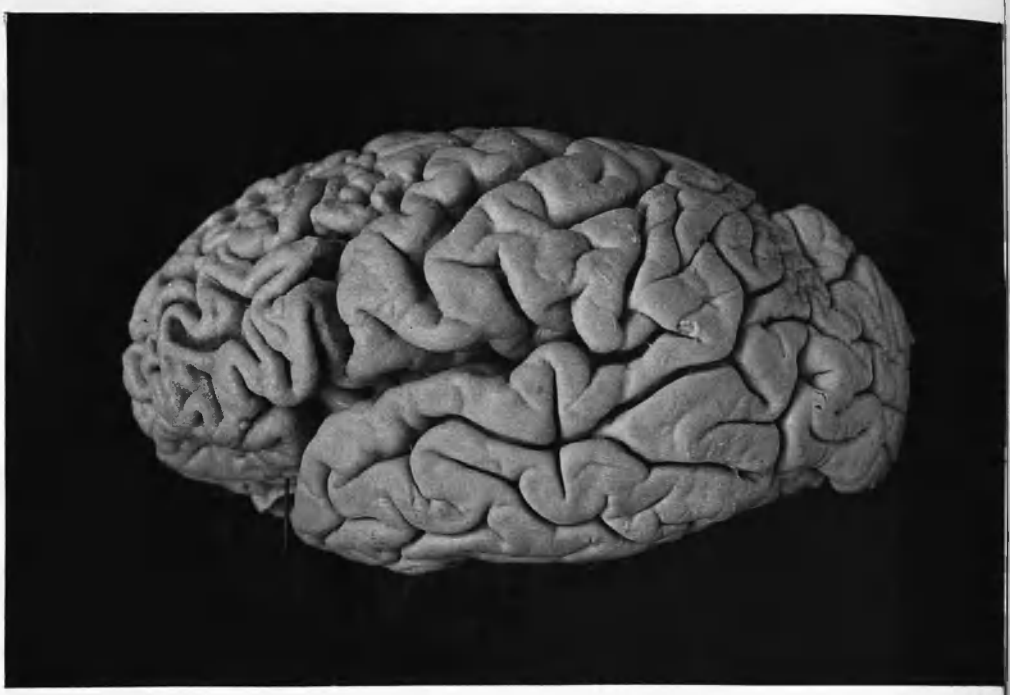
On the whole the left hemisphere is less affected than the right. The Main fissures are practically the same as those of the right.

Frontal Lobe:-

The First Frontal convolution for its posterior 25 to 30 millimetres is in a more or less healthy condition, the remainder (lateral surface) is extremely microgyrous, as it was on the opposite side; it must be noticed that where the healthy and diseased areas join, occur islets of substance which are not wasted, and these may be completely isolated.

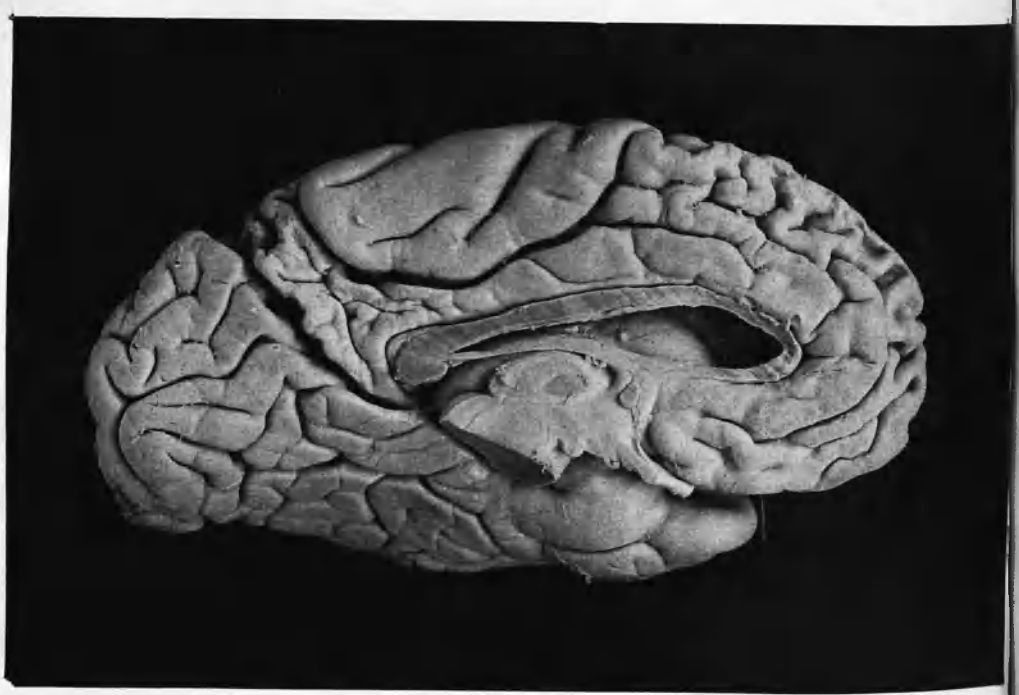
The Second Frontal convolution for a very small portion is healthy; the next 12 to 15 millimetres is wasted and puckered, next comes an islet of fairly healthy substance

Case No. 1600.



Left Hemisphere. Outer Surface.

The condition of parts is similar to that seen in the Right Hemisphere



Left Hemisphere. Inner Surface.

The wasting of the Quadrate Lobe is extreme

substance

forming the upper lip of the Ramus Horizontalis of the Sulcus Precentralis Inferior: the rest of the convolution is extremely microgyrous.

On the Third Frontal convolution there exists a separate Sulcus Diagonalis, and anteriorly to this sulcus the whole of the convolution, including the Pars Triangularis and Pars Orbitalis, is considerably reduced in size, somewhat puckered and unduly firm. That part of the Operculum lying behind the Sulcus Diagonalis and also the whole of the Ascending Frontal convolution is practically unaffected.

On the mesial surface the whole Paracentral lobule is again untouched, but the whole Marginal gyrus, with the exception of a small portion annexed to the Paracentral lobule and that portion at the other extremity lying next to the Optic nerve, is microgyrous.

The Orbital convolutions are practically the same as those of the right hemisphere.

Parietal Lobe:-

The whole Ascending Parietal convolution is again untouched, and so is the Supra-marginal convolution, but of the Superior Parietal lobule, that portion bounding the Ramus Horizontalis of the Intra-parietal sulcus, the portion forming the anterior lip of the so-called External Parieto-

Parieto-occipital fissure and also the portion annexed to the Precuneus are extremely microgyrous and depressed below the general surface level.

Just as on the right side, the whole Precuneus and the whole of the subjacent posterior Limbic lobe right down to the Corpus Callosum and the stem of the Calcarine fissure are extremely microgyrous and again depressed below the general surface level.

The Precuneus only measures 12 mm. in breadth. The whole of the Angular gyrus, save a small portion which is annexed to the Supra-marginal gyrus, is shrivelled, and so is the adjoining part of the Posterior Parietal convolution.

Occipital Lobe:-

On the whole the Occipital lobe is of fair size and the external convolutions, although small in size, are much less affected than those of the opposite hemisphere.

The Cuneus is of fair size and entirely escapes microgyrous changes.

Temporal Lobe:-

In this hemisphere the whole of the Temporal lobe escapes affection.

Insula:-

The convolutions of the Insula are well represented

well represented.

Limbic Lobe:-

This, as on the right side, although rather small all along, is specially microgyrous in its posterior portion.

The Corpus Callosum is thin all along.

Anterior and Posterior Commissures are present and all other structures found normally in this region are represented, and it can only be said of them that they are small.

The Anterior Horns of the Lateral Ventricles are markedly dilated; the other horns cannot be seen.

Cerebellum:-

It is interesting to note that portions of this organ have undergone changes precisely the same as those just described in the Cerebrum.

Thus on the upper surface of the left hemisphere, mainly affecting the Lobus Clivi, is a strip 35 mm. broad by 12 mm. deep, in which the folia are much atrophied and of increased firmness. Also bordering the great horizontal fissure posteriorly, parts of the convolutions show similar although less marked changes.

The Pons and Medulla though small actually, are of relatively normal size and free from special peculiarity.

SUMMARY.

Taking into account the clinical history of this case, it is quite impossible to assign the cause of the condition to the fall, sustained at the age of sixteen. It might be that those observers who place stress on the " trauma causation " would quote this case in support of their view, but it must be perfectly clear to all that no trauma could cause such a change in so large a surface, and, in what are to all practical purposes definite symmetrical areas, and which pathologically showed no signs of a meningitic affection.

This case, as has already been mentioned, points out very clearly the way in which the Central convolutions seem to escape this affection, and it is on this account, in my opinion, one of the strongest cases yet described supporting the developmental theory, as here we have those higher centres, which are so characteristic of the human brain, and which place it on a level so much higher than that of the lower animals, entirely affected.

It is not to be wondered at, that before death the patient led simply an "animal existence" as only the "animal centres" remained healthy.

Another point of interest is that the Lateral Ventricles (Anterior Horns) were dilated, this fact according to Otto being common in this condition.

condition.

This case is the only one in this collection in which there is the slightest tendency for the Insula to remain open,- the Frontal Operculum being small - a state of matters which Meirzejewski states is common and the grounds of which he bases his views that the condition is due to a developmental arrest.

It is also interesting to state, en passant, that microscopically, the Interradiary Plexus is fairly rich in thin varicose fibres but very deficient in transverse association fibres, and the disappearance of the association system may be regarded as the most important point brought out in the sections, which were taken from the first Frontal convolution, at a place severely affected, and stained for nerve fibres by the method of Wolters-Kulschitzky.

CASE.No.18.

S.C.(female) aet.17. adm.19th.July.1889.

Died:-5th.May.1891.

Cause of Death:- Phthisis pulmonalis.

On Admission:-

She can only articulate a few words, and is incapable of answering even the simplest questions. She has no power of walking and can hardly crawl. She sits all day in the same position, and gives little or no trouble. She is wet and dirty in her habits.

Later:-

She became if anything more degraded, and died in a state of Absolute Amentia.

Post Mortem Examination.

Skull:-

There was marked flattening in the Frontal and Temporal regions. The thickness was also much increased and consisted for the most part of cancellous tissue of a deep red colour, this was most marked in the region of the Orbital plate on each side of the Ethmoid bone where the bones were raised quite half an inch, and received the brain in a deep furrow corresponding to the Ethmoid.

Ethmoid.

The floor of the Basal fossae were also markedly irregular.

Encephalon:-

There is a marked general roundness of the whole brain viewed from above; the convolutions of both Frontal regions, the inner surface of both hemispheres and the Left Occipital region are considerably diminished and altered.

The Cerebellum is of comparatively large size. The Medulla and Pons are small.

Right Hemisphere:-

Frontal Lobe:-

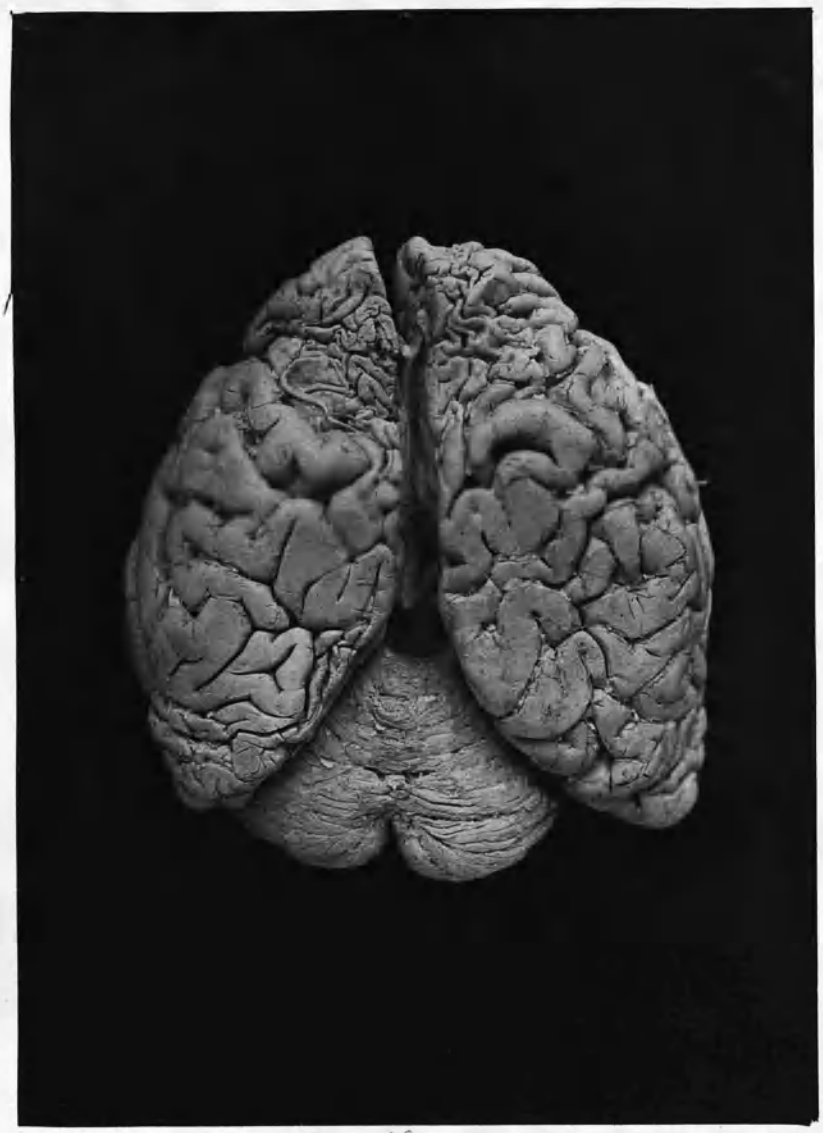
The First Frontal convolution measures about 5 ctm. in length, the upper third of which is of fairly round ed shape, the remaining two thirds are atrophied and withered; the shape of the convolution, however, is retained, although it is very much reduced in size.

The Second Frontal convolution measures about 4ctm. in length, and in the upper fifth of its course, is much the same as the upper part of the First above described, the atrophy is, however, more marked; the shape etc,. of the convolution is retained.

The Third Frontal convolution is composed of a series of parallel gyri lying almost at right angles to the

Case No. 18.

Left.



Right

18

View of Encephalon from above.

Note:- The condition of the frontal convolutions of both hemispheres. The condition of the left Occipital region.

to the

First and Second, at the lower third there is a somewhat better developed convolution which partly occupies the region of the Second Frontal.

At the Anterior margin of the hemisphere the convolutions are very much atrophied in the region of the First and Second Frontal but on turning the edge the convolutions rapidly increase in size.

The Orbital surface is peculiar in that the Outer two thirds are small, well-formed and regular, but the inner third, occupying the Internal convolution chiefly and parallel to the Mesial sulcus, projects as a ridge beyond the plane of the rest of the surface, by about two-thirds of an inch.

The Ascending Frontal convolution is well-developed, and the limiting sulci are quite distinct.

The Paracentral lobule, as far as can be seen without destroying the specimen, is not in any way affected.

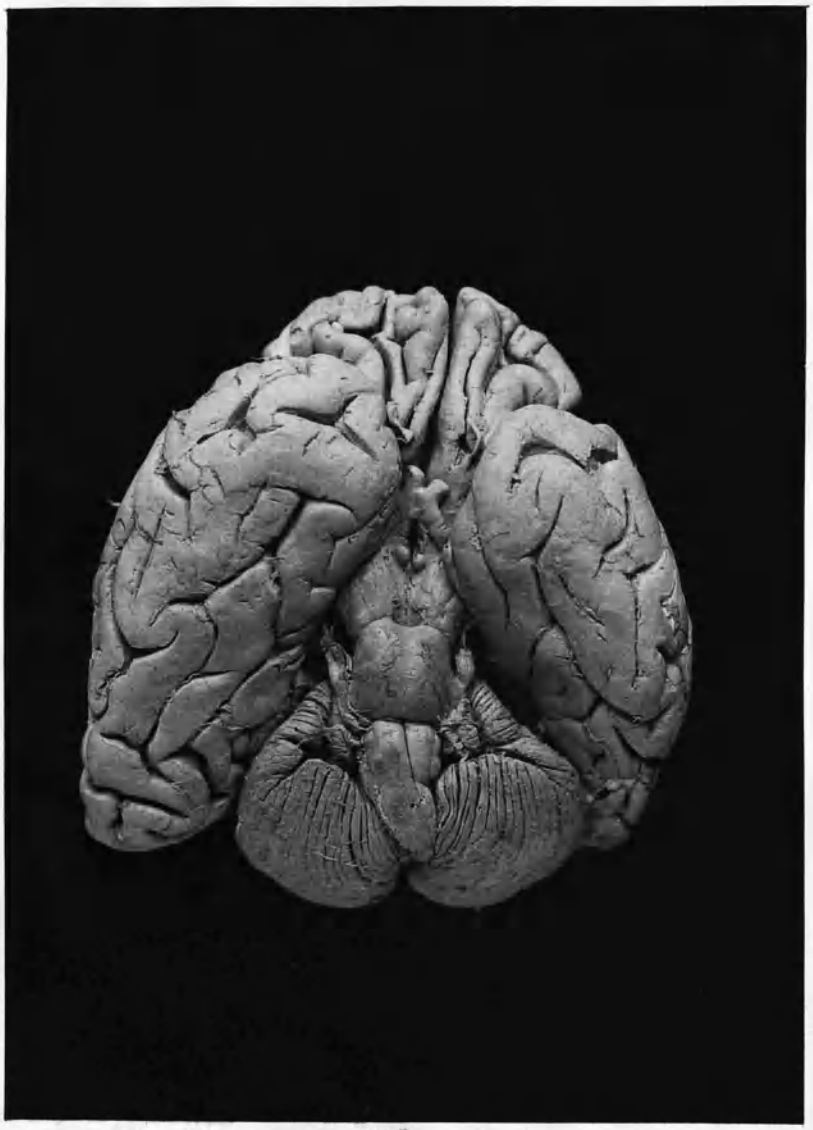
Parietal lobe:-

The convolutions, on the whole, in this region are of fair size but inclined to be irregular and intersected unduly by secondary sulci.

Occipital Lobe:-

In the right hemisphere there is little to be

Case No. 18.



18

View of Encephalon from below.

Note:- The Orbital ridges. The Cerebellum is large in proportion to the Cerebrum, while the Pons is small.

normal.

The Orbital Surface is practically the same as that of the right side, except that the ridge does not stand out in such a marked manner.

Parietal Lobe:-

This is practically the same as that of the right hemisphere.

Occipital Lobe:-

This is marked by great reduction in size, being considerably smaller than that on the right. The distribution and course of the convolutions does not vary in any marked degree from normal.

There is nothing of interest in either of the Temporal lobes.

SUMMARY.

The condition of this case is one which I can put forward with confidence as one due to some form or other of arterial occlusion. It also demonstrates very prettily the point which I have drawn attention to all through this chapter, namely, the peculiar manner in which the Central Convolutions excape this condition; here the Paracentral lobules escape, although sharing practically the same blood supply as the more anterior affected gyri. Again I fail to understand how any form of trauma could cause this condition.

CASE.No.461.

M.H.H.(female), aet.18. adm.Sept.10th.1891.

Died:- Jan.13th.1894.

Cause of Death:- Phthisis pulmonalis.

Imbecility bordering on Idiocy.

On admission:-

She was very dull and could give no account of herself. She rambled in conversation and took little interest in her surroundings.

Previous to Death:-

Her power of attention was poor; she had little idea of time or place. At times she was irritable and obstreperous without any discoverable cause. She had no memory and took no interest in associated entertainments.

Post Mortem Examination.

Encephalon:-

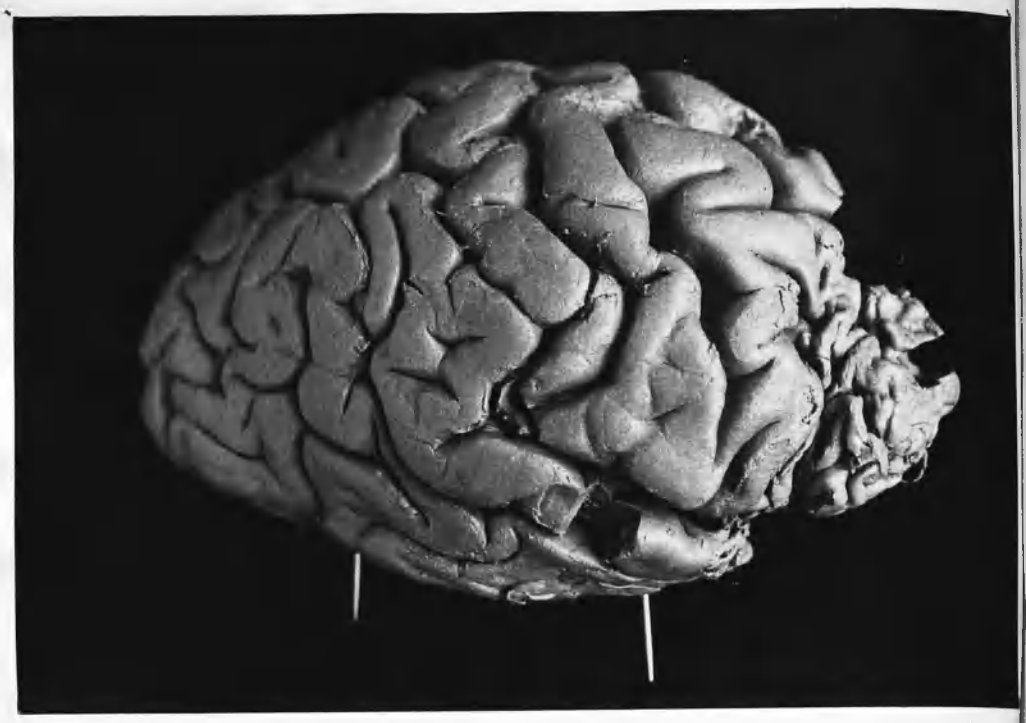
Weight 990 gms.

CEREBRUM:-

Right Hemisphere:-

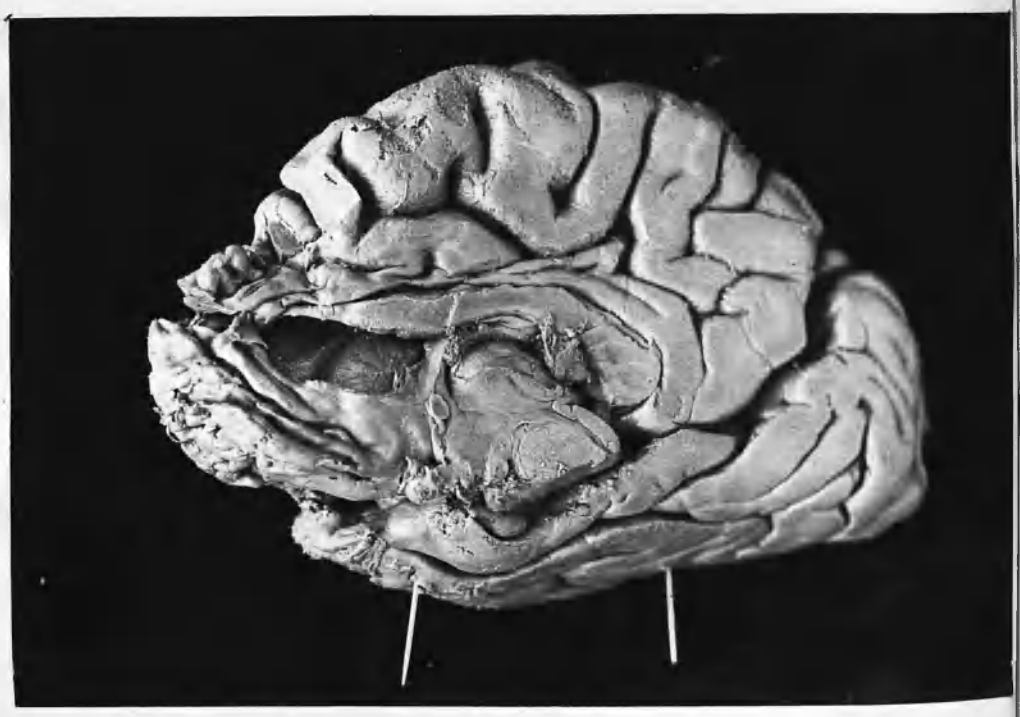
The entire Frontal segment with the exception of the Ascending Frontal, a small part of the adjacent Second Frontal and the Posterior two inches of the First Frontal convolutions is of extremely small size and worm-like in ap-

Case No. 461.



Right Hemisphere. Outer Surface.

The Frontal convolutions are represented by a small worm-like mass. The tip of the Temporal Lobe is similar.



Right Hemisphere Inner Surface.

Note:- The appearance of the tip of the Tempora Lobe, the dilated Ventricle, the approaching di appearance of the anterior extremities of the Gyrus Fornicatus & the Corpus Callosum

worm-like in ap-

earance but of firm consistence, with the exception of the central part which is semi-transparent in colour and of a gelatinoid consistence.

The corresponding part of the Coppus Callosum is greatly reduced in size and thickness.

The Anterior Cornu of the Lateral Ventricle is dilated and the white matter as well as the cortex is obvious -ly thinned.

A like change has affected about one and a half inches of the anterior ends of the First and Second Temporal convolutions.

The remainder of the hemisphere is fairly well developed and to all practical purposes normal.

The Sulcal arrangement is a simple one. The Intra-parietal fissure joins that of Sylvius. The Parieto-occip-ital is deep and the Calcarine joins the Collateral.

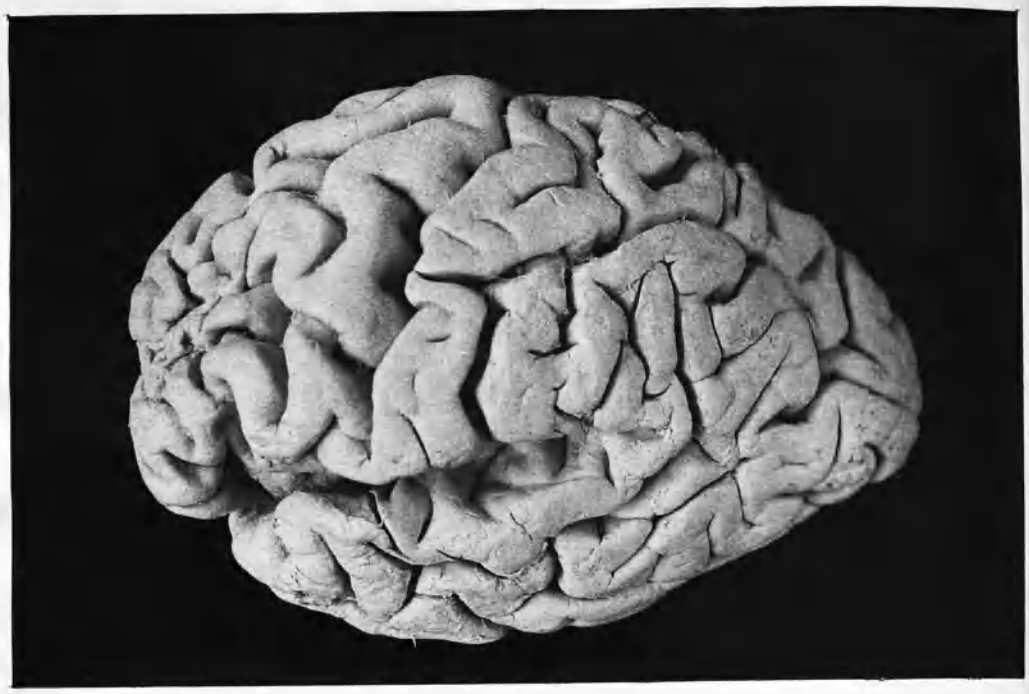
The Gyrus Fornicatus, as far back as the anterior end of the Precuneus is reduced in size.

Left Hemisphere:-

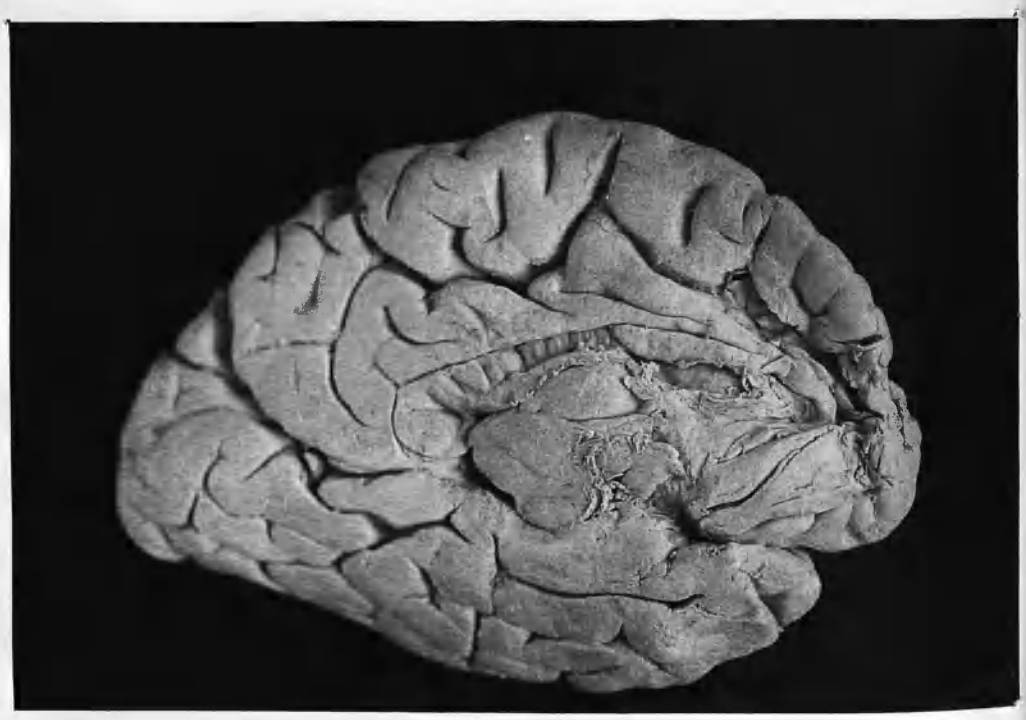
The convolutions of the entire Frontal seg-

ment- excepting the Ascending Frontal are small; while on the exterior aspect the anterior tip of the First Frontal for one inch and the same part of the Second Frontal for two

Case No. 461.



Left Hemisphere. Outer Surface.
The Prefrontal region is shrunken and the convolutions composing it are greatly attenuated.



Left Hemisphere. Inner Surface.
Note the appearances of the Prefrontal convolutions, the Gyrus fornicatus & the Corpus Cal-
um

for two

inches are extremely small and represented by thin worm-like convolutions. In the corresponding parts of the First Frontal, on the inner and lower aspect, a like condition is present.

The Corpus Callosum is thinned at its anterior end and the central portion of this part is semi-transparent and gelatinoid.

The Lateral Ventricle does not show the same dilatation as that on the right side.

The rest of the hemisphere and particularly the motor area is well developed.

A section taken from the most gelatinoid part and stained to show nerve cells showed:- A distinct though thin first layer with homogenous or faintly granular matrix, supporting a few oval or rounded cells with large faintly stained nuclei and a thin circle of protoplasm. No distinct processes can be seen springing from these cells, but they were noticed to contain some bright refractive spots. Beneath this follows a deep stratum of closely packed oval or irregularly shaped cells, most of which possess a definite nucleus which however, does not stain well; all are destitute of processes, many are vacuolated or contain fat-like specks. The matrix

The matrix stains normally, but throughout it are scattered a large number of round, small, highly refractive bodies which in places arrange themselves in clumps. This is followed by a broad layer of more or less pure sclerosis, showing an almost homogenous whitish ground work with a few fatty oval or round ed nuclei and some colloid bodies. These colloid bodies appear again under this layer, and finally the white matter is reached, in which these bodies are again seen.

SUMMARY.

The points of interest in this case are practically the same as those of cases No. 1600. and 18, except in the fact that the process has been of greater severity and not to have extended over such a large area.

Otto's point that the lateral Ventricles may be dilated is again well demonstrated.

The Motor area is again untouched.

It is noticed that the Corpus Callosum is in part affected, a fact which, as far as I am aware, no observer has yet called attention.

MICROCEPHALIC IDIOCY.

By the term Microcephaly is understood, according to the definition of Ireland, any cranium which is less than seventeen inches in circumference; it must, however, be clearly understood that even with so small a skull as the above, it is quite possible that the subject may be of average intelligence.

We find that the size of the head which is too small to permit of the intellectual faculties being present in a normal degree, is from thirteen to fifteen inches in circumference and nine inches from the root of the nose to the posterior extremity of the Occipital bone; these figures being those of Felix Voisin.

Turning now to the brain in Microcephaly, it is stated by Thurnan that all under the weight of 1000 gms. in males and 920 gms. in females belong the Microcephalic order, but if such is the case we have here several brains under these weights which by no stretch of imagination could be termed Microcephalic, thus showing that these figures require some considerable modification.

Taking now into account the pathology and causation of this condition, we find the opinions and theories of the various observers very divergent; the following may be quoted

be quoted

as being those most prominent.

The condition has been put forward as one of Atavism, and the observer who is the most emphatic on this point is Charles Vogt; on the other hand, this opinion is actively combated by Gratiolet who bases his argument on the phylogenetic development of different parts of the brains of man and the higher apes. He states "The temporo-sphenoidal convolutions appear first, and the frontal last in the brain of the ape, while in man exactly the opposite takes place. From this fact, which was rigorously verified, there flows a necessary inference; no arrest of development can make the human brain more nearly resembling that of the ape's than it is in the adult; far from that, it will differ so much the more, the less developed it is." He then goes on to take up the details of the difference between the brain in Microcephaly and that of the higher apes, and points out the impossibility of the former being a form of the latter.

Another and probably the correct view is that the Brain of the Microcephalic Idiot is one which is too small to allow of any degree of intelligence, and one which has only reached a certain degree of development, inside a skull also too small; in plain words, the condition is due to some form of developmental arrest, this according to some observers

some observers

is stated to take place between the third and fifth month of intra uterine life.

Another view which at one time attracted a considerable amount of attention, but one which at the present day might almost be called obsolete, is that this condition is due to premature synostosis of the cranial bones; this statement, however, has been proved over and over again, notably by Bournville who quotes over 300 cases, to be false; another point towards refuting this theory is that enlarging the skull by operative interference has been performed many times by different observers, with never, as far as I am aware, any improvement in the patient's mental condition. (Bournville quotes 80 cases of the same.).

As regards the various parts of the brain which are affected in Microcephaly; we find that almost any part of the Encephalon may be abnormally small or even wanting; in general the deficiency consists of a smallness of the hemispheres in every part. If any portion of the hemisphere is more prone than others to be affected in this condition it is the Frontal lobe; some observers notably Bischaff and Hubner state that in Microcephaly the Third Frontal convolution is always rudiamentary.

We find that the brains coming under this heading

this heading

can be divided into two classes, namely True and False Microcephaly. In the former we find no definite localised pathological lesion, simply a general smallness of the Encephalon, the gyri are comparatively large, and the sulci few and extremely simple; in fact cases have been described in which the sulci numbered only about five in number. In the latter, there is some definite lesion whic, at least, helps to bring about the diminution in size.

Case No. 228.

225

Calvarium seen from above.

CASE. No.228.

M.C.(female), aet.12. adm.17th July.1890.

Died:- 5th June.1892.

Cause of Death:- Phthisis pulmonalis.

Idiocy.

On admission:-

She had no intelligence and she could not understand anything that was said to her. She could not speak and only made inarticulate sounds. She was not able to perform the simplest acts for herself and was wet and dirty in her habits.

Previous to death:-

Her mental condition showed no improvement and she became if anything more degraded in her habits.

Post Mortem Examination.

Skull:-

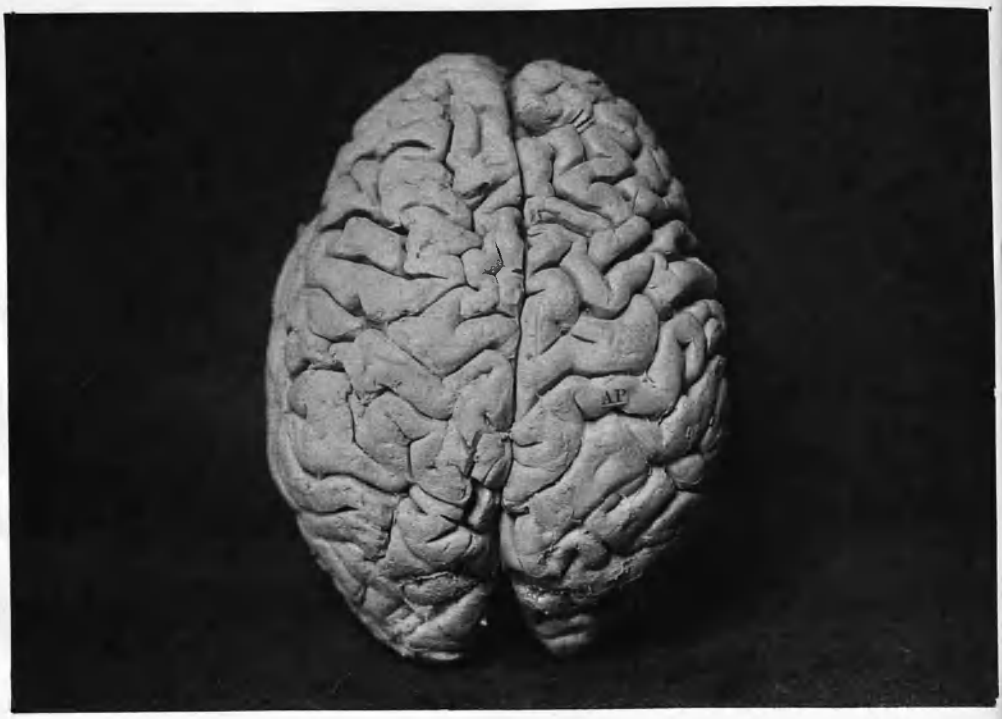
Horizontal Circumference 16 1/2 ins.

Transverse diameter 4 1/2 ins.

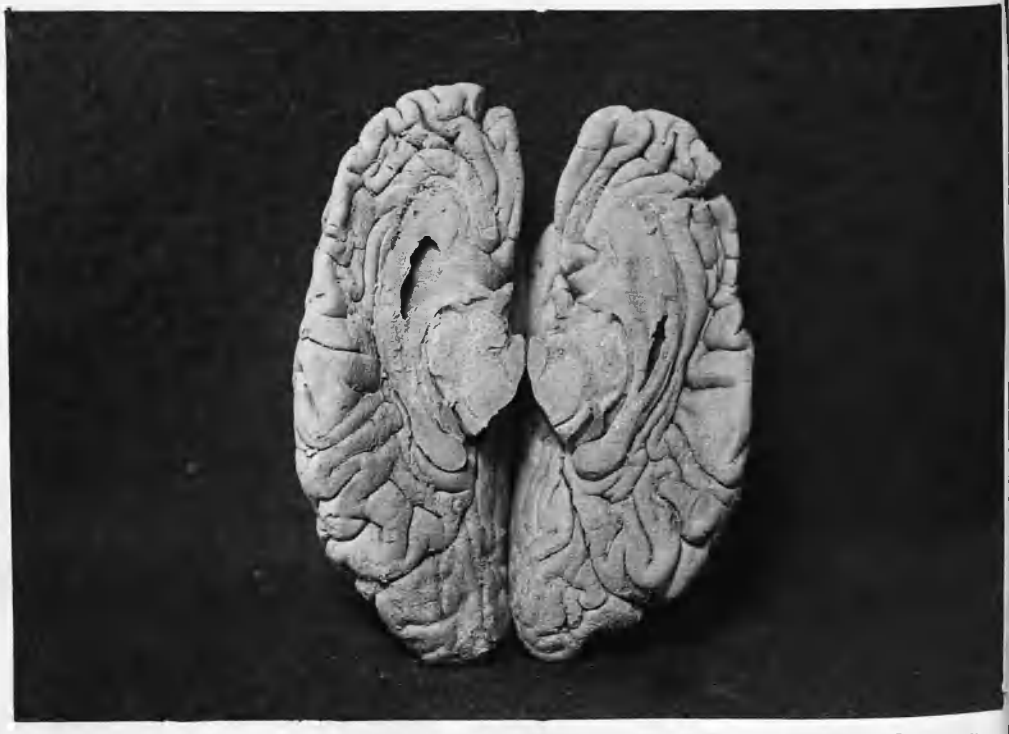
Antero-posterior diameter 6 ins.

This was dolio- and micro-cephalic in shape. Its symmetry was to all practical purposes perfect and its thickness and sutures normal.

Case No. 228.



Both Hemispheres viewed from above.
AP. represents the Ascending Parietal convolution of the right hemisphere.



View of the Inner Surfaces of Both Hemispheres.
Note:- The atrophied portion of the Gyrus Fornicatus and the interrupted Calloso-marginal fissure on the left side.

Encephalon:-

Weight 725 gms.

Cerebrum:-

Left Hemisphere. Outer Surface:-

The shape on the whole is practically normal. The convolutions are small and the arrangement of the same is rather complex.

It is to be noted that there is a small portion of the First Frontal convolution close to the inner margin of the hemisphere and one centimetre in front of the Precentral sulcus, which is distinctly atrophied to about half the size of the normal average size, otherwise the outer part of this convolution is normal. The rest of the Frontal lobe shows no distinct lesion.

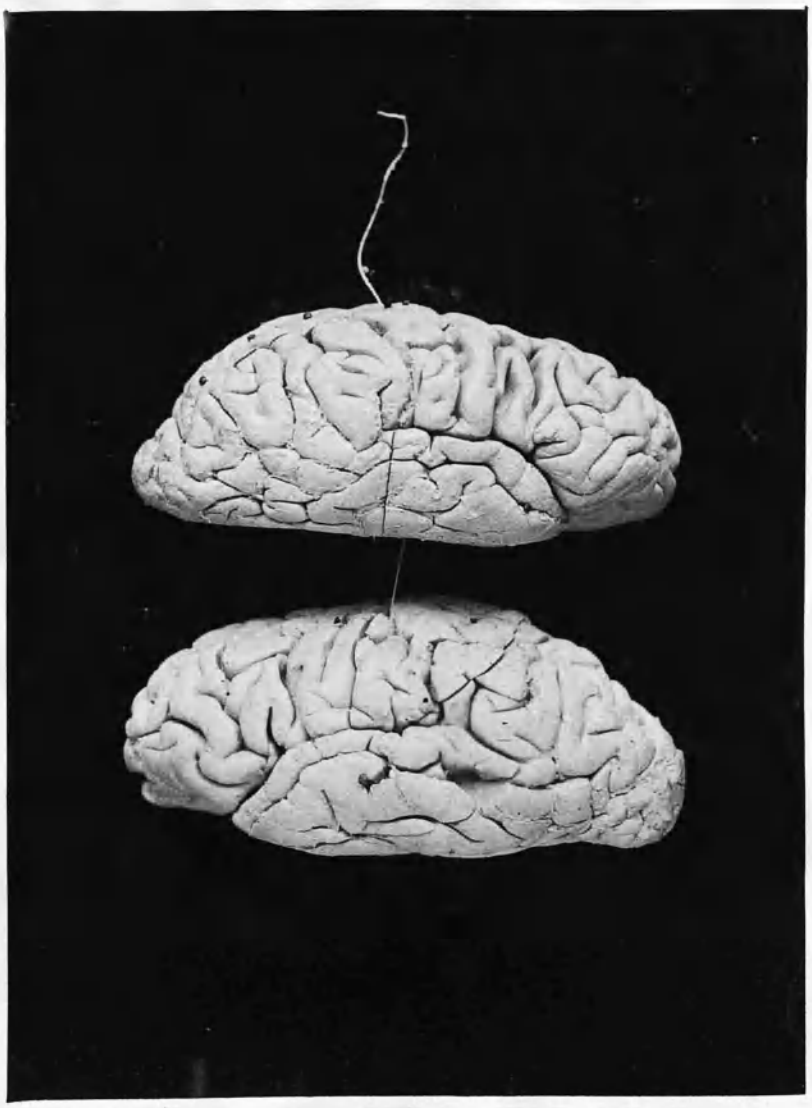
The Insula is not exposed and the remainder of this surface of the hemisphere is practically normal in convolutional arrangements.

As regards the sulci, these are normal in distribution, and it will be noticed that they tend to gape a little in the neighbourhood of the atrophied portion.

Inner Surface:-

It will be noticed that the anterior half of the Gyrus Fornicatus, excepting that part bounding the anterior end of the Corpus Callosum, is atrophied to about half the

Case No. 228.



226

View of Outer Surfaces of Both Hemispheres.

Note:- The smallness and the general regularity of the convolutions.

half the

size one would expect in a brain of this weight. The same may be said concerning that part of the First Frontal convolution and the Paracentral lobule, seen on the inner surface, which adjoin the Celloso-marginal sulcus.

The Cuneus etc., are practically normal.

The Calloso-marginal sulcus is interrupted at about the middle of its course, and the Parieto-occipital fissure runs rather far out on the external surface. The other great fissures follow a normal distribution.

The Orbital surface is normal.

Right Hemisphere:-

This differs very little from that of the other side; the atrophied Frontal portion is again well seen.

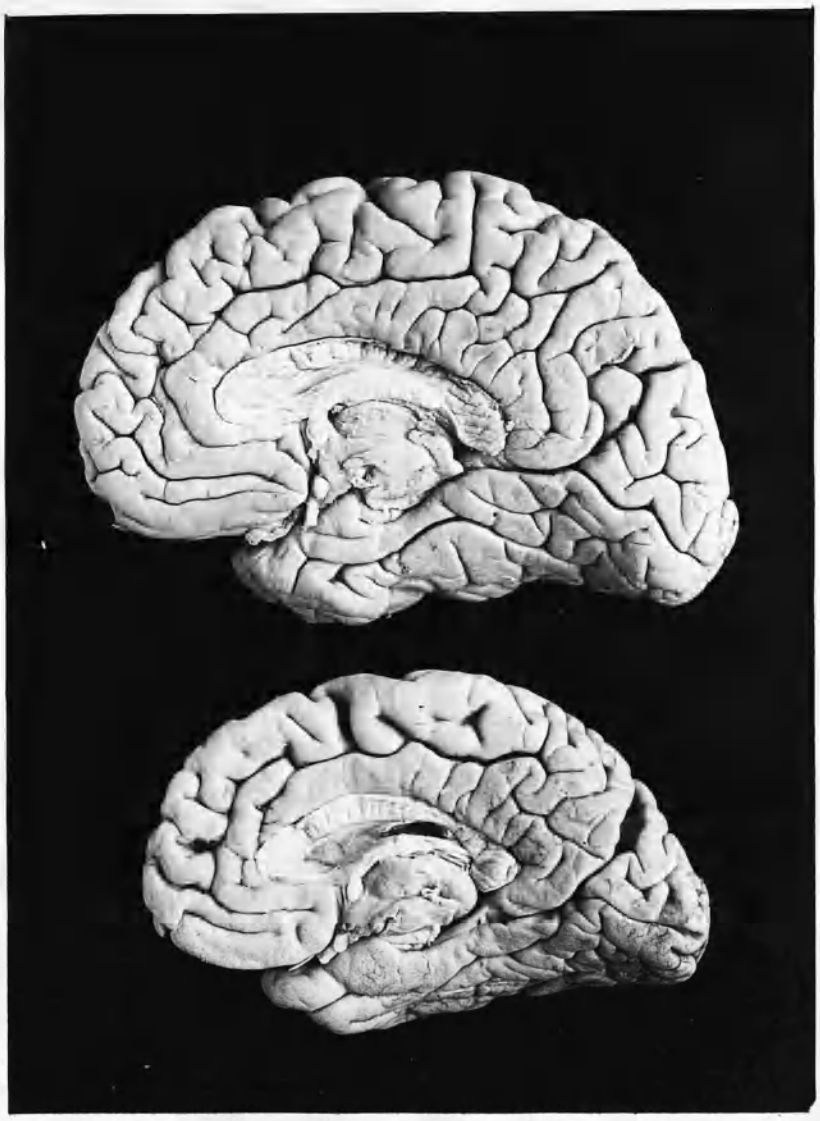
On the inner surface, however the Calloso-marginal fissure is not interrupted.

The Corpus Callosum etc. are normal in shape and appearance.

SUMMARY:-

That this case is one of Microcephaly, according to the definition of Ireland, there is no doubt, and it is of interest in representing that form which is known as False Microcephaly, as there is an atrophic lesion combined with the diminution in size of the hemispheres. This lesion was without doubt due to some occlusion of the Anterior Cerebral artery.

Case No. 1972.



Right Hemisphere. Inner Surface (above). Compared with the same of a normal brain (below).

MACRO.- or MEGALO-CEPHALY.

The following case illustrates extremely well this rare condition:-

CASE.No.1972.

E.M.(male) aet.15. adm.Feb.12th.1889.

Died:- May 30th.1903.

Cause of Death:- Gangrene of Lungs.

On admission:-

He was idiotic in appearance and smiled fatuously when spoken to. He could give no information at all about himself. He spent his time wandering aimlessly about the ward. He scratched and bit at all within reach. He was wet and dirty in his habits.

Previous to death:-

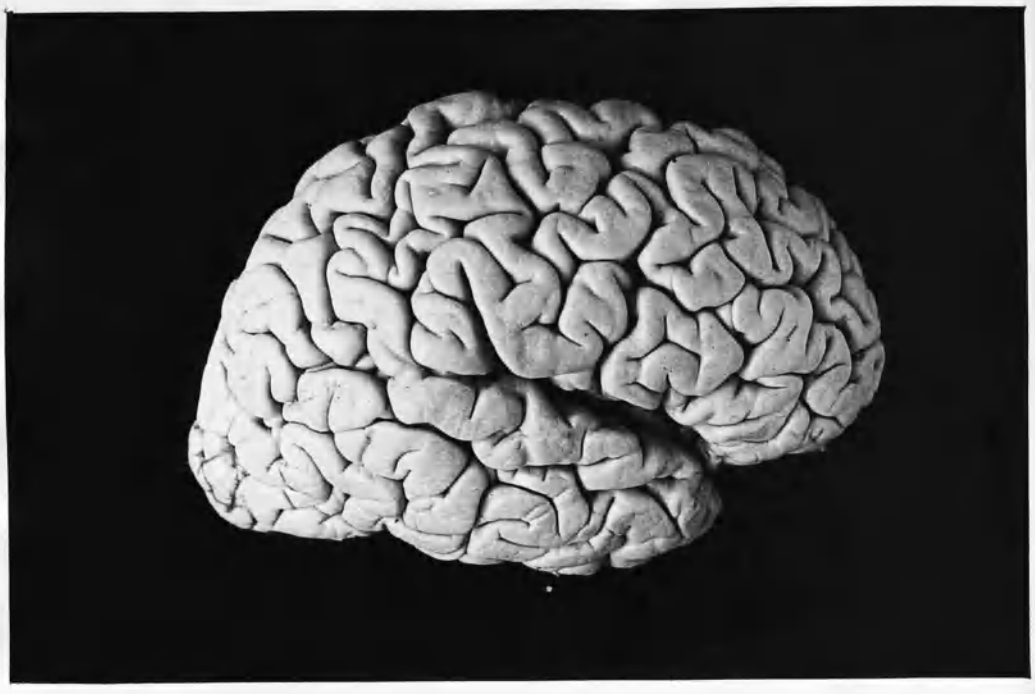
He never showed any signs of improvement mentally and became if anything more degraded in his habits.

Post Mortem Examination:-

Skull:-

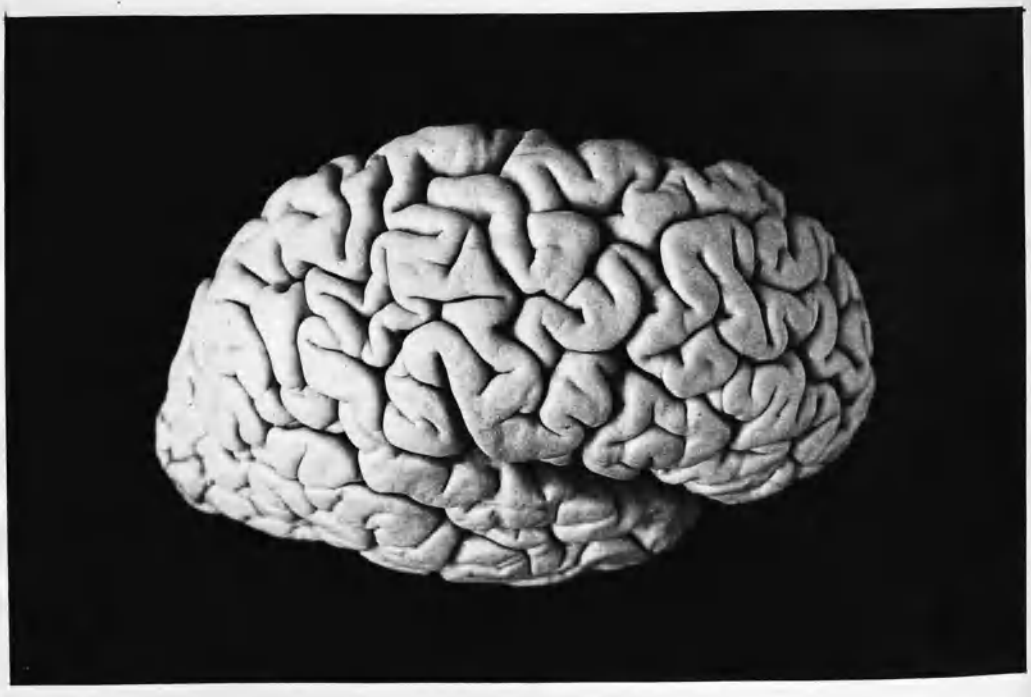
This was large and rounded in shape; the vault high; the frontal protuberances were not prominent and the sutures were close.

Case No. 1972.



Right Hemisphere. Outer Surface.

Note:- The complexity of the gyri and sulci.



Right Hemisphere. Outer Surface.

Another View of same.

Encephalon:-

Weight 1775 gms.

Cerebrum:-

Both hemispheres of the above are to all practical purposes alike, although there is a slight disparity in the weights.

Weight of right hemisphere 816 gms. Stripped 775 gms.

" " " 775 " " 747 "

The hemispheres are well developed in all directions and are symmetrical in shape.

The convolutions are normal in appearance and position; their shape is normal, the edges being well-rounded; their complexity is an extremely interesting point, in fact, every annectant gyrus seems to come to the surface.

The sulci are intricate and of normal course; they are of average depth, and such as one would be likely to find in the brain of an intelligent man. In places it may be noted that their course appears to be broken, this is due to an annectant gyrus coming up to the surface, which in a normal brain would be depressed below it.

In consistence the cortex is firmer than normal and of a slightly darker colour.

The Corpus Callosum, Fornix, etc., are all of a correspondingly large size.

Microscopic Examination.

Pieces of the Ascending Frontal convolution were taken and stained for nerve cells by the method of Nissl:-

These showed that the cell-lamination was good and that the cells were present in practically normal numbers, and the interesting point is that they were normal in shape and contents (i.e.the processes were numerous; the nucleus central in position and the Nissl bodies well marked.).

Pieces of the same part stained to demonstrate nerve-fibres showed that these were present in normal numbers and size.

By the method of Golgi, the Neuroglia was found to be present in more than normal abundance.

SUMMARY.

This case becomes of interest by the fact that here we have a brain, the convolutions of which are normal and intricate, the sulci deep, yet the subject was an absolute Idiot.

As regards the aetiology of this condition, this is absolutely obscure.

The condition has been looked on by many observers as a Hypertrophic Sclerosis; in this case we certainly have an over-abundance of fibrous tissue, in the form of neuroglia but it is of interest that with this increase no change in

change in

the structure and wealth of the nerve cells and fibres.

This case throws no further light on this condition but simply adds to the few so far described.

Cerebral ~~CEREBRAL~~ ASYMMETRY or DISPARITY.

By the above heading is implied Disparity in weight and size of each cerebral hemisphere without difference in shape or outline; i.e.the convolutions and sulci being of the same outline or shape on each side.

The following case is one of extreme interest, as it is the only one of its kind which, so far as I am aware, has been described.

From a pathological point of view it may be described as a Sclerosis affecting the hemisphere in its entirety, no parts escaping and all being affected to the same degree.

The cause of the above state is extremely obscure, and I must confess that I can put forward no pathological process which would cause such a great and uniform change in the surface of the brain. Some observers may be prompted to voice the opinion, that it might be due to some arterial occlusion and might quote the fact that the Anterior Cerebral artery was found to be smaller in calibre on the affected side, as a basis for their view, but if such were to be taken as the cause, one would expect that those parts in the neighbourhood of the anastomotic branches from the other side would either escape, or at least be less affected than those parts lying at a distance from the same, such,

such,

however, we see, is not the case; this fact in my opinion is quite sufficient to put that view out of consideration.

Again it might be put forward that the state is due to the bony asymmetry of the skull, but if such was the position of affairs then one would expect some parts to be more affected than others and also some abnormality in shape on the affected side; on this question we have the opinion of Bournville, the greatest authority on the Pathology of Idiocy, that the growth and shape of the skull is directly proportional to the growth of the brain, also that too early synostosis etc,. is due to, and is never the cause of, some abnormality of the growing brain.

Case No.845.

A.W.(female), aet.15, adm.19th.Feb.1890.

Died:- 25th.May.1896.

Cause of Death:- Phthisis pulmonalis.

Idiocy with Epilepsy.

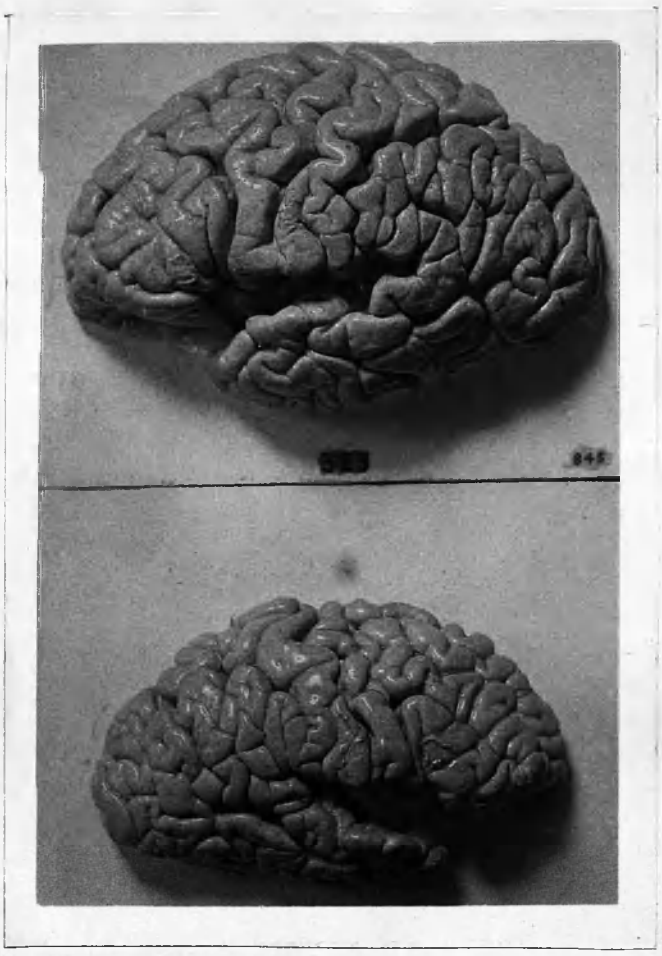
On admission:-

She could not speak and never attempted to move, but lay all day long curled up in a basket chair. She was wet and dirty in her habits.

Previous to death:-

She became more degraded in her habits, eating

Case No. 845.



Outer Surface of both Hemispheres.

Note:- The minature appearance of the Right Hemisphere compared with the Left.

eating,

if not watched, her own faeces. She became very destructive and required careful watching.

The fits averaged 29.3.per mensem.

Post mortem Examination:-

Skull:-

Shape:- narrow frontally.

Symmetry:- Right half considerably smaller than the left. (This applies to the bones of the vault as well as to those of the basal fossae.).

Encephalon:-

Weight 800 gms.

Cerebrum:-

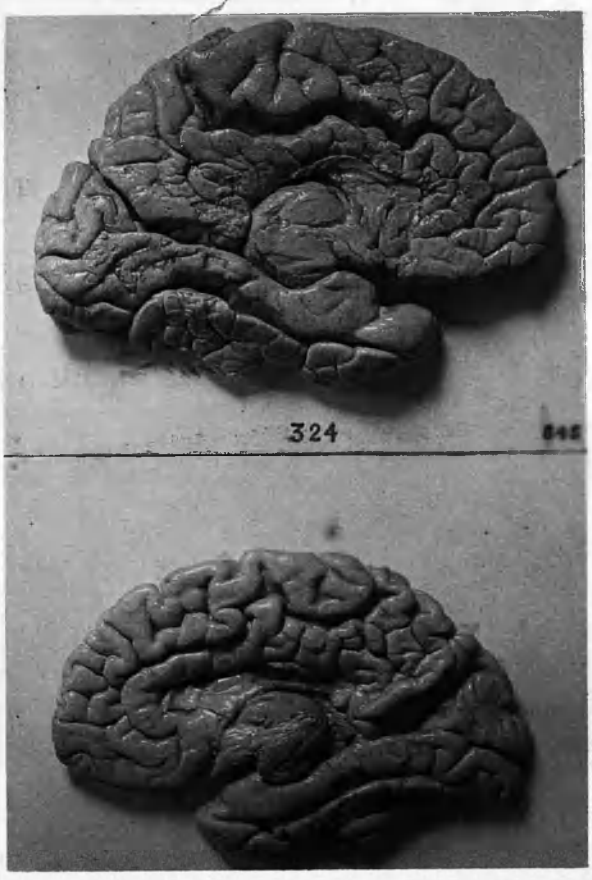
There is marked asymmetry, the right hemisphere being much smaller than the left. The conformation of both hemispheres being normal and to all practical purposes identical. (It was noted at the beginning of the Post-mortem examination that the Anterior Cerebral artery of the right side was of smaller calibre than the left. The remaining vessels seemed normal and symmetrically disposed.)

Right Hemisphere:-

Weight 218 gms. stripped 198 grms.

The consistence of this hemisphere is in every

Case No. 845.



Inner Surface of both Hemispheres.

Note:- The disparity in size.

in every

segment very firm and contrasted with the left hemisphere, which is apparently healthy, the difference is very striking.

This Sclerosis, if it may be called so, seems to pass right through the hemisphere and to affect all parts alike.

The surface is smooth. There are no gross lesions to be detected and the atrophy is apparently equal all over, no convolutions being found which are markedly smaller than others.

The cortex is thin and not well striated.

Left Hemisphere:-

Weight 417 gms. stripped 405 gms.

The size of this hemisphere is, on the whole, good. The convolutions are well developed and are arranged in a normal manner. The cortex is of normal depth and the sulci healthy, in fact no changes are apparent in it except, that it may be said to be on the small side for one of her age.

Cerebellum:-

The left lobe is somewhat smaller than the right.

Microscopic Examination:-

Portions of the cortex were taken from correspondin parts of the Ascending Parietal convolutions and stained by

stained by

the method of Nissl.

These showed striking differences, the main change appeared to be a diminution in the number and size of the nerve cells, a diminution without special implication of any particular layer or layers.

Comparing for instance the large pyramidal cells of the left side, they were numerous (normal) and morphil-ogically perfect, while on the right, although some were seen which appeared healthy, still the total number was great-ly below normal.

CONCLUSION.

-%-%-%-%-%-%-%-

In conclusion, I have to thank Dr. Wiglesworth, the Medical Superintendent of the County Asylum Rainhill, for permission to use the pathological material here described and also for access to the Medical Journals of the Institution.

I also acknowledge with thanks A.W.Campbell M.D., Pathologist to the above Institution, for many suggestions in treating the above subjects, also for placing his library at my disposal and for translations of extracts of some of the following references.

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