A CLINICAL STUDY

OF

3 Cases of Lipodystrophia Progressiva,

With a

Review of the Literature.

A THESIS,

Presented for the M.D. Degree,

BY

JAMES BAIRD.

SUNDERLAND :

PRINTED BY R. ATTEY & SONS, 17 NORFOLK STREET.

ProQuest LLC.
789 East Eisenhower Parkway
P.O. Box 1346
Ann Arbor, MI 48106 – 1346

A CLINICAL STUDY
OF
3 CASES OF LIPODYSTROPHIA PROGRESSIVA.

This condition being described as exceedingly rare, I may consider myself very fortunate in having had the opportunity of having under my own observation in my practice, the following typical cases :—

CASE I.—D.....B..... a female æt. 19 years (23-1-27.)
Married ; has one child, a healthy female, æt. 2 years.
Complaint.—"Thinness" of 3 years duration.

She states that she first noticed her cheeks becoming thin and that this was accompanied by an itching sensation inside the ears. She describes this sensation as proceeding downwards on both sides underneath the tongue and into the floor of the mouth between the teeth and the tongue.

This sensation she still has ; it comes on two or three times daily and lasts for a few minutes. It is present on first waking as a rule and she can abolish it by obliterating the external auditory meatuses and stroking the cheeks over the masseters

The emaciation proceeded progressively downwards over the neck and chest wall and then stopped short just above the navel.

Pari passu with this there was a wasting of the shoulders and arms.

This stage was reached in about a year and has remained stationary since.

She was scalded at æt. 9 months on the right side of the neck.

Had measles at æt. 7 years ; and "tonsillitis" at 14 years (uncomplicated.)

Menstruation began at the age of 13 for 3 months and then ceased for 6 months. She has menstruated regularly since with the exception of her pregnancy and she has corresponded faithfully to the 6/28 type ; without pain or other abnormality.

She has never had any employment other than household duties.

Father and Mother alive and well ; Father æt. 45 suffers from occasional attacks of gallstones—lost an eye due to an accident.

Mother æt. 43—obese—in good health and has never had any illnesses.

2 Brothers dead : æt. 21 killed and 1 month pneumonia.

Has 2 Sisters living : æt. 13 measles at æt. 6 and has had tonsillectomy done ; the other æt. 8 healthy.

Fig. 1
See index of illustrations. (p. 17)

Paternal Grandfather obiit æt. 76—Maternal Grandfather alive and well.

Paternal Grandmother no history. Maternal Grandmother obiit "old age."

No relative ever suffered from epilepsy, migraine, asthma, etc.

Patient has always been in comfortable working class surroundings; gets a sufficiency of good wholesome food; is well clothed and lives under modern improved sanitary conditions.

Her usual practice is to rise at about 5-30 a.m. she has a cup of tea and makes her husband's breakfast and occasionally goes back to bed for an hour; but always gets up at 7 a.m. to get the morning meal ready.

This meal consists invariably of bacon and eggs, tea, with white bread and butter.

She then does ordinary household duties until 12 noon when she has a dinner consisting of soups, beef, mutton, mince, potatoes, green vegetables, turnips, yorkshire puddings, boiled suet puddings, milk puddings, fresh fruit, prunes.

She takes very little exercise except in summer when she goes swimming once or twice a week.

Takes 3 or 4 cups of tea or coffee daily.

Total abstainer.

Retires between ten and eleven p.m.

States she never feels cold.

Since I have known her, viz.: 4 years, her average weight has been 8 stones 6½ lbs. Height 5ft 1½ins.

She is an intelligent, bright girl; carries herself well with shoulders square. There is no lordosis or scoliosis and she has no limp or other peculiarity of gait.

Her facial appearance is very striking; she has the appearance of a woman at least 15 years older who is recovering from a long wasting illness; the infra-malar hollows and the drawn appearance of the face generally are well brought out in the accompanying photographs.

The eyes are bright; there is no strabismus, nor inequality of the pupils.

The complexion is pale except for some local cyanosis of the hands; she has a few spots of acne vulgaris, and the sebaceous secretion in the usual situations is rather excessive.

The hair is dry but of normal female distribution and there is no excessive growth of hair. The nails are normal.

Perspiration is present but not excessive.

The skin is elastic and appears normal; the musculature is well developed and rather above the average.

Fig. 2.

The breasts are well formed and there is no wasting of the subcutaneous fat in this situation.

The striking features of her appearance consist in the absolute loss of the subcutaneous fat from the tissues of the face, the neck, the arms, the thorax and the abdomen as far as the umbilicus. Below this level the appearance of the patient differs in no respect from any normal individual except that the loss of the fat above gives the relative appearance of increase of the subcutaneous fat below.

The loss of subcutaneous fat is not absolute on the posterior aspects of the arms and the chest wall; but though not absolute it is well marked and moreover the loss extends to a lower level than in front: namely as far as the iliac crests.

There are no glandular enlargements; the thyroid is not palpable.

Pulse averages 84 beats per minute. Respirations 15 per minute.

ALIMENTARY SYSTEM.

Good appetite—no excessive thirst.

Does not eat between meals.

Bowels move once daily.

Teeth are bad—the molar teeth are all carious and the gums are spongy and bleed easily—the tonsils are enlarged and the tonsillar tissue fibrotic—she has a high narrow arched palate.

The lips are red—the tongue moist and red—there is nothing abnormal to note with regard to the odour of the breath.

Palpation of the abdomen reveals nothing abnormal—the spleen is not enlarged.

The total acidity of the gastric contents was 48. Free HCl was present altering the colour of Congo red to light blue.

The fæces appeared normal and there was no "neutral fat" detected microscopically on a mixed diet.

CIRCULATORY SYSTEM.

She has no objective symptoms and does not complain of præcordial pain or breathlessness. She sleeps well and has neither palpitation nor giddiness.

The pulse is 84 per minute—regular—beats similar—vessel wall soft—pulse pressure good and there are no secondary waves.

The average blood pressure is 130mm Hg Systolic and 90mm Hg Diast.

The apex beat is in the 5th interspace ¾in. internal to the nipple line; upper border lower edge of 4th rib; right border left edge of sternum (deep cardiac dulness).

Fig. 3.

There is visible pulsation in the epigastrium but not at the root of the neck. She has a soft v.s. mitral murmur not propagated and heard best at the apex.

Hæmoglobin 75% Red cells 4750000. White cells 5800. The red cells appeared normal and a differential white cell count revealed no departure from the usual proportions.

RESPIRATORY SYSTEM.

Expansion of the chest wall is regular and symmetrical. There is neither increase nor diminution of vocal fremitus or resonance. The breath sounds are vesicular throughout and there are no adventitious sounds. No sputum.

GENITO-URINARY SYSTEM.

Passes on the average 48 oz. per 24 hrs. 32 oz. by day, 16 oz. night urine.

Spec. Gravity 1012 slightly acid.
No albumen present.
,, sugar ,,
,, blood ,,
,, bile ,,
,, acetone ,,

Urea concentration 1·80%. 3 hours after 15 gms. urea by mouth 1·6%.

Menstruation regular of the 6/28 day type; no dysmenorrhæa.

She has one child born normally; 1st stage 10 hrs.; 2nd 2¼ hrs.; 3rd 20 mins. The secretion of milk ceased after 12 days absolutely; the puerperium was otherwise uneventful.

NERVOUS SYSTEM.

Right handed.
Memory—for recent events good—past events fair.
Sleeps well—does not dream—not unduly emotional.
There are no motor speech defects and there is no aphasia.
The cranial nerve functions are unimpaired.
There is no hippus and the cilio-spinal reflex is intact.
Motor functions are normal and co-ordination is perfect.

Epicritic and protopathic sensation and the muscle-joint sense are present and unimpaired all over the body: and thermal and tactile sensations are normal.

There is in no situation any hyperidrosis or other vasomotor disturbance: and there are no trophic lesions.

Apart from the excessive sebaceous secretion mentioned above and the curious form of paræsthesia already noted in the distribution of the 3rd division of the 5th nerve the nervous system is otherwise normal.

Fig. 4.

CASE II.—A..... S..... female, æt. 34 (21-1-27), married 11 years. No children, never any miscarriage.

Complaint – States that she continually feels sick—feels the cold badly and though she has noticed her thinness would not have complained of it.

She was 16 years of age when she began to menstruate, but a few months later she had amenorrhœa for a period of 5 months; about this time she had "bilious attacks which lasted for 24 hours and recurred at intervals of 2 to 3 months; with these attacks she had pain in the pit of the stomach."

About the age of 18 years she noticed that she was getting thin at the temples; then her cheeks and neck became thin and the emaciation proceeded progressively downwards affecting the arms at the same time, and stopped short at the navel. The whole process was complete so far as she can recall in about 2 years.

At the age of 21 and again at the age of 26 she had an attack of "bleeding from the stomach" associated with acute pain. On each occasion she was laid up in bed for 5 weeks. She still has attacks of pain in the "stomach" mostly in the morning.

She had measles at the age of 7 followed by scarlet fever but both diseases were uncomplicated.

She complains of several times recently having noted a sensation of numbness in the tongue and cheeks. She feels as if her tongue were swollen and she can abolish the sensation by rubbing the cheeks.

The only other illnesses she has had were "enlarged glands" in the neck at 14 and an attack of influenza at 25.

Her father died at the age of 66 from acute lobar pneumonia.

Her mother is alive at the age of 65; she suffers from valvular disease of the heart.

She has 5 sisters; all are living:—

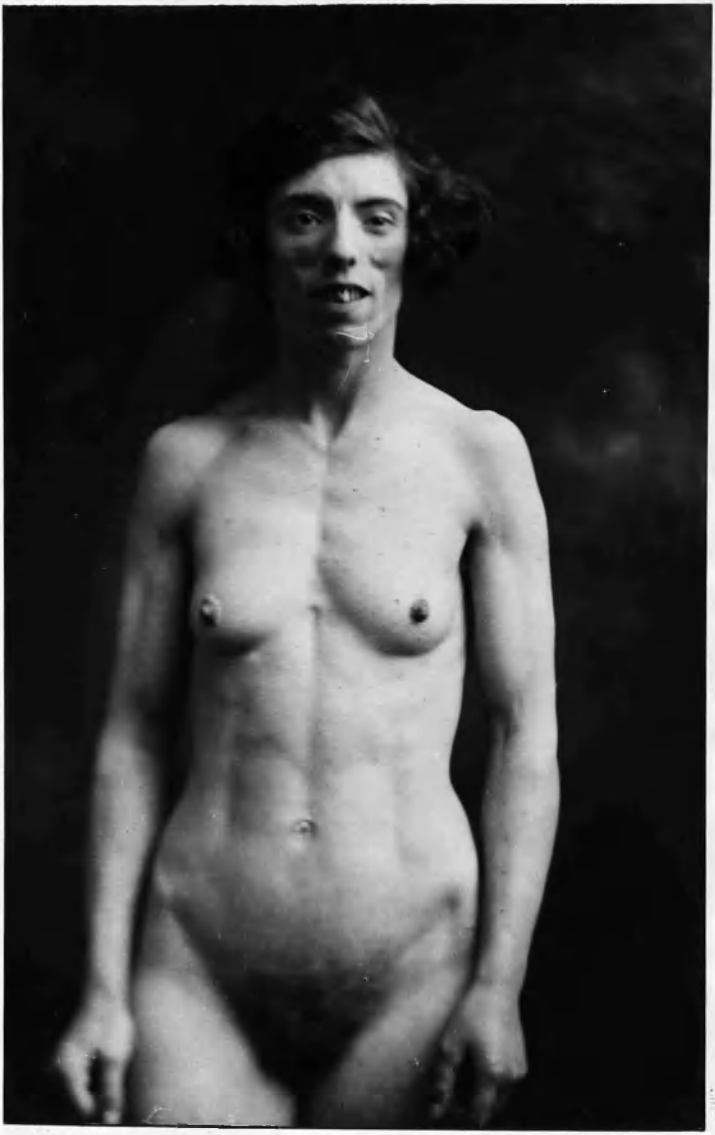
1. Aet 44, married, healthy—has 1 child, female, ? tubercular.
2. Aet 32, married, healthy—has 4 children alive and healthy and 1 dead from ? convulsions æt 1 year.
3. Aet 28, married, healthy—has 2 children healthy and 1 dead from tubercular meningitis.
4. Aet 24, single—Chronic otorrhœa.
5. Aet 20, single—Pulmonary Phthisis—Alive.

She has 3 brothers alive and healthy:—

1. Aet 40, married, 1 child—healthy.
2. Aet 35 ,, ,,
3. Aet 26, ,, ,,

One brother died from Dementia paralytica æt. 37.

Fig. 5.



The only other family history obtainable is as follows :—

A maternal aunt died from puerperal fever at æt. 28 and another at æt. 44 from epilepsy. Two maternal uncles died from "heart disease" at æt. 44 and 28 respectively.

She worked as a confectioner for a period of 6 months and at dressmaking for another 6 months; apart from this she has always been employed at domestic duties in her own house.

Before her marriage she was always very comfortable and was regular in her habits; since then her habits of life have been very much the opposite. She rises between the hours of 10 and 12 noon, never has any breakfast; at about 12 noon she has a meal consisting of bacon with brown bread and butter and tea. At 6 p.m. she has tea and white bread and butter; then at varying periods between 10 p.m. and 2 a.m. she has what she calls dinner, and this meal consists of cocoa, fried fish, milk pudding, fruit and vegetable.

These conditions are due to the "idiosyncracies" of her husband; an individual who lives upon an allowance from his mother and who has never worked in his life!

She lives under modern sanitary conditions; walks 3 or 4 miles daily, occasionally goes to "pictures" and does needlework.

Her average weight is 6 st. 11 lbs. and her height 5 ft. 2in.

She is of normal intelligence for the education she has had.

She has a good carriage with square shoulders and no lordosis, scoliosis or kyphosis.

She has the characteristic facial appearance; drawn haggard facies with temporal and infra-malar hollows. The skin is well nourished but rather oily and the hair is greasy. The eyes are bright and there is neither strabismus nor inequality of the pupils.

The distribution of the subcutaneous fat dystrophy is similar to case 1, but the loss is more evenly distributed and is as marked over the back of the chest wall as it is in front.

She is pale but there is no apparent anaemia. The thyroid gland is impalpable; the hands are cyanosed; there is no excessive perspiration. The nails are normal; hair is present in the usual situations and is of normal female type of distribution; though there is rather an extensive growth of hair on the extensor aspects of the forearms.

The breasts are well formed and the muscular development is good though not excessive as in the previous case. There are no glandular enlargements. Respiration is of the thoracico-abdominal type about 15 respirations per minute.

The lower limbs are normal except for the presence of slight varicose veins.

Fig. 6.

The teeth are bad—molars carious and septic gums slightly retracted. The lips are red, the tongue is cyanosed; the side papillae are enlarged and inflamed; the tonsils are small and buried ; the palate is normal.

ALIMENTARY SYSTEM.

There is no pecularity of the odour of the breath.

The appetite is capricious and poor; there is no excessive thirst.

She does not eat between meals; there is no pain but occasionally she has a sense of fullness in the stomach after meals, coming on in 5 or 10 minutes after a meal and passing off in a few minutes. This is not influenced by any particular kind of food and there is never any vomiting, flatulence, or eructations.

The bowels are moved only every other day and then with aperients.

There are no haemorrhoids ; there is some deep tenderness in the right iliac fossa and I think I can feel a thickened appendix. There is nothing else abnormal to note.

The total acidity of the gastric contents after a test meal was 50 : free HCl was present. The faeces appeared normal and no neutral fat was seen microscopically.

CIRCULATORY SYSTEM.

She has occasional attacks of dyspnoea mostly when sitting talking; there is no præcordial pain; no palpitation and she sleeps well.

She sometimes gets attacks of vertigo but only when her "stomach is out of order."

There is no oedema of the feet.

Pulse rate 80 per minute regular; good volume and tension.

The apex beat is in the 5th interspace ½in. internal to the nipple line.

There is no epigastic nor episternal pulsation. The upper border of the deep cardiac dulness is at the lower border of the 4th left rib and the right border at the left edge of the sternum. There no murmurs or other abnormalities on auscultation.

The red cells numbered 5200000 and the white cells 6200 per cmm.

There were no abnormal forms of red cells noted and the differential white cell count was within normal limits.

Fig. 7.

RESPIRATORY SYSTEM.

There is nothing abnormal to note here; the chest wall moves equally all over and expands normally. The percussion note is resonant throughout and there are no adventitious sounds, the respiratory murmur being vesicular: and there is neither diminution nor increase of the vocal fremitus or vocal resonance.

GENITO-URINARY SYSTEM.

Menstrual periods began at the age of 14 and are of the 1/28 day type. She had premenstrual pain until æt 21 but none since.

Her marriage has never been consummated and she is virgo intacta.

She passes only 24 oz. of urine in the 24 hours and the day and night quantities are equal; there is no strangury.

The urine is of a pale straw colour slightly acid in reaction and the specific gravity is 1012.

No albumen present.
,, sugar ,,
,, blood ,,
,, bile ,,
,, acetone ,,

Urea concentration 1·1%. 3 hours after 15 gms. urea by mouth 1·5%.

NERVOUS SYSTEM.

She is right-handed; the intellectual functions might be described as below the average but there is no actual mental deficiency; her memory for distant and recent events is good; she is rather of the "colourless" unemotional type.

The cranial nerve functions are unimpaired and not perverted: with the following exceptions.

The peculiar form of paræsthesia noted above in the distribution of the 3rd division of the 5th nerve.

Some slight nerve deafness on the right side.

The functions of the cervical sympathetic are normal and the cilio-spinal reflex is intact. There is no hippus.

The peripheral nervous system is normal; epicritic and protopathic sensation are present and normal all over the body; the kinæsthetic and stereognostic senses are unimpaired; there are no hyperæsthetic areas.

There does not appear to be any vasomotor disturbance, with the exception of the cyanosis of the tongue and hands.

The superficial and deep reflexes are present and not exaggerated; the plantar response is flexor.

Fig. 8.

Muscular function is unimpared and co-ordination is perfect.

She complains of an itching sensation in the pit of the stomach; but her description was vague and I could not detect any area of hyperæsthesia or analgesia.

Thermal and tactile sensation are normal and there are no trophic lesions.

CASE III.—M..... P..... female æt 11. (? Feby., 1923).

Unfortunately I did not know the condition for what it really was when I had her under observation and this case has not been fully investigated. The photographs were simply taken as a matter of interest.

The emaciation began at the age of 6 years after an attack of scarlet fever and the distribution is similar to the previous case.

I have lost touch with the case and cannot trace the patient.

REVIEW OF THE LITERATURE OF THE DISEASE.

Index of Bibliography on pp. 16 & 17.

The disease known as Lipodystrophia progressiva was noted by Osler (1) in 1895 but was not named by him at that time; he noted that the disease was confined almost entirely to females and that the breasts were spared.

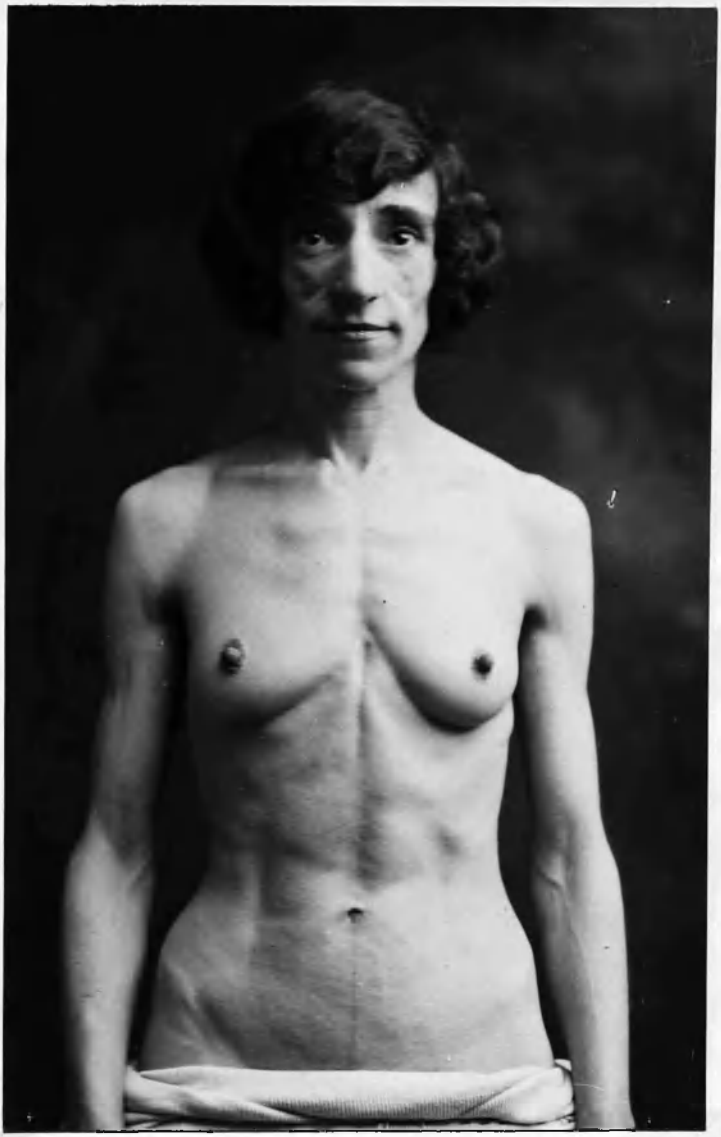
The earliest published case appeared in 1906 and was by Barraquer (2) of a woman æt 25 in whom the disease commenced at the age of 18. In 1911 Simons (3) recorded a case in a woman æt 21 in whom the disease began at the age of 11; and he gave the name of Lipodystrophia progressiva to it.

Since that date some 50 odd cases have been described; by far the greater number being females; the best descriptions are probably those of Lee Smith (4) and Parkes Weber (5).

Lee Smith describes the onset as insidious and as occasionally coming on in early life with vague aches and malaise but nothing of a definite character. The characteristic feature in all the cases is a slowly progressive and almost complete disappearance of the subcutaneous fat from the face and neck, the upper extremities and from the trunk as far as the iliac crests and folds of the groin. Here the fat absorption ends abruptly

In a few cases, nearly always a male subject, the emaciation stops short at the clavicular region or at the costal margin.

Fig. 9.



He states that there is an excessive accumulation of fat in the buttocks which makes the clinical picture a very striking one; while Osler was of the opinion that the appearance of excessive fat deposition in the buttocks and thighs was merely a relative one.

Lee Smith says the affection is not hereditary and is consistent with good health and well being.

Prior to 1914 the disease was thought to be peculiar to females but in that year Husler (6) described 2 cases in boys of 10 and 9 respectively. Male cases had been noted before this however; Batty Shaw (7) had in 1905 described a case of "bilateral atrophy of the face" in a boy of 10 and Bury (8) a similar case in 1912 in a boy. Hertz and Johnson (9) had a case in a man of 38 in 1913 but this was attributed to chronic plumbism. Since 1914 seventeen further male cases have been described; but this still leaves the incidence amongst females much greater.

Watson and Ritchie (10) state that the affection is neither racial nor familiar or hereditary. They believe that it is more common than is supposed and instance the case of a woman who came undert heir observation: who had been treated for ulcer of the stomach, appendicitis, peritoneal adhesions, uterine fibroids, pulmonary tuberculosis: had been subjected to special dietetic treatment, and finally treated for neuro-circulatory asthenia. She presented the typical appearance of the disorder.

The first signs of the disease are as a rule noted between the ages of 5 and 8 years (64%). The earliest recorded age of onset is 2 years Trömner (11) and the latest Hertz and Johnson 37 years. The active stage lasts as a rule from 2 to 3 years but it may last only a few months or be prolonged for a period of 4 years, (Pic and Gardère) (12.)

Recrudescence is rare; in one of Christiansen's cases activity was renewed after erysipelas (13) and in one of Watson and Ritchie's it was renewed after pregnancy and again 14 years later without obvious cause.

Recovery is unknown.

Adiposity of the lower limbs is symmetrical except in one recorded case by Laignel-Lavastine and Viard in which the labium minus and lower limb were larger on the left than on the right side (14.)

Parkes Weber's description of a typical case is worth quoting:—"One need only imagine a grotesque figure, the lower part of which seems to be modelled after an extraordinarily florid Venus of the "ultra-Rubens" style, whilst the face and upper part of the trunk might—bring to mind the popular idea of one of the witches in Macbeth."

Fig. 10.

The loss of subcutaneous fat is in the vast majority of the cases noted to have extended as far as the iliac crests; though it has been described as having been confined to the face by Batty Shaw, Hertz and Johnson, and Husler. In other cases again the lipodystrophy has been noted to extend beyond the usual limits; and in each case the patients have been males; Gerhartz's case of a man æt. 29 (15) showed wasting of the subcutaneous fat of the front of the thighs; both lower limbs were affected in Christiansen's case; and in Feer's case (16) the distribution was complete from face to feet.

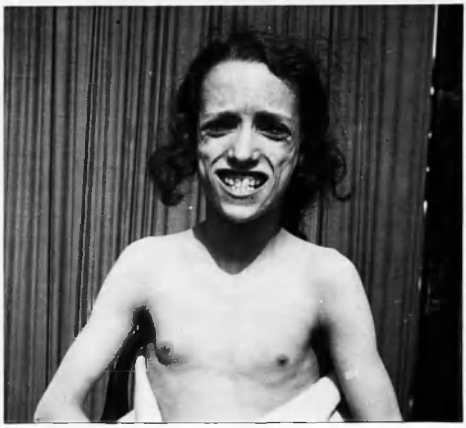
It is stated that the adiposity below the level of the lesion is increased in typical cases; but this seems to be a moot point.

The skin is normal in texture; Lee Smith refutes the suggestion that "elastic skin" is the same disease by showing that in that disease there is an alteration in the elastic fibres of the skin, while the other tissues remain normal; four cases have had portions of the skin and subcutaneous tissue from dystrophic areas excised and in each case the appearances have been similar, namely, the skin was normal and there was complete disappearance of the fat from the subcutaneous tissue though fat globules persisted in the sebaceous glands; References: Simons, Christiansen, Feer and Lee Smith.

The bones are unaffected and clinical evidence and occasionally radiographic evidence have distinguished the disease from bilateral facial atrophy; Frank (17) had a case in a girl æt. 11, however, who was the subject of fragilitas ossium and who possessed in addition blue sclerotics. Several members of her family were the subject of blue sclerotics and fragilitas ossium but not of lipodystrophia.

It will thus be seen that the cases I have reported are typical; the female incidence; the otherwise good health; the onset at an early age especially in case three; and the distribution are perfectly typical. Other features corresponding to the usual description are the sparing of the breasts in the first two cases; the fact that the maximum intensity was reached in 1-2 years: and that the wasting was progressive and that there have been neither remissions nor exacerbations of the disease.

Fig. 11.



ÆTIOLOGY OF THE DISEASE WITH SPECIAL REFERENCE TO THE CASES DESCRIBED.

Campbell (18) put forward the suggestion that excessive activity of the sebaceous glands might be responsible for the draining away of the subcutaneous fat; excessive sebaceous secretion has not been recorded in any subsequent case and though it was rather excessive in my first case, I am not prepared to lay any stress on this point, much less to agree that it is a causal factor.

Laignel-Lavastine and Viard suspected tuberculosis as a cause—there certainly is a fairly strong strain present in my second case, but the patient herself presents no clinical evidence of the disease; and in both the first and the second cases the Von Pirquet cutaneous test was negative absolutely.

Syphilis as a causal agent is unlikely in my cases; in the first two the Wassermann reaction in the blood serum was negative and the third case showed no stigmata of congenital infection.

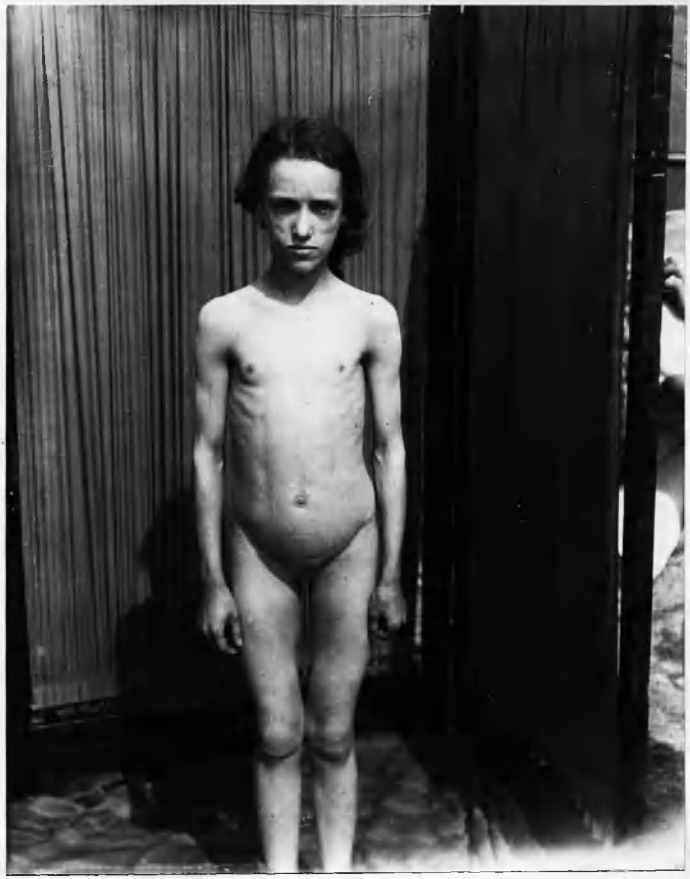
The condition is certainly not due to malnutrition in my cases; and this is supported by the evidence obtained by over-feeding experiments in the cases of Mirallié and Fortineau (19) and Hartenberg (20). These patients developed great increase in the adipose tissue of the unaffected parts while the affected parts remained unaltered.

Faulty or deficient fat assimilation has little evidence in the literature to support it as a possible cause and this is borne out by the absence in my first two cases of free fat in the stools. Watson and Ritchie note the presence of an abnormally low content of split fat in the fæces of one of their patients; but in no case published has there been any steatorrhœa noticed and Simons found that there was no increase of the blood lipase.

Carbohydrate metabolism has been attacked and the results seem to have acquitted it as being at fault; in my first case the blood sugar level, as estimated by McLean's method was 0·09% and 2½ hours after oral ingestion of 50gm. glucose it was 0·106%. The figures in my second were respectively 0·100 and 0·093%. References in the literature to glucose tolerance and glycosuria are to be found in the work of Spear (21), Klien (22), Feer and Lee Smith.

Disturbance of the endocrine balance has been put forward as a cause but it is very difficult to see how the absence, excess, or perversion of a secretion which is circulating throughout the body can give rise to the typical case. No doubt in a case such

Fig. 12.



as that of Feer, where the lipodystrophy was complete the theory is attractive, but it does not explain the majority of these cases. It has struck me that an ardent supporter of the endocrine theory might still uphold his theory on the grounds that while the fat dystrophy was not complete, that which remained being fluid at the body temperature, would naturally accumulate in the dependent parts. This would account for the excessive adiposity described in the thighs and buttocks and also for the sparing of the hands in some cases This attractive theory is easily refuted when it is remembered that fat is an inta-cellular content and therefore not subject to the laws of gravity taking the body as a whole,

Myxœdema was noted as a concomitant by Christiansen; exophthalmic goitre by Lee Smith and Hartenberg and Strauch (23) reported a tumour of the right lobe of the thyroid. The basal metabolic rate has been investigated by various writers; it was normal in the cases investigated by Simons, Watson and Ritchie; increased due to hyperthyroidism in Smith's case.

Klien believes that the causal lesion is in the pineal but the experimental work of Izawa (24) has corroborated the clinical findings in pineal lesions, namely, precocious sexual development, gigantism, general adiposity, and somnolence; the classical picture given by Dickens in the "fat boy" in "Pickwick Papers"; anything more unlike the picture of lipodystrophia can hardly be imagined.

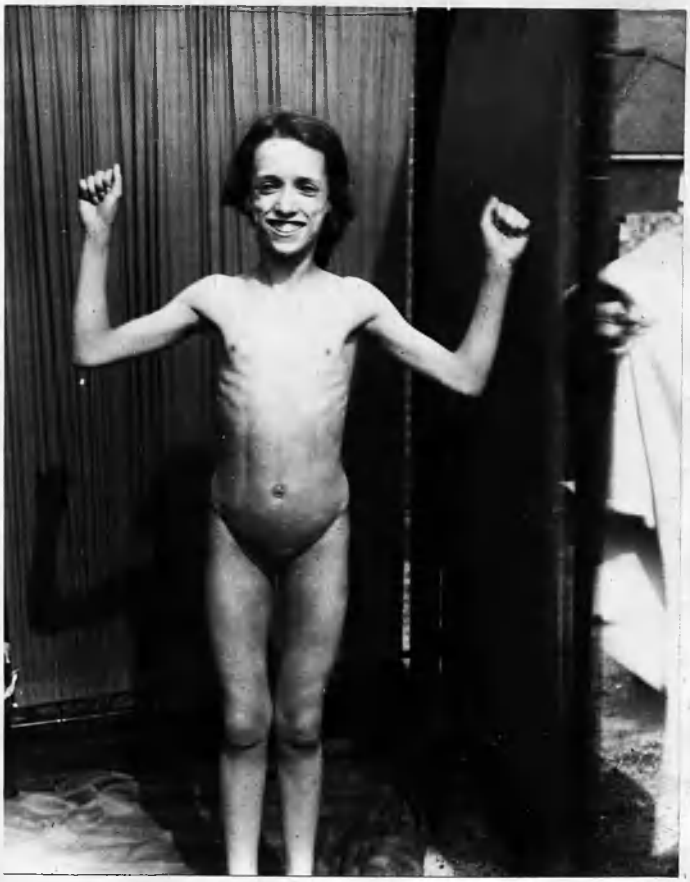
Klien's patient also showed polyuria; enuresis and rhinorrhœa, all these symptoms clearing up on exhibition of pituitary extract; without having any effect, however, on the lipodystrophia. Oliguria was noted by Gerhartz as in my second case. Simons, Christiansen, Smith and Husler had the pituitary fossa radiographed when it was found to be of normal size.

Two cases have come to post-mortem examination, one of Parkes Weber's, and one of Husler's; the former showed normal thyroid, pituitary, ovary and adrenal; in each case the fat in the body cavities was normal in amount and distribution; neither case threw any light on the cause of the disease.

Parkes Weber considers the disease to be due to an obscure lesion of the autonomic nervous system whereby a redistribution of the fat in the subcutaneous tissues takes place, and he uses the analogy of the redistribution of pigment in the skin in vitiligo.

My own observations lead me to agree with Parkes Weber up to the point that the lesion is an obscure one of the autonomic nervous system.

Fig. 13



The curious form of paræsthesia to which my first two cases is subject seems to be the only new point that I have elicted. I have made enquiries and cannot find anything similar amongst other patients who have consulted me for the many and varied complaints that present themselves to a practitioner in a busy industrial area like Sunderland, and the point that struck me in both cases was that this sensation was over the distribution of the 3rd division of the 5th nerve, in other words, in the area which was first affected (roughly) by the lipodystrophy.

This led me to speculate that the disease might be due to some obscure lesion of the autonomic system affecting the upper part of the system alone. Fat is probably synthesised from fatty acid and glycerol by an intra-cellular lipase [Halliburton (25)] after these components have passed through the cell-membrane. This lipase has two components, an inactive portion resident in the fat cell and an activator portion circulating in the blood.

Now it is conceivable that this action is under the control of certain fibres of the autonomic system and a lesion disturbing the function of these might result in a constant process of fat hydrolisis going on; the process being an isothermic one no loss of energy would take place. Alternatively it might be that no fat synthesis takes place; the explanation being again a disturbance of the nervous process regulating fat synthesis in the regions affected.

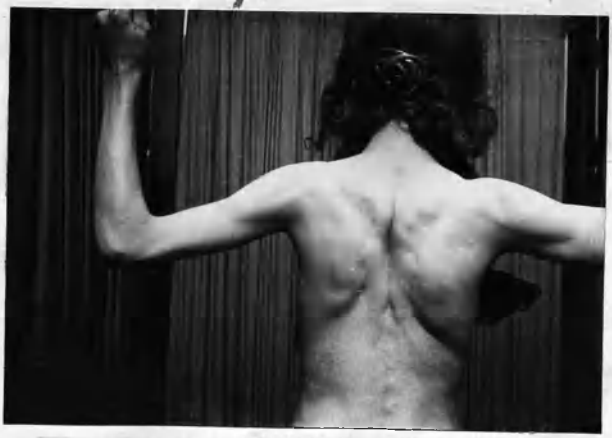
The rarity of the condition, the placing of these typical cases on record and so adding to the literature, together with the interest I had taken in the cases once I had recognised them for what they really were, led me to believe that if I could throw any light on the condition it might be of value to me as a subject to present as a Thesis for the M.D. Degree.

Whilst I am somewhat disappointed that I cannot definitely throw a clear flood of light on the lesion responsible for the condition, and so put the seal of finality on my work, I can only state that I have tried, with the limited material and facilities at my disposal, to elucidate the riddle and like the distinguished authorities I have quoted, failed.

I submit this Thesis for your consideration as a candidate for the M.D. Degree.

Fig. 14.

Fig. 15.



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PATIENT II.

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