The cyst was large enough to produce local midbrain pressure symptoms and internal hydrocephalus, probably intermittent.

Details of the histology are given.

Our thanks are due to our colleagues, particularly to Major R. C. Connolly, R.A.M.C., to Prof. Dorothy Russell for giving us her opinion on the sections, and to Dr. P. Daniel for taking the photomicrographs. We are grateful to our commanding officer, Col. G. D. Gripper, for permission to publish this case.

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AMYOPLASIA CONGENITA ASSOCIATED WITH HYPEROSTOSIS FRONTALIS INTERNA

BY

R. N. HERSON, M.B., B.S.

[WITH PHOTOGRAPHURE PLATE]

The absence of any reference in the literature to the combination of amyoplasia congenita and hyperostosis frontalis interna and the striking clinical picture resulting from this compound syndrome prompt me to record the following case.

Case Report

The patient was a woman aged 61 years. Since birth her lower limbs had been abducted at the hips and flexed at the hips and knees, her arms had been extended at the elbows and flexed at the wrists, and movements at all the joints had been severely limited. She learnt to shuffle round the floor, to feed herself, write, sew, knit, and do other forms of handicraft. She went to school from the age of 7½ to 9 years, was fond of reading, and was as apt as children of her own age. She was of a cheerful disposition, being despondent only when afflicted by pain. She had had headaches behind the eyes and over the frontal region as long as she could remember, the more severe attacks being accompanied by nausea and vomiting, but had never complained of her sight or of seeing double. At the age of 9 she had measles. Menstruation started when she was 11, and, although at times periods were regular every 28 days, she was subject to long spells of amenorrhoea even up to four years. She ceased to menstruate at the age of 54. When she was 19 she gained weight rapidly, and soon attained a weight of 11 stone (70 kg.). At the same time she developed in the beard area and on the upper lip a growth of hair which became sufficiently noticeable to require the use of a razor or depilatories.

At the age of 41 she began to get severe attacks of colicky pain in the upper part of the right side of the abdomen, radiating round to the right scapula. A diagnosis of gall-stone colic was made. Eighteen months later she was found to have a duodenal ulcer. This required two years' medical treatment. When 50 years old her gall-bladder was drained but not removed. Through the operation scar there developed a large incisional hernia, which has been a constant source of pain and distress.

At the age of 52 she complained of excessive thirst; sugar was found in her urine. She was given 15 units of insulin morning and evening, without any special diet, for four months. In 1941 she was admitted to Southmead Hospital, Bristol, on account of an abscess in the pubic region. Three years later she was again admitted with multiple boils and an anal fistula, for which she was operated on. Sugar was again found in the urine, and she was discharged on 40 units of insulin a day but no dietary restrictions; this did not render her sugar-free. She was re-admitted to Southmead Hospital on June 6, 1946, because of recurrent attacks of severe pain in the incisional hernia associated with vomiting. There was heavy glycosuria but no ketosis. In the surgical ward, in an attempt to control the glycosuria, the insulin was increased to 40 units four-hourly, but in spite of this each specimen of urine was loaded with

sugar. She never had an overdose. So far as can be ascertained from the notes she has never had ketonuria.

Family History.—Her father died at the age of 80. Her mother, who died at the age of 84, had nine pregnancies. The first was a boy, who died at 14 months after an accident; the second child was a girl, who is still alive, and whose only complaint has been cholecystitis. The next in the family was the patient, who was born when her mother was 36. There is no knowledge of any complications during the pregnancy or confinement. All the remaining pregnancies ended in miscarriages at the fourth month. There was no consanguinity either in the parents or in the grandparents.

Physical Examination.—The gross obesity coupled with the limb deformities made a striking picture (Plate, Fig. 1). Sitting height 32 inches (81 cm.), weight 11 st. 9 lb. (74 kg.). She had a large fat ruddy face with hair on the chin and upper lip. The lobes of both ears were webbed. The neck was short, and the neck and body were grossly obese, which contrasted with the thin upper and lower limbs. Abdominal fat sagged down and partially covered the drawn-up legs. The breasts were normally developed, and the hair on the head and body was of normal female distribution. The skin was greasy and subject to acne, but there were no striae or abnormal pigmentation.

Passive and active movements of the joints were approximately equal. Abduction of the shoulder-joints was limited to 15°. The arms were extended at the elbows and the forearms pronated. Movements of the elbow-joints were limited to 5°. The wrists were fixed in flexion and ulnar deviation. The finger-joints were flexed. She was able to flex fingers and thumbs to about 60°, but had practically no power of extension in the fingers. She was able to oppose her thumbs to any of her fingers, and thus grip an object between the fingers and thumbs. There was webbing of both axillary folds and of the first and second fingers of both hands. There were flexion and abduction at the hip-joints, flexion at the knee-joints, and bilateral equinovarus. She could flex the right hip-joint about 40°, and had moderately good power of abduction and adduction of the right femur. Movement of the left hip was more limited than that of the right. Movement was extremely limited in the knees and was absent in the ankle-joints. There was limited flexion of the toes. She had slight scoliosis, and mobility of the back was very limited.

The pupillary responses to convergence and accommodation were normal. Eye movements were full and the media and fundi were normal. Peripheral and central fields of both eyes were full to 3/330 and 3/1,000 mm. white object, respectively. Her visual acuity in the right eye was 6/9 and in the left 6/6 without glasses, and 6/6 in both eyes with correction. She had complete upper and lower dentures. There was no thyroid enlargement and no abnormal pulsation in the neck. The pulse rate was 70 per minute and the blood pressure 170/90 mm. Hg. The heart and lungs were normal. There was a large incisional hernia on the right side, painful on palpation. The liver and spleen were not felt and there was no glandular enlargement. Nothing abnormal was found on examination of the central nervous system.

Investigations.—Blood: Hb, 116% (Haldane); R.B.C., 5,720,000; W.B.C., 8,200 (neutrophils 62%, lymphocytes 31%, eosinophils 2%, monocytes 5%). Urine: specific gravity, 1027; acid; no albumin; sugar, 260 mg. per 100 ml.; no ketone bodies; urobilinogen not increased; bilirubin absent; deposit not remarkable. Blood calcium, 10.4 mg. per 100 ml.; blood phosphorus, 4.3 mg. per 100 ml.; serum phosphatase, 9 units; 24-hourly urinary excretion of calcium, 320 mg. Blood Wassermann and Kahn reactions negative. Both the glucose-tolerance and the insulin-tolerance estimations were performed at a time when the patient had been receiving a normal mixed diet. There was an insulin-resistant diabetes, as the following figures show: The blood cholesterol was 185 mg. per 100 ml. It was estimated on many occasions, but was not found to vary appreciably with the blood-sugar levels. The 24-hourly 17-ketosteroid excretion was determined on two occasions, and gave figures of 2.3 and 4.1 mg.

X-ray Examination.—Heart and lungs were normal. A congenital deformity of the first and second ribs on the right and a short first rib on the left were present. There were a bony bridging between the vertebrae, slight scoliosis, and fixation of

Blood-sugar Level (mg. per 100 ml.)

At	After Administration of 50 g. Glucose (Fasting, 210 mg.)	After 60 g. Glucose Orally and 10 units Insulin Intravenously (Fasting, 299 mg.)
$\frac{1}{2}$ hour	246	255
1 hour	275	290
1½ hours	250	310
2 "	236	294
2½ "	222	290

the right transverse process of L.5. The bones of the upper limbs were small. There were localized periosteal thickenings and deformity of the lower end of the right radius. The bones of the lower limbs were small and rarefied and there was bowing of the tibiae and fibulae. The tibial vessels were calcified. A skiagram of the pituitary fossa did not show any definite enlargement. Calcification was seen on its floor, extending anteriorly, and there was some erosion of the base posteriorly. The films were shown to Dr. J. R. Gilmour, who suggested a diagnosis of hyperostosis frontalis interna, and this was confirmed by a lateral skiagram of the whole skull (Plate, Fig. 2). This shows hyperostosis on the internal surface of the frontal bone without involvement of the outer table, large frontal sinuses, and over-pneumatization of the mastoid cells.

Insulin therapy was stopped and she was given a diet of 10½ oz. (297 g.) of meat or fish supplemented with green vegetables. On this diet there was considerable reduction in the amount of sugar excreted. The blood-sugar level three-quarters of an hour after a midday meal was 160 mg. per 100 ml., and at three and a quarter hours was 163 mg. The polyuria was largely controlled. She lost 9 lb. (4 kg.) in a month on this diet.

Discussion

The congenital deformities of her limbs are those of amyoplasia congenita. Apart from headaches, which she has had as long as she can remember, the other main features of her condition point to a pituitary or a pituitary-hypothalamic disturbance starting at the age of 19. These features are a large fat ruddy face, hirsuties, a buffalo distribution of fat, greasy skin, acne and boils, a history of irregular menstruation, insulin-resistant diabetes (detected at the age of 52), hypertension, and a haemoglobin of 116%. The combination is suggestive of Cushing's syndrome but lacks some of its important features. The lateral radiographs of the skull establish the diagnosis of hyperostosis frontalis interna; in a few cases of this condition calcification has been seen, as in this patient, in the sella turcica. It is now generally accepted that this cranial dysplasia is a manifestation of pituitary dysfunction (Knies and Le Fever, 1941). In the description and classification of a large series of pituitary cranial dysplasias by Mortimer, Levene, and Rowe (1937) there are references to hyperostosis frontalis interna, though they do not use the term themselves. Both basophilic adenomatous masses and eosinophil adenomata have been found at necropsy in the pituitary glands of some of these cases. The low excretion of 17-keto-steroids is probably accounted for by the patient's age: Frazer *et al.* (1941) found that the daily output was low in the elderly. Henschen (1936) has suggested that in hyperostosis frontalis interna a hereditary influence determines the frontal localization of the bony changes, and it may be that there is in this case a common genetic factor responsible for both the limb deformities and the cranial dysplasia.

I wish to thank Dr. Phillips, Medical Superintendent, Southmead Hospital, Bristol, for permission to publish this case, and Dr. A. C. Crooke and Dr. J. R. Gilmour for their valuable advice.

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TUBERCULOSIS OF THE GREAT TROCHANTER

BY

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[WITH PHOTOGRAPHIC PLATE]

Tuberculosis of the great trochanter with involvement of the overlying bursa was originally described by Teale in 1870. Since that time references to this condition in the literature had been meagre until Meyerding and Mroz (1933) reported nineteen cases. The present series comprises twelve cases treated in Liverpool and Wrightington during the last fifteen years.

Incidence

None of the previous papers on this subject have given any indication as to its frequency. In this collection of case histories I have found the incidence to be just under 2 per 1,000 cases of tuberculosis. It has been stated by Clapton (1919) that it does not occur in children, but in this series the youngest patient was 7 years old and there was also one patient of 14 years. The average age of the remaining patients was 28 years, which is comparable with the average of 35 years quoted by Meyerding and Mroz. Nevertheless, as the total number of cases recorded in the literature is only 41 this figure cannot be considered large enough to indicate a definite age period.

Aetiology

Two factors are of importance. The first is the question of trauma: the superficial position of the great trochanter makes it peculiarly liable to injury, and severe blows on this bony prominence are an everyday occurrence. However, a history that recent trauma was followed by swelling or pain associated with tuberculosis of the great trochanter does not immediately suggest this as the principal aetiological factor, since the underlying disease may have already been present for some time, and the trauma may merely have been responsible for drawing the patient's attention to it. Nevertheless, three of the patients in the present series gave a history of considerable trauma to the great trochanter, although two of them remembered having had occasional aching pains referable to the hip for several months before the accident. Secondly, a considerable proportion of these patients were suffering, or had suffered, from tuberculosis in other situations. In the series described by Meyerding and Mroz (1933) 73% had other tuberculous lesions. In the present series, however, only five patients out of twelve (42%) had any other demonstrable tuberculous lesion. The site of this lesion varied: two patients had pulmonary tuberculosis, one had cervical adenitis, one a retropharyngeal abscess without a bony focus, one tuberculous arthritis of the knee, and one tuberculosis of the sacro-iliac joint of the opposite side.

Symptoms and Signs

This condition may present in two ways: usually the patients notice a swelling of increasing size over the great trochanter, or they may complain of pain or aching of a mild type over the outer side of the thigh, apparently referred to either hip or knee. On examination a fluctuating swelling of varying size is usually found, covering the outer aspect of the greater trochanter, and extending backwards over the buttock and downwards for a varying distance (in two cases it reached the level of the lower

RHEUMATOID ARTHRITIS AND AMYLOID DISEASE: W. YEOMAN AND J. V. WILSON

FIG. 1.—Kidney. Showing degenerative changes in glomeruli, tubules, and interstitial tissue.

FIG. 2.—Liver. Amyloid spreading in from periphery of lobule. Liver cells show degenerative changes.

FIG. 3.—Spleen. Malpighian corpuscle showing amyloid infiltration.

FIG. 4.—Suprarenal cortex. Showing amyloid infiltration and some cellular degeneration.

AMYOPLASIA CONGENITA ASSOCIATED WITH HYPEROSTOSIS FRONTALIS INTERNA: R. N. HERSON

FIG. 1.—The patient at the age of 61.

FIG. 2.—Lateral radiograph of skull showing hyperostosis on internal surface of frontal bone and calcification of base of pituitary fossa. The frontal sinuses are large and the mastoids over-pneumatized.