Short Notes and Clinical Cases.

A CASE OF ADIPOSO-GENITAL DYSTROPHY OCCURRING IN A MONGOL.

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IN spite of the popularity of the view that mongolism is in some way related to a disturbance of the function of one or other of the ductless glands definite evidence of endocrine disorder belonging to a recognised clinical type has seldom been recorded, and it may therefore be of some interest to describe the following instance of hypopituitarism occurring in an adult mongol.

CLINICAL CASES.

History.—E. S., a typical mongolian imbecile, age 33, was admitted to Darenth Training Colony on January 26, 1922. Both parents were dead and no previous history could be obtained. On her transfer to Leavesden Mental Hospital in May, 1923, she weighed 7 st. 4 lbs. At that date there was no evidence of obesity, but early in 1926 she commenced to put on weight and has continued to do so in a slow and progressive manner ever since, and this in spite of prolonged treatment with thyroid extract.

Present Condition.—The patient is an adult female mongol, 52½ inches in height, and 14 st. 2 lbs. in weight. The extreme degree of obesity displayed by the patient hampers her movements and makes physical examination somewhat difficult. Her head is small, rounded and bradycephalic; circumference 19¾ inches; cephalic index 77. The palpebral fissures are small but not narrowed or oblique: there is marginal blepharitis and conjunctivitis. The lids are thickened and there is a slight degree of ectropion. The eyes are closely set and slightly prominent. Her nose has a depressed bridge and is short, squat, and with the nostrils directed forwards as well as downwards. The tongue is large with irregular fissuring near the tip and shows hypertrophy of the circumvallate papillæ. Her ears are small, rounded and shell-like. With the exception of two incisors and one molar, all the teeth are missing. The cheeks are enormous and have a leaden flush; the upper lip is everted. The palate is high and narrow; adenoids and enlarged tonsils are present. The patient suffers from rhinitis and bronchitis, her voice being deep and husky. The hands are broad, clumsy and flabby, with short thumbs and little fingers. The digits are slightly tapered and the nails ribbed longitudinally. The feet are broad, swollen and cyanosed; the cleft between the first and second toes is large.

Both upper and lower limbs are relatively short and show excessive mobility at the larger joints. The abdomen is large and protuberant. The skin is rough and dry, showing pigmentation on the neck. The hair on the scalp is abundant but there is none in the axillæ and little on the pubes. A slight moustache is present and there is hair on the

sacral region. The extremities are blue and cold; chilblains, and cuticular mottling on the trunk are present. The heart sounds are muffled and weak; pulse 76 per min. Blood pressure 90/45 mm. Any sustained effort such as climbing stairs produces severe dyspnoea.

Her pupils are small and react rather sluggishly to light. Ophthalmoscopic examination shows partial optic atrophy on both sides. Vision is impaired, though how much it is difficult to say owing to lack of co-operation on the part of the patient.

A case of adiposo-genital dystrophy occurring in a mongol.

Abdominal reflexes absent; plantars normal; knee jerks present. Her gait is slow, clumsy and waddling. Menstruation, which was always scanty and irregular, ceased in August 1930.

As will be seen from the accompanying photograph, a remarkable degree of obesity is present. Her cheeks are huge, giving her a marked double chin, and the circumference of her neck is only a quarter of an inch less than that of her head. The breasts are very large and pendulous. Although the girth of the arms is increased, it is particularly on the pectoral, girdle and femoral regions that the chief deposition of fat has occurred.

No actual œdema is present, but the subcutaneous tissue has a boggy feeling. Owing to the enormous girth and weight of her abdomen, she has difficulty in maintaining the erect posture and if asked to stand upright after a few seconds she allows her body to sag slowly forwards.

Laboratory Findings.—The serum Wassermann reaction is negative. Red blood corpuscles 3,860,000; white blood corpuscles 8,800; hæmoglobin 85 per cent.; colour index 1.1. Differential leucocyte count: Polymorphs 63.4 per cent.; small lymphocytes 24.5 per cent.; large lymphocytes 6.4 per cent.; eosinophils 3 per cent.; mast-cells 1.5 per cent.; basophil myelocytes 0.75 per cent.

Glucose tolerance tests gave the following values:—

Fasting.—116 per cent.; half an hour after glucose, 153 per cent.; one hour after glucose, 241 per cent.; two hours after glucose, 219 per cent.

The urine contains no abnormal constituents. X-ray examination shows no enlargement of the pituitary fossa.

Mental State.—The patient is a low-grade imbecile who requires assistance in dressing and washing. She calls both a halfpenny and a penny by the name of the former and is unable to use a knife and fork properly. Formerly clean in her habits, she now neglects the calls of nature. She remains seated for hours on end in a state of somnolence. Her appetite is moderate.

DISCUSSION.

According to Gondal¹ the adiposo-genital pituitary syndrome of the adult is characterised by (1) rapid and progressive obesity; (2) atrophy of the genital organs and disappearance of the sexual functions; (3) association of cerebral and visual disturbances. This triad of symptoms is shown in the case reported above; the patient's weight has almost doubled in the last five years, menstruation has ceased, and vision is failing owing to partial optic atrophy. In addition, the patient shows a marked tendency to sleep during the day. There can be little doubt therefore, that the patient is suffering from hypopituitarism of the Fröhlich type, possibly the consequence of disease of the posterior lobe of the pituitary body. With the exception of one doubtful case reported by Galant², no similar examples appear to have been previously reported, but the possibility of some connection between mongolism and the ductless glands has for long been entertained, and some writers have brought the pituitary gland under suspicion. Timme³, for example, who has called attention to the frequency with which mongols show subnormal and disproportionate body growth coupled with lack of genital development on X-ray examination, found abnormality of the anterior portion of the pituitary fossa in twenty-three mongols—the change consisting in an excavation under the anterior clinoid processes and communicating with the anterior portion of the fossa itself. On the basis of these findings he suggests that a disturbance of the anterior portion of the pituitary body might readily produce many of the symptoms shown by mongolian idiots. Timme's roentgenological findings have not been corroborated and it is more than doubtful whether slight deviations in the size and form of the pituitary fossa are of any particular significance, and as Enfield⁴ has pointed out, unless evidence of erosion of the long structure of the sella turcica can be demonstrated it is hazardous to draw any conclusions.

Moreover, since hypopituitarism is said to be a symptom of implication of the posterior lobe Timme's speculations concerning the anterior lobe are in no way supported by the presence of the Fröhlich syndrome and it seems legitimate to conclude that the association of the latter with mongolism is purely accidental.

REFERENCES.

- ¹ GONDAL, *Thèse de Paris*, 1918.
- ² GALANT, *Med. Klin.*, 1927, vii, 250.
- ³ TIMME, *Arch. of Neurol. and Psychiat.*, 1921, v, 568.
- ⁴ ENFIELD, *Jour. Amer. Med. Assoc.*, 1922, lxxix, 934.