Acromegaly Presenting with Hemiplegia

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SUMMARY

A case of acromegaly presenting with hemiplegia is described. The radiological features, including cerebral angiography, are discussed.

Acromegaly is uncommon in the Black population of South Africa. Experience at Baragwanath Hospital (2 400 beds) confirms the rarity of the disease, and reference to the *South African Medical Journal* over the past 25 years has failed to reveal a single case.

In all published series no case of 'stroke' in acromegaly is specifically described. This is unusual because of the many underlying aetiological factors of potential stroke present in acromegaly.

S. Afr. Med. J., **48**, 684 (1974).

The following is a report of acromegaly in a Black woman with the unusual presentation of a cerebrovascular lesion.

CASE REPORT

A 48-year-old Black woman was admitted to Baragwanath Hospital in January 1973 with a history of left-sided paresis which had developed over a period of 8 hours. She had complained of headaches for the past 5 years, and had recently suffered from palpitations and backache. She had noticed progressive enlargement of her hands and feet during the previous 9 years and now needed shoes which were 3 sizes larger. All her teeth were removed in 1960. She had been menopausal for 3 years and there was no family history of diabetes mellitus.

On examination she was found to have an acromegalic facies, large hands, feet and tongue. The blood pressure was 175/110 mmHg and the heart showed left ventricular hypertrophy with no evidence of failure. There was a flaccid left hemiplegia with hemi-anaesthesia to pin-prick on the same side. The visual fields on the Bjerrum screen showed non-specific changes due to bilateral early cataracts. The left fundus was normal and the right fundus showed mild sclerosis of the arteries. The optic discs were normal. There were no other cranial nerve abnormalities.

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Laboratory Investigations

The full blood count was normal. Blood urea was found to be 46 mg/100 ml, serum sodium 137 mEq/litre, serum potassium 3,9 mEq/litre, serum chlorides 103 mEq/litre, serum calcium 4,5 mEq/litre, and serum phosphate 4,8 mg/100 ml.

Cerebrospinal fluid analysis showed pressure to be 140 mm H_2O; protein 85 mg/100 ml; sugar 73 mg/100 ml; chloride 118 mEq/litre, and cell count 5 lymphocytes. Kolmer reaction was negative.

Tests of thyroid function revealed the protein-bound iodine to be 8 μg/100 ml; T_3 uptake was 108% (normal range 94 - 124%); and T_4 iodine was 5,3 mEq/100 ml.

Repeated tests of plasma cortisol showed normal levels.

Gonadotrophin tests showed urine luteinising hormone excretion to be 51 IU/24 hours; urine 17 - hydroxy-corticosteroids 12,5 mg/24 hours; and urine 17 - ketosteroids 11,7 mg/24 hours.

Serum lipid tests showed the serum as slightly opalescent, and total lipids were markedly raised to 1 375 mg/100 ml; cholesterol 236 mg/100 ml; triglycerides 670 mg/100 ml and phospholipids 350 mg/100 ml.

Initial glucose tolerance tests showed normal glucose tolerance and subsequently showed evidence of diabetes mellitus.

During a glucose tolerance test serum growth hormone levels were raised, with failure of significant suppression. Water deprivation test was normal.

An electrocardiogram showed a normal frontal plane QRS axis and no evidence of left ventricular hypertrophy. The T waves were rather low to inverted in leads V4 - V6, standard leads I, II and III, leads AVL, AVR and AVF. These changes were thought to be non-specific.

Radiological Findings

Skull: There was enlargement of the pituitary fossa with a double floor (Fig. 1), undercutting of the anterior clinoid processes, and thinning of the dorsum sellae. The frontal and parietal bones were thickened. Prognathism was present. Bilateral carotid angiography demonstrated ectasia of the internal carotid artery. In addition, the supraclinoid part of the internal carotid artery was elevated (Fig. 2).

Chest: Cardiomegaly was present with left ventricular enlargement.

Hands: The ends of the phalanges were widened with squared margins and characteristic proximal pointing of the tufts. The fourth metacarpals were hypoplastic. The sesamoid index was increased and measured 42.

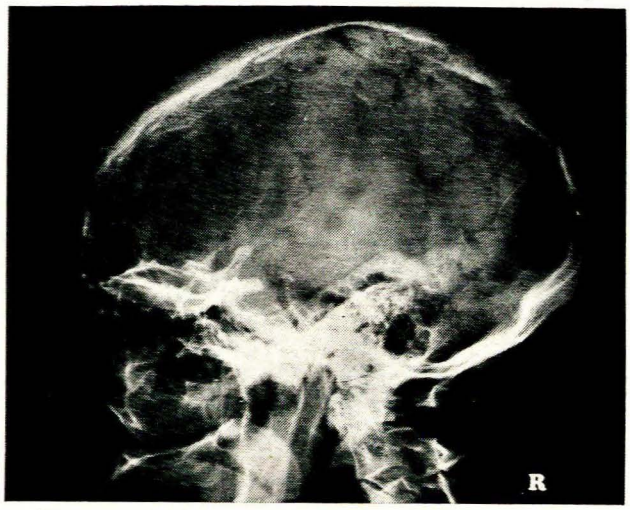

Fig. 1. Lateral radiograph of the skull, demonstrating enlargement of the pituitary fossa with a double floor, undercutting of the anterior clinoid processes, and thinning of the dorsum sellae.

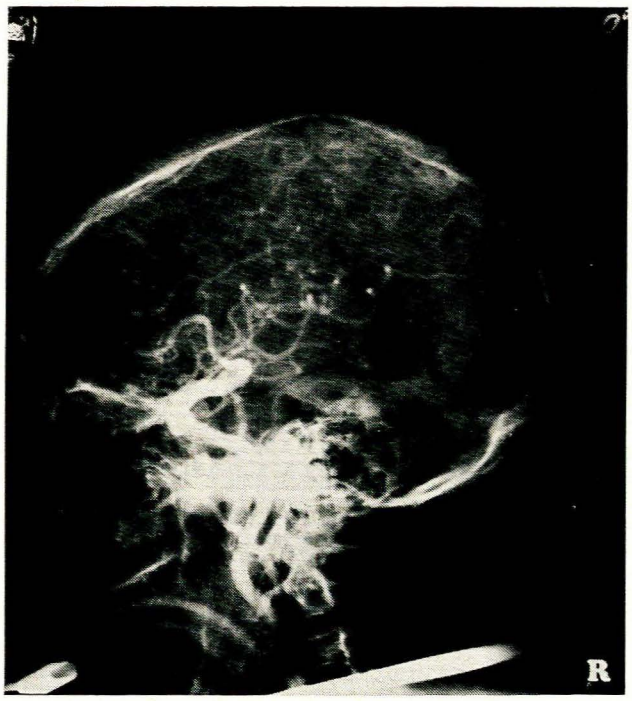

Fig. 2. Lateral view of skull, demonstrating ectasia of the internal carotid artery.

Dorsal vertebrae: The middle and lower dorsal vertebrae were increased in their anteroposterior diameter. Scalloping was present posteriorly.

Pelvis: Spurring was present at the margins of the acetabulae. Thinning of the pubic bones was present, with breaking at the superior margins of the symphysis.

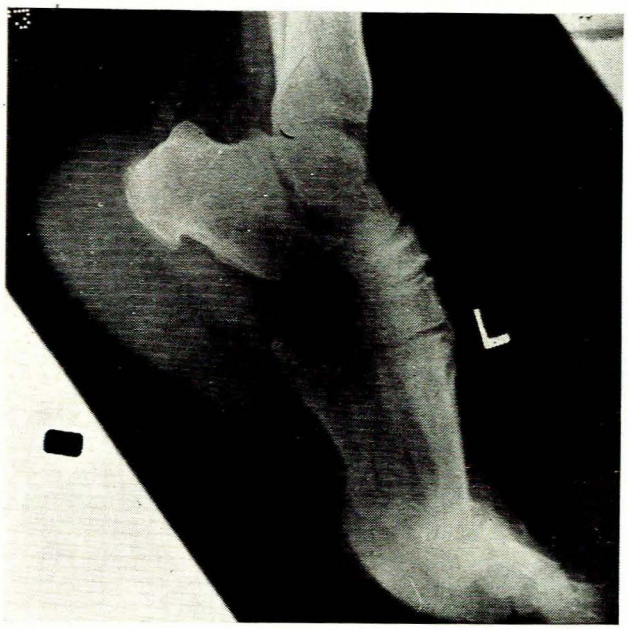

Fig. 3. Lateral radiograph of the foot and heel, demonstrating increased heel pad thickness.

Heel pad thickness: The heel pad was thickened, measuring 36 mm (Fig. 3).

Treatment

The patient was given a course of 5 000 rads of ^{60}Co teletherapy to the pituitary gland, with little improvement. She refused surgical intervention.

DISCUSSION

The patient presented with the typical clinical features of acromegaly. Indicative of activity were the signs of progressive acral enlargement, persistent headaches, elevation of the serum inorganic phosphate, and the raised levels of serum growth hormone with failure of significant suppression after oral glucose.

The patient also had evidence of hypertension and cardiomegaly. Hypertension is commonly observed and the reported incidence approximates 30%.[1,2] The aetiology of the hypertension is unknown, but treatment of the primary disease is often associated with an improvement.[2]

The cardiomegaly may be a reflection of visceromegaly, hypertension or coronary artery disease. The increase in heart size may, however, be out of proportion to the general splanchnomegaly, and be due to a myocardiopathic effect of excess growth hormone. Myocardial failure has also been reported in the absence of other causes of heart disease.[3]

Pepine and Aloia[4] reported a case with severe myocardial dysfunction which showed significant improvement following hypophysectomy, thus indicating a probable myocardiopathic effect of excessive growth hormone.

Radiological Findings

The classical features of acromegaly are demonstrated.

Dilatation of the carotid arteries: Hatam and Greitz[5] have shown that dilatation of the cerebral arteries is a common finding in individuals with eosinophilic adenoma and acromegaly. In their series no correlation between the dilatation of the arteries and blood pressure was noted. They postulate that a more plausible explanation would be that arteriectasis is part of the splanchnomegaly and hence governed by the growth hormone secretion.

Significance of the sesamoid bones: Steinbach *et al.*[6] described large sesamoid bones in patients with acromegaly. Measurements were first used by Kleinberg *et al.*[7] The technique is performed by taking an X-ray film with palms flat on a cassette. The greatest diameter in millimetres of the medial sesamoid at the metacarpophalangeal joint of the thumb is measured and multiplied by the greatest diameter perpendicular to the first one. In the series of Anton,[8] the upper limit of normal was 40 for males and 32 for females. The sesamoid index in this patient was increased to 42. Use of the sesamoid index is suggested as an aid in the diagnosis of acromegaly.

Significance of the vertebral changes: The vertebral changes are due to new growth of bones and intervertebral discs (mainly anteriorly) on the middle and lower thoracic regions. The lumbar vertebrae are usually not affected. There may be scalloping of the vertebrae posteriorly. This is thought to be due to soft-tissue hypertrophy in the neural canal, producing secondary erosion.

Heel pad thickness: Measurement of the heel pad thickness appears to be one of the most accurate radiological methods available in the diagnosis of acromegaly. Other causes of soft-tissue thickening must be excluded, viz. injury, infection, obesity, peripheral oedema due to cardiac failure, venous obstruction and myxoedema. An increased heel pad thickness may also be found in Black subjects. The upper limit of normal is considered to be 22 mm. Kho *et al.*[9] have shown that the heel pad thickness is related to the serum level of growth hormone and to the duration of acromegaly. Their findings suggest that a reduction in the serum growth hormone level greater than 50% is necessary for a significant reduction in heel pad thickness to occur.

Hemiplegia

In our patient this is of special interest. The hemiplegia developed fairly suddenly over 6 - 8 hours, with arm and face more involved than the leg. There was associated hemi-anaesthesia. The accompanying headache was not severe and did not last longer than a few hours. The lesion responsible for the hemiplegia was probably a thrombus or an embolus in the area of the middle cerebral artery.

A thrombus could be secondary to atheroma caused by hypertension, a diabetic or hyperlipaemic state. An embolus could have arisen in the heart or carotid artery. A carotid arteriogram was performed to see whether lateral extension of the pituitary tumour could have encroached on the internal carotid artery, thereby compromising the distal blood supply or causing distortion which may be the origin of an embolus. It was found that the arteries were markedly ectatic on both sides and no intimal roughening was observed.

The association of acromegaly and hemiplegia in this case may indeed be fortuitous. However, the presence of so many of the precipitating factors for the development of a cerebrovascular accident would suggest a causal relationship. It is surprising that this manifestation has not been recorded more frequently.

Therapy

Treatment should be instituted whenever the disease is active. Progressive visual impairment is an indication for surgical resection, either by craniotomy or via the sphenoidal sinus. Patients who do not have visual defects are treated by high-voltage external irradiation. External irradiation achieves a satisfactory fall of growth hormone levels in only about half the patients, and was without appreciable effect in this patient.

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