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ACROMICRIA, AN INTERESTING CLINICAL PICTURE

BY

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Acromegaly is widely known, but the opposite condition — acromicria — has been the subject of very few reports in the literature.

Brugsch was the first to describe acromicria, in 1927. He published a description of a case, a woman of 23, who had been quite normal up to the age of 13, when her fingers and toes gradually became smaller and thinner. At the age of 20 the menses, which had been normal up till then, ceased, and diabetes insipidus was diagnosed. Intense acrocyanosis appeared in the extremities, especially in the hands. Roentgenological examination showed a distinct reduction in the calcium content of bone in the terminal phalanges of the fingers and toes. The sella turcica was small. These roentgenograms are unfortunately not included in the report. *Brugsch* assumed the syndrome to be due to aplasia of the adenohypophysis *Bársony,* in 1933, described the clinical picture of acromicria more fully, and in addition to the symptoms mentioned by *Brugsch,* observed that the nose, ears and lips became sharp, and thin and the chin less prominent. He found that the symptoms were spontaneously relieved during pregnancy and were sometimes absent after delivery for as long as eight years, but then relapse occurred.

As far as I know, such patients have not been subjected

to clinical-chemical or hematologic examinations, apart from routine blood studies, nor have they been examined for histological changes in the anterior lobe of the pituitary gland, as no autopsy had been performed in any of the cases reported.

A fairly extensive search of the Scandinavian literature failed to reveal any reports dealing with this disease, though *Wahlberg* in 1934 described 3 patients with »asthenia gravis hypophyseogenea«, all of whom had madonna fingers or, in his own words »the upper extremities showed a certain tendency to acromicria«.

As this disease is rare but, on the other hand, extremely interesting, it seems desirable to publish details of 2 cases observed by me.

Case 1. The patient was a machine draughtsman, aged 30, unmarried, with nothing informative in his family history. He complained that his ears, fingers and toes had been sensitive to cold as early as the time when he was attending elementary school. Since 1939 — when the patient was 18 years old — the hands and to a lesser extent the feet, had been bluish and cold. He was admitted to the Hospital of the Finnish Red Cross, the diagnosis being dystrophia hypophysaria. A bilateral cervicothoracic ganglicctomy was performed. After this the hands showed only slight cyanosis for two years and were not so cold, but then the condition became worse again. In addition to cyanosis and a sensation of cold, the hands were stiff and the fingers had gradually become thinner since 1938; both index fingers especially had shortened and their nails had become short and convex. A kind of tightness resembling cramp often developed in the toes. Since the late summer of 1949 the patient had experienced fatigue and lassitude. He had a curious feeling of tightness in and under the chin. The appetite was normal and he had not lost any weight. Bowel function was normal. The secretion of urine showed nothing abnormal. The patient denied venereal infection. Libido and potency were normal. He was a nonsmoker and teetotaller. He was admitted to the Central Military Hospital I on Jan. 16, 1950.

Fig. 1.
Case 1.

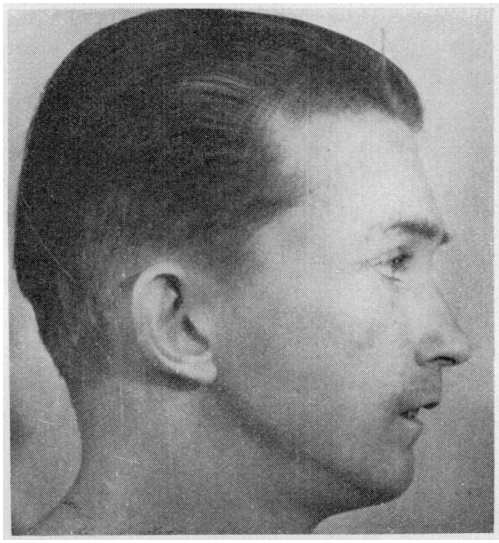

Fig. 2.
Case 1.

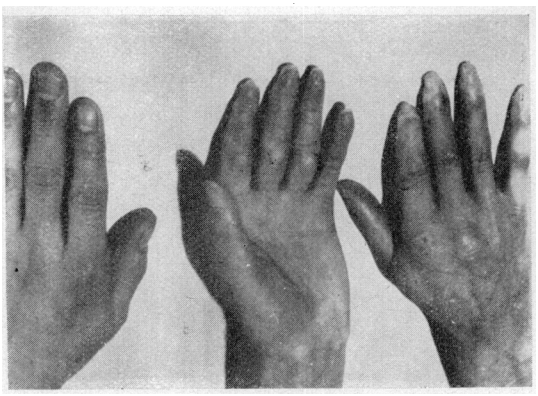

Fig. 3.
The hands of Case 1, compared with a hand from a normal person (to the left).

The patient was of delicate build, height 177 cm., weight 60 kg., arm span 173 cm. Thyroid gland was normal. On both sides of the neck, in the area of the supraclavicular fossa, there was an operation scar. The hair and beard were normal, and there was a masculine distribution of hair. Colour and moistness of skin were normal, apart from the cyanosis of the hands starting somewhat proximal to the wrist. The dorsal surface of the feet was only slightly cyanotic. The skin of the hands was thin, smooth and shiny, the nails were small and deformed, and the terminal phalanges of the index fingers especially appeared short. The fingers were thin and tapering (Fig. 3). Sensitivity to touch, pain and heat was normal. There was no difficulty in making a fist. The range of movement of the fingers was normal. The patient had a clear handwriting. The changes in the toes were similar to those in the fingers but less pronounced. His walk was normal. The lips were thin. The upper anterior teeth were partially carious and prominent. The mouth appeared pursed. The lower anterior teeth were too close together and arranged crosswise. The palate was high and the tongue thin and pointed. The patient spoke clearly. He had a bird-like profile (Figs. 1 and 2).

The sella turcica was very small, and in the lateral view

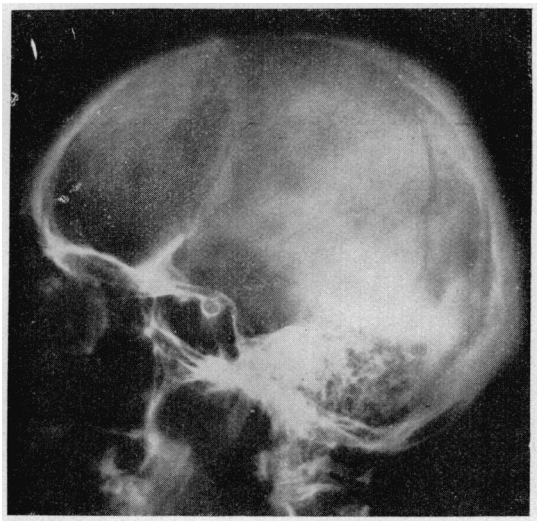

Fig. 4.
Skull of Case 1.

presented a circular appearance. The clinoid processes were very close to each other. No bone destruction was seen in the sella (Fig. 4). In the terminal phalanges of the fingers some changes appeared; malformation was most conspicuous in the

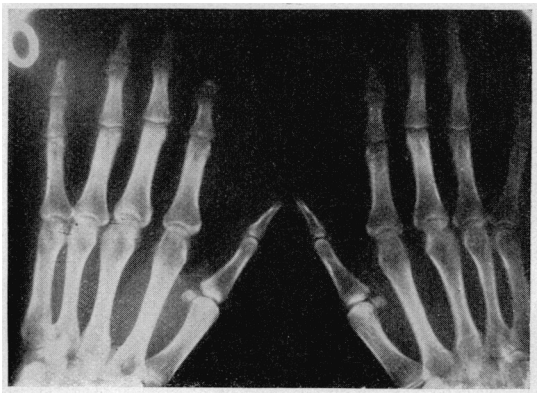

Fig. 5.
Phalanges of Case 1.

right index finger. Only the proximal end of the phalanx and a small part of the corpus remained; the tuberositas unguicularis was completely absent (Fig. 5). In some other fingers and in the left big toe the tuberositas unguicularis was beginning to disappear.

Examination of the lungs yielded nothing of importance. The heart was slightly displaced medially; on auscultation no pathologic changes were observed. The electrocardiogram was normal and the blood pressure 130/80. The radial artery was straight, and its wall smooth. The pulse was normal. The a. dorsalis pedis was normally palpable. Reflexes were normal, eyegrounds normal, pupils reacted normally. Trousseau's and Chvostek's tests were negative. The abdomen showed no abnormality. Genitalia were normally developed.

Hemoglobin (Sahli) was 93 per cent, Erythr. count 5.55 mill., Index 0.89, Leukoc. 8900. The differential count showed no abnormal changes, blood platelets numbered 261,000/cu. mm. The sedimentation rate was 2 mm. Wassermann, cholesterol Wassermann and Kahn tests were negative. The prothrombin index was 97, galactose test normal, Takata-Ara test negative, acid serum phosphatase 0, alkaline phosphatase 1.7 Bodansky units, inorganic phosphorus 3.0 mg. per hundred ml., serum calcium 11.1 mg. per hundred ml., sodium chloride 633 mg. per hundred ml., non-protein nitrogen 28 mg. per hundred ml., total cholesterol 180 mg. per hundred ml., ester cholesterol 128 mg. per hundred ml., and serum lipid phosphorus 8.5 mg. per hundred ml. — The blood sugar was 82 mg. per hundred ml.; it rose during the sugar tolerance test (1 Gm. glucose per kg. of body weight) to a maximum 142 mg. per hundred ml. in 30 min. and fell in 2½ hours to the fasting value (no sugar appeared in the urine during the test).

Case 2. The patient was a widow aged 37, who was admitted in 1948 to the Second Medical Clinic, the diagnosis being sclerodactylia (M. Raynaud). Insuff. valv. mitr. cord. et insuff. cordis.

Unfortunately I have had no opportunity of examining the patient and therefore quote from the clinical report.

Development in childhood had been normal. Menstruation began at the age of 16. The patient has three children. She ceased to menstruate at the age of 30.

At the age of 27 the patient had polyarthritis and rheumatic heart disease. The joint symptoms were initially severe but for a few years before admission had been milder; all signs of infection disappeared and the sedimentation rate became normal. However, her congestive heart failure and contractures were disabling.

At about the time when polyarthritis started she noticed that the skin of the hands gradually became thin and shiny.

The patient's height was 160 cm., weight 57 kg. Flexion contracture of the finger joints of both hands was observed. The thumbs were slightly less affected. Wrist movements were limited. The toe joints showed contractures. The skin of the hands was pigmented, hard, shiny and atrophic. Examination of the heart showed mitral regurgitation. According to a report from the Otolaryngologic Clinic the soft parts of the nose were atrophic suggesting lupus, but the skin was free from

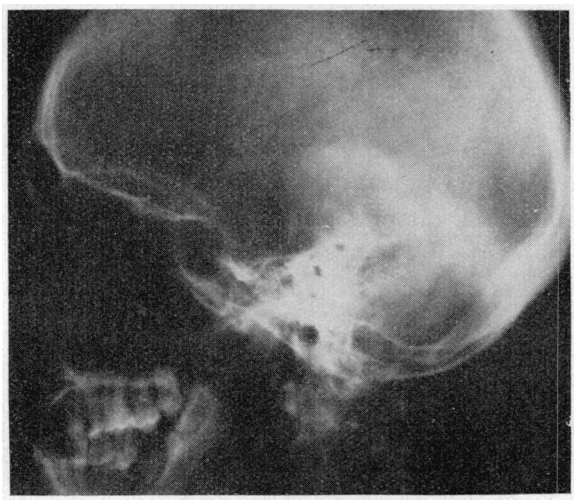

Fig. 6.
Skull of Case 2.

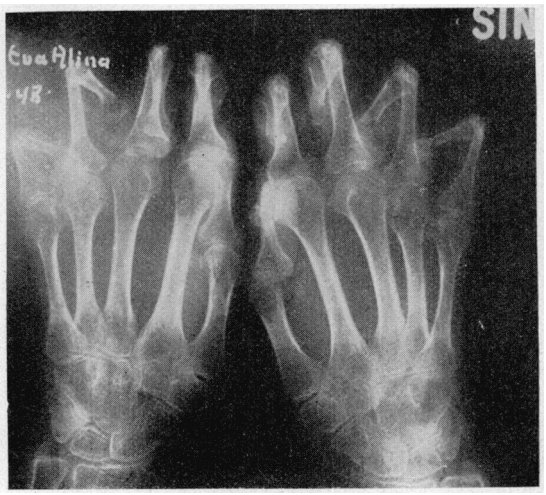

Fig. 7.
Phalanges of Case 2.

scars and elastic; the mucous membrane of the nose was atrophic, the tongue small. Both auricles were small, and of low elasticity. Gynecologic examination showed small, firm ovaries.

Roentgenological examination showed a fairly small sella turcica, osteoporosis of the clinoid processes, but no definite destruction (Fig. 6). Roentgenograms of the fingers showed contractures; in the terminal phalanges of some of the fingers only the proximal part remained while in others it was wholly absent and the pathological process extended in places to the peripheral part of the middle phalanx. In the soft tissues of the left thumb and index finger there were granular calcium deposits resembling those found in calcium gout (Fig. 7).

Hemoglobin (Sahli) was 80 per cent. Erythr. count 4.64 mill., Index 0.87, Leukoc. 4600. The differential count showed no changes. The sedimentation rate was 2 mm. Wassermann, cholesterol Wassermann and Kahn tests were negative. Total serum protein was 8 per cent, chlorides 599 mg. per hundred ml., calcium 9.4 mg. per hundred ml. The basal metabolic rate

was 32 per cent, which probably could be attributed to the congestive heart failure.

In Case 1 pituitary grafting was performed on Jan. 25, 1950: the hypophysis of a calf which had been killed half an hour previously was implanted in the pectoral muscle. The next day the patient stated that the feeling of tightness under the chin had disappeared and his hands felt warm. He stayed at the hospital for one week and during this time the cyanosis in the hands disappeared almost completely. The wound healed by primary intention. Three weeks after the implantation the patient wrote: »The hands continue to be warmer and more supple than previously. Their colour is also good, and they become slightly blue only after lunch but their normal colour returns later in the afternoon. The blue tinge of the skin on the feet has disappeared, they have become warm and I have no more cramps. The sensation of tightness on the neck and chin has disappeared. Before the operation I suffered from extreme fatigue and from a feeling of oppression. I was afraid I should soon lose my reason and not live long. If my present condition continues I shall soon be very well«.

On May 2, 1949, i. e. more than four months after the operation, the patient was seen again and said that he still felt all right. His condition corresponded with the description given in the letter. – – Treatment with growth hormone Phyol (Benzon) was instituted at the beginning of June 1950.

In Case 2 the syndrome was not clearly that of acromicria. The symptoms were complicated by a mitral regurgitation and congestive heart failure. The changes in the bones and joints seen in the roentgenogram may have been due to the previous polyarthritis, but acromicria appears to be a contributory factor. It is conceivable that the rheumatic infection had inhibited the secretion of pituitary somatotropin, possibly by producing thrombi and embolisms.

Case 1, on the other hand, is a genuine deficiency condition but it is difficult to decide what was the primary cause of the reduced production of somatotropin from the anterior lobe of the pituitary. It is difficult to believe that the number

of cases of acromicria is actually as low as that indicated by the literature. Probably the majority of cases have been diagnosed as a sclerodermia syndrome or as sclerodactylia or acrosclerosis. In view of the general recognition given to the term acromegaly, it seems reasonable to pay more attention to the corresponding state of hypofunction, acromicria.

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