Medical Memoranda

Albright's Syndrome

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CASE REPORT

A Hindu girl aged 7 presented to the casualty department on 18 August 1965 complaining of pain in the right hip and gradual deformity of her legs. Menstruation had begun about four months previously, and her breasts had gradually developed during the previous two years.

For two years the mother had noticed that the patient's legs were becoming bent. Her knees were now knocking each other. Since birth the child had had a peculiar pigmentation of the skin of the right arm and the right side of the chest, both front and back. It was also noticed that for the past two years both breasts were becoming enlarged. In April 1965 she had her first period, lasting for two days; she was then 6 years and 8 months old. Her next periods were in June and September, each lasting for three days.

The family history did not reveal any such abnormality on either side. Both parents were quite healthy. The mother, aged 25, had another child, a boy aged 5, who was normal.

On examination the patient was 44½ in. (113 cm.) tall and seemed older than her actual age. She was rather spare-built and was intelligent. She had genu valgum deformity, with a gap of about 5½ in. (13 cm.) between the heels. Internal rotation at the hips seemed slightly restricted on the right side. The spine was normal. Both breasts were fairly well developed, with brown pigmentation of nipple and areola on both sides. The nipples were quite developed and pointed. On compression, there was a white discharge from both nipples. The labia majora were well developed, the clitoris was enlarged, and a slight growth of hair could be detected. Flat brown pigmentation was widely distributed over the right arm, the right side of the chest, and the right buttock. The pigmentation was restricted to the right side (see Photograph). Other systems, N.A.D. Rectal examination did not reveal any abnormality.

Photograph showing pigmentation.

Blood examination showed: Hb 10.5 g./100 ml.; total white-cell count, 9,300/c.mm. (differential count not significant); and E.S.R., 15 mm. in first hour. Stool examination, N.A.D. Vaginal smear showed mostly uncornified epithelium of intermediate type and about 10% cornified epithelium. Basal cells and leucocytes were not seen. Urine examination: N.A.D.; serum calcium, 10 mg./100 ml.; alkaline phosphatase, 25 units/100 ml. (Bodansky); Sulkowicz test of urinary calcium estimation, within normal limits; blood urea, 30 mg./100 ml. 17-Ketosteroid excretion, 2.7 mg./24 hours.

On x-ray examination all bones appeared slightly radiolucent. The tubular bones, especially metacarpals, metatarsals, phalanges, tibia, fibula, radius, ulna, and femur of both sides, showed discrete areas with loss of bony texture. These areas had a homogeneous density slightly less than the density of normal bone. Multiple cystic changes were present in metacarpals, metatarsals, phalanges, and ulna, with slight expansion of the overlying cortex. Both femora showed coxa vara deformity with incomplete fracture at the neck. Pelvis showed deformity due to bending of both femora at the neck. Spine did not reveal any abnormality. Skull showed gross thickening of the base, especially of the roof of orbit, and of the sphenoid bone.

The bone age of the patient seemed to be about 12 years, as the secondary centre of ossification for trochlea, olecranon, and pisiform already showed, but the centre of ossification for external epicondyle of humerus and lesser trochanter of femur had not appeared. This suggested skeletal precocity.

Albright's syndrome was diagnosed because of polyostotic fibrous dysplasia; precocious development of sex organs with secondary sexual characteristics in a female patient; precocious skeletal development; and flat pigmentation of the right arm and right side of the chest and right buttock.

Differential Diagnosis.—Tumour of ovary (theca) leading to precocious sexual development and menstruation at the age of 5 years has been reported. It has also been reported that tumour of ovary has produced precocious development of both sex organs and secondary sex characteristics. Flat cutaneous pigmentation in a theca tumour is unknown. Bony deformity as seen in Albright's syndrome, but not so severe, has been reported.

Novak's Syndrome.—Bony deformity and pigmentation are absent in these cases. Sexual precocity is present.

COMMENT

Albright, Butler, Hampton, and Smith (1937) published five cases with multiple cystic areas in the bones, patches of pigmentation in the skin, and precocity.

The condition, in which precocity and pigmentation do not exist but bony changes are present, has been described by Lichtenstein (1938) as polyostotic fibrous dysplasia. Monostotic and polyostotic varieties, with or without pigmentation and with or without sexual precocity, have been grouped under the same heading as fibrous dysplasia of bone. A satisfactory explanation of the cause has not yet been given. The group that includes pigmentation and sexual and skeletal precocity suggests that the endocrine system may play some part in the causation. Falconer and Cope (1942) suggested that it is the result of a congenital disorder of development involving the skeleton and hypothalamus. Probably there is an increase in prolactin in this condition. There is some evidence suggesting that the sexual precocity is caused by lesions in the wall of the third ventricle and in the hypothalamus (*Textbook of X-ray Diagnosis*).

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REFERENCES

- A Textbook of X-ray Diagnosis* (1959). 3rd ed., vol. 4, edited by S. C. Shanks and P. Kerley. Lewis, London.
- Albright, F., Butler, A. M., Hampton, A. O., and Smith, P. (1937). *New Engl. J. Med.*, **216**, 727.
- Falconer, M. A., and Cope, C. L. (1942). *Quart. J. Med.*, **11**, 121.
- Lichtenstein, L. (1938). *Arch. Surg.*, **36**, 874.