Polyostotic Fibrous Dysplasia with Acromegaly
(Albright's Syndrome)

A Case Report

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SUMMARY

The association of polyostotic fibrous dysplasia and acromegaly, presenting with gross deformity of the skull and fractures of the left femur and right humerus, is described in a 42-year-old Black woman. Possible causative factors are discussed.


CASE REPORT

A 42-year-old Black woman was referred to this hospital with fractures of the left femur and right humerus, which she had sustained 9 months previously when she fell into a bath. Her complaint on admission was of backache and some pain in the left thigh.

The patient's menarche was at the age of 11 years, and she had menstruated irregularly for about 2 years. She had never fallen pregnant. At the age of 12 years she noticed a lump on her head which had occurred spontaneously and had slowly increased in size. The discomfort caused by her fall had cleared over some weeks of rest, and she did not consult a doctor.

On admission the patient was found to be in fairly good general condition. She weighed 90 kg and was just under 2 metres in height. There was a bony deformity on the right forehead laterally (Fig. 1), about the size of half a tennis ball. The prominence of her brows and zygomatic arches and the enlargement of her hands and feet and of her facial features were consistent with acromegaly. The right humerus appeared thickened throughout its length and there was an angular deformity of a considerably thickened left femur. In other respects the patient was essentially normal, with blood pressure 120/80 mmHg.

The patient had a mild normochromic normocytic anaemia, with normal blood urea and electrolyte concentrations. Her serum calcium and phosphorus levels were also normal. Alkaline phosphatase was 69 IU/litre. The fasting growth hormone level was 50 ng/ml, and was raised after ingestion of glucose.

Radiological examination of the skull, right humerus and left femur showed the features of polyostotic fibrous dysplasia (Fig. 2). In addition, acromegaly was thought to be present.

Fig. 1. See text.

DISCUSSION

Fibrous dysplasia is a condition of unknown aetiology in which the normal replacement of bone is taken over by fibrous tissue. It is monostotic when it occurs in one bone or polyostotic when it occurs in more than one bone. The occurrence of fibrous dysplasia with endocrine anomalies in the form of precocious puberty with café au lait areas on the skin was first described by Albright et al. in 1937. Subsequently the association of this condition with thyrotoxicosis was reported by Lichtenstein and Jaffe in 1942. Later, the occurrence of diabetes mellitus, hypercalcemia, basophil adenoma of the pituitary, and acromegaly with fibrous dysplasia were described. This association of fibrous
TABLE I. COMPARISON OF THE FEATURES OF ALBRIGHT'S SYNDROME AND MULTIPLE ENDOCRINE NEOPLASIA

<table>
<thead>
<tr>
<th>Albright's syndrome</th>
<th>Multiple endocrine neoplasia</th>
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<tbody>
<tr>
<td>Thyrotoxicosis</td>
<td>Thyrotoxicosis with or without goitre</td>
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<tr>
<td>Hypercalcuria</td>
<td>Parathyroid adenoma</td>
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<tr>
<td>Diabetes mellitus</td>
<td>Islet cell tumour of the pancreas</td>
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<td>Basophil adenoma of the pituitary</td>
<td>Chromophobe adenoma of the pituitary</td>
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<tr>
<td>Acromegaly</td>
<td>Phaeochromocytoma</td>
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<tr>
<td>Café au lait areas on the skin</td>
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</tbody>
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and which, while having no overt endocrine manifestations, is influenced by thyrocalcitonin.

Of interest here is the hypothesis of Pearse and Polak, of a peripheral neuro-endocrine system (APUD system), phylogenetically old and common to all vertebrates, the cells of which derive from the neural crest of the embryo. According to this theory these cells produce a variety of polypeptide hormones, which, together with hormones of the gastro-intestinal tract and lung and thyrocalcitonin, may be responsible for the endocrine conditions mentioned above. The oat-cell carcinoma of the lung is believed to be an undifferentiated argentaffin cell tumour, and its various ectopic hormones fulfil the criteria of the APUD series. A comparison of associations and disease patterns may show certain bone conditions such as fibrous dysplasia and Paget's disease to be involved in this hypothetical system, and may elucidate the causative factors in clubbing and pulmonary osteo-arthropathy.

REFERENCES