OSTEOSARCOMA IN ENCHONDROMATOSIS (OLLIER'S DISEASE)

Report of a Case

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Dyschondroplasia, or Ollier's disease, is best called enchondromatosis, and was defined by Jaffe (1958) as the presence of either circumscribed foci or large masses of cartilage in the interior of bones. It may be confined to limb bones and often almost to one side of the body, as in the patients described by Ollier in 1900.

The possibility of malignant change in a focus of enchondromatosis was considered to be rare by Fairbank (1948), whereas Jaffe (1958) found that chondrosarcomata developed in half his cases, and bore no relation to the severity of the enchondromatosis. Dahlin and Henderson (1956) and Dahlin (1957), although finding chondrosarcoma a common complication of multiple chondromata, stated that chondrosarcoma secondary to a proven benign enchondroma did not occur in their series. Murray and Cruickshank (1960) thought that malignancy did not occur unless the enchondromatosis was associated with cutaneous haemangiomata (Maffucci's syndrome). None of the above authors nor Hunter and Wiles (1935), Geschickter and Copeland (1949), Laurence and Franklin (1953), Cleveland and Fielding (1959), Lichtenstein (1959), Marjolis (1959) and Coley (1960) referred to the possible development of osteosarcoma.

The following case report describes an osteosarcoma developing in a man of sixty-eight who suffered from enchondromatosis.

CASE REPORT

The patient, a man aged sixty-eight, complained of an aching pain over the outer aspect of the left hip, present for three weeks. He said that a swelling had been excised from the right leg when he was sixteen years old, but apart from this there was no relevant history.

On examination he was in apparently good general health. There was left genu varum of 20 degrees and a bony swelling, thought to be an exostosis, was visible and palpable on the upper third of the left tibia. There was faint brown pigmentation on the outer aspect of the left thigh. There was tenderness over the left greater trochanter, and, although movement at the left hip was full, forced flexion caused pain in the groin. There was three-quarters of an inch of shortening in the left lower limb. The other systems were normal.

Radiographic examination—Unusual bony changes were present in the left femur, tibia, fibula and right fourth metatarsal: they took the form of areas of focal calcification, sometimes with alteration of the texture and outline of the surrounding bone (Fig. 1), and were interpreted as lesions of enchondromatosis. There was an area of increased density in the neck of the left femur above the lesser trochanter, and adjacent to it was a soft-tissue mass containing flecks of calcification (Fig. 2): this was interpreted as a malignant tumour, possibly a chondrosarcoma.

Arteriography—There was an abnormal plexus of fine vessels either in or sweeping round the soft-tissue mass (Fig. 3). The appearance was thought to be consistent with the development of a chondrosarcoma at this site, as suggested by plain radiography.

Biopsy—Biopsy of the left femoral neck was done. At operation the cortex was found to be partly replaced by soft tumour tissue which was invading the tissues surrounding the posteroinferior aspect of the neck.

Histological examination of the material confirmed that the lesion was a malignant tumour, but showed it to be an osteogenic sarcoma, not the expected chondrosarcoma. The
bulk of the tumour tissue had a pleomorphic spindle-celled structure (Figs. 4 and 5), but there were also conspicuous areas of tumour bone (Fig. 6), some of which was heavily calcified. Immersed in the tumour tissue were fragments of calcified cartilage and bone, presumably derived from the old areas of enchondromatosis. There were also several small nodules of more cellular cartilaginous tissue whose interpretation was not so clear: they appeared to be more cellular foci of enchondromatosis, rather than part of the malignant tumour.

Course—Three days after operation a pathological fracture occurred in the left femoral neck, and two days later he developed pneumonia. When his condition improved the upper half of the left femur was excised, with those muscles surrounding it that were involved, and it was replaced by a massive homologous bone graft.

On waking from the anaesthetic he had a hemiplegia and again developed pneumonia. Despite intensive treatment, he died two weeks after operation with fulminating pneumonia.

Necropsy findings—The lungs showed confluent bronchopneumonia in the lower lobes with widespread thrombosis of small pulmonary arteries. The air passages contained thin green pus. Numerous nodules of firm white tumour tissue were uniformly distributed in all lobes: they measured up to two centimetres in diameter. The brain was oedematous. The right frontal and parietal lobes were soft and swollen due to extensive haemorrhagic cortical infarction, which was also seen in the medial part of the right occipital lobe.

The upper half of the left femur, removed by an oblique saw cut in its midshaft, had been replaced by a similar human bone which had been fixed by a medullary nail. The original upper part of the left femur is shown in Figure 7.
The subcutaneous tissue over the left costal margin contained three vascular yellowish calcareous nodules up to 0.8 centimetre in diameter: a fourth nodule was present on the posterior wall of the fundus of the stomach. The coeliac and right para-aortic lymph nodes were moderately enlarged and of a spongy consistency.

**Histological examination**—The lung nodules had the structure of anaplastic spindle-celled sarcoma (Fig. 9). The calcareous nodules from the costal margin and the posterior wall of the stomach were shown to be cavernous haemangiomata, with large calcified foci due to old thrombosis. The para-aortic lymph nodes were replaced by lymphangiomatous tissue.
When the lower half of the femur and the tibia were sawn through, numerous small nodules of cartilage were seen in the medulla of both bones. Larger masses of cartilage were present, expanding and thinning the cortex of the upper part of the shaft of the tibia in the neighbourhood of the tuberosity (Fig. 8) and the lower part of the shaft of the femur.
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DISCUSSION

The main interest of this case is the development of an osteosarcoma in a focus of enchondromatosis in a man of sixty-eight. The incidence of malignant change appears to vary widely in different series, and those who accept it confine their remarks to chondrosarcoma. The occurrence of rapidly metastasising chondrosarcomas is mentioned by Jaffe (1958) but is usually confined to young adults.

The frequent occurrence of vascular anomalies in association with enchondromatosis was described by Carleton, Elkington, Greenfield and Robb-Smith (1942). Haemangiomata were seen in the subcutaneous tissues but not in the skin as in the Maffucci syndrome. Necropsy in this patient revealed several intercostal and retroperitoneal haemangiomata, together with angiomatic changes in the aortic lymph nodes.

The demonstration that the apparent "exostosis" of the tibia was actually an expansion of the overlying cortex by a central chondroma accords with the view that enchondromatosis is quite distinct from diaphysial aclasia (multiple exostoses).

SUMMARY

1. A patient of sixty-eight suffering from enchondromatosis (Ollier's disease) is described.
2. A malignant tumour developed in the region of the lesser trochanter of the femur. Histological investigation established that it was an osteosarcoma.
3. Other features of interest are the radiographic appearance of Ollier's disease at an advanced age, and the presence of subcutaneous subcostal and retroperitoneal haemangiomata.

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REFERENCES