CYSTIC DEGENERATION OF FIBROUS DYSPLASIA
MASQUERADING AS SARCOMA


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An area of fibrous dysplasia of bone may undergo rapid enlargement which may be due to either cystic degeneration or malignant transformation. These complications may be clinically and radiologically indistinguishable and, unless both are borne in mind, incorrect management may follow. Magnetic resonance imaging was used in one of our cases and was the only imaging modality to demonstrate the true nature of the condition.

Fibrous dysplasia of bone has been recognised as a separate entity since it was described by Lichtenstein in 1938. There are monostotic and polyostotic forms. Polyostotic fibrous dysplasia often follows an aggressive course and may be associated with skin pigmentation, endocrine abnormalities and precocious sexual development in Albright's syndrome (1937). The monostotic form is often thought to be innocuous, but certain complications are well recognised; these include recurrent fractures and progressive deformity.

Malignant change is a rare but well recognised complication in both the monostotic and polyostotic forms. Schwartz and Alpert (1964) reviewing the literature, found 28 cases of malignancy arising in fibrous dysplasia, 14 in monostotic cases. Eleven of the 28 patients had previously had radiotherapy for the fibrous dysplasia itself, and the malignancies included 18 osteosarcomas, seven fibrosarcomas, two chondrosarcomas and one giant cell sarcoma. They estimated the risk of malignant change for the monostotic form to be 0.4% and for an individual with the polyostotic form to be 4%.

We report three cases of fibrous dysplasia that presented with clinical features and plain radiographs suggestive of malignant change. All three cases were subsequently shown to have benign fibrous dysplasia complicated by cystic degeneration.

CASE REPORTS

Case 1. A 39-year-old man complained of pain in the lower right side of his chest and had a tender swelling of the ninth rib just in front of the anterior axillary line. His blood count, ESR and alkaline phosphatase level were normal. Radiographs showed an expanding lesion of the ninth rib 9 x 2.5 cm with poorly defined borders (Fig. 1). A chest radiograph taken nine months previously had shown the lesion to be under half this size with well defined borders. This lesion was active on bone scintigraphy.

Sarcomatous change was suspected and the ninth rib was excised (Fig. 2). The specimen contained an irregular expansion (3.5 x 1.5 x 12 cm) which included a cystic area (3 x 1.5 x 0.9 cm). Histological examination showed benign fibrous dysplasia with cystic degeneration.

Case 2. A 46-year-old woman with Albright's syndrome who was known to have lesions of the pelvis, many ribs and vertebral bodies, presented with gradual onset of weakness of her legs. Clinical examination revealed an incomplete paraparesis.

Her blood picture was normal except for a raised alkaline phosphatase of 207 SI units (normal <85). Radiographs showed marked disorganisation and erosion of the proximal ends of the left fourth to sixth ribs (Fig. 3), with a left pleural effusion. Tomograms showed an expanding lesion with indistinct borders; this was thought to be sarcomatous change in an area of fibrous dysplasia. Myelography revealed a partial block to the cranial flow of contrast at T6.

At a left thoracotomy the spinal cord was decompressed and a mass of abnormal bone was encountered replacing the left fourth, fifth and sixth ribs. A large cystic cavity, 5 x 6 cm, had excavated the fourth and fifth vertebral bodies leaving the disc intact. Histological examination showed benign fibrous dysplasia with cystic change.

Case 3. A 32-year-old housewife gave a three week history of aching behind her left knee. There was a firm swelling 4 x 6 cm attached to bone but not to skin, lying posterior to the distal third of the femur, with no increased warmth or abnormal vessels. Her knee was...
normal except that flexion was restricted by the mass to 95°. No other abnormalities were found on general examination.

Her blood count, ESR and alkaline phosphatase level were normal. Plain radiographs (Fig. 4) showed a poorly-defined lesion which had eroded through the posterior cortex of the distal femur and produced a periosteal reaction. There was also a soft-tissue mass in the popliteal fossa which contained streaks of calcification. On bone scintigraphy, the lesion was active and solitary.

In view of these findings a primary bone malignancy was suspected. Computerised tomography (CT) of chest and distal femur showed no evidence of pulmonary metastatic disease but revealed numerous fluid levels in the intramedullary component of the lesion (Fig. 5). Magnetic resonance imaging (MRI) confirmed the fluid levels but also showed, on T2 weighted images, that the entire lesion was surrounded by a low-level 'rind' (Fig. 6).

Biopsy was performed through an anterior window, bearing in mind the possibility of a later limb-sparing procedure. The specimen revealed fibrous dysplasia which had undergone extensive cystic degeneration. The lesion was subsequently excised, curetted and bone grafted through a posterior approach. Definitive histological examination confirmed the biopsy findings (Fig. 7).

**DISCUSSION**

Cases of rapid enlargement of fibrous dysplasia attributable to cystic enlargement have been described by Jaffe (1946) and by Schlesinger, Keats and Ruoff (1949). One of these cases had a rapidly enlarging rib lesion, similar to that in our Case 1, while the other had a lesion in the proximal tibia which was diagnosed as a sarcoma on the basis of rapidly increasing size, increased warmth of the tissues and radiological appearance. This latter patient had an amputation without biopsy because of what was considered to be overwhelming evidence of malignancy, but subsequent histology demonstrated that the lesion was benign.

The plain radiographic appearance in each of our cases suggested an aggressive pathology: malignant transformation of fibrous dysplasia in Cases 1 and 2 and a primary bone malignancy in Case 3. The CT and MRI
appearances of cystic degeneration of fibrous dysplasia have not to our knowledge been documented. In Case 3 fluid levels were identified on both CT and MRI, and there was a rind of low intensity signal on MRI. Fluid levels have been demonstrated on both CT and MRI in cases of aneurysmal bone cyst and telangiectactic osteosarcoma (Hudson 1984; Zimmer et al. 1984; Beltran et al. 1986). It would seem likely that the fluid levels seen in such cases represent layering of suspended solid material, such as clotted blood within cystic spaces. A low-level signal rind on MRI around a lesion has been reported as an indication of a benign process (Zimmer et al. 1985). This feature was present in our Case 3; MRI was the only imaging modality to indicate the benign nature of the lesion correctly.

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REFERENCES


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