ANEURYSMAL BONE CYST IN ASSOCIATION WITH FIBROUS DYSPLASIA

A CASE REPORT

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A patient with polyostotic fibrous dysplasia had several fractures of the right lower limb. An above-knee amputation was eventually performed, followed by arthrodesis of the hip. Five years later the stump became painful and swollen with dramatic rapidity. Biopsy showed that this was not due to malignant change, but that an aneurysmal bone cyst had developed in association with the fibrous dysplasia.

A 23-year-old female with a long history of fibrous dysplasia (see Table 1) was first seen at our hospital when she was four years old; at that time she had sustained a pathological fracture in the subtrochanteric region of her right femur. Radiographs of the limb suggested fibrous dysplasia; a skeletal survey was then made, revealing other lesions in the right ilium, the distal femur, the tibia and the fibula (Figs 1 to 3). She clearly had polyostotic fibrous dysplasia involving the right lower limb and hemi-pelvis (Figs 4 to 6).

Histological examination of a biopsy specimen confirmed the radiological diagnosis. The fracture, and all later fractures, were treated conservatively and healed within normal time-limits. Despite weight-relieving devices and bone-grafting operations, severe deformity resulted and the right leg became functionless. At the age of 19, her right leg was therefore amputated through the upper third of the femur (Fig. 7); subsequently an arthrodesis of the right hip joint was performed, with bone-grafting. After this the patient functioned well socially and she married.

Five years later, shortly after the fitting of a new prosthesis, she felt pain in the stump. Clinical examination revealed no abnormalities, and there were only minimal radiological changes in the proximal femur; these were thought to be of little significance (Fig. 8). Three weeks later, however, she presented with unbearable pain in the stump. It was now extremely painful on palpation and the bone was enlarged. The skin seemed normal, there were no local or systemic temperature changes, the ESR was 12 mm in the first hour and further blood analysis showed no abnormalities. A further radiological examination showed impressive changes (Fig. 9). Malignant degeneration was considered, but the speed of the changes, the very rapid expansion of the bone, a faintly visible cortical shell and the fluid level on the CT scan (inset, Fig. 9) suggested the development of an aneurysmal bone cyst.

An open biopsy was performed, revealing a cavernous lesion with a thin bone shell filled with blood. Histological investigation showed mesenchymal tissue with woven bone typical of fibrous dysplasia, but there were also cavernous spaces filled with blood and parts of cyst walls with haemosiderin deposits, as seen in aneurysmal bone cysts. No sign of malignancy was found.

Table 1. Chronology of fractures and operations

<table>
<thead>
<tr>
<th>Age</th>
<th>Fracture or operation</th>
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<tbody>
<tr>
<td>4</td>
<td>Fractured right femur</td>
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<tr>
<td>6</td>
<td>Curettage and bone graft</td>
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<tr>
<td>9</td>
<td>Valgus osteotomy right femur with bone graft Epiphysiodesis proximal end left tibia</td>
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<tr>
<td>12</td>
<td>Fractured right femur</td>
</tr>
<tr>
<td>18</td>
<td>Fractured right femur</td>
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<tr>
<td>18</td>
<td>Fractured right tibia</td>
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<tr>
<td>19</td>
<td>Fractured right ischium and pubis</td>
</tr>
<tr>
<td>19</td>
<td>Right above-knee amputation</td>
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<tr>
<td>20</td>
<td>Resection of right femoral head and neck, and arthrodesis with bone grafting</td>
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</tbody>
</table>

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Figure 7—The patient's femur one month after amputation (age 19 years). Figure 8—Five years later only relatively minor changes are seen. Figure 9—The femur three weeks later: blow-out expansion of bone is clearly seen with only faintly visible cortical margins. A CT scan (inset) of the stump area shows a fluid level consistent with liquid blood within the bone. Figure 10—The femur three months after an open biopsy: the cortical margins are outlined more clearly and the lesion looks quiescent, with the septa and ridges more regularly distributed.

After biopsy the pain disappeared and two months later there were radiological signs of reappearance of the old structure (Fig. 10).

DISCUSSION

In this patient, rapid expansion of the bone in a fibrous dysplastic lesion at first suggested malignant change. Such change occurs in 4% to 10% of cases of fibrous dysplasia (Dabska and Buraczewski 1972; Huvos, Higinbotham and Miller 1972; Bejui-Thivolet, Patriicot and Vauzelle 1982). However, the radiological evidence of extremely rapid disappearance of bone structure in less than three weeks, the apparent “blow-out” expansion of bone and the appearance of a fluid level on the CT scan all suggested a diagnosis of secondary aneurysmal bone cyst formation. Fluid levels have been described by Hudson (1984) in six out of 17 cases of aneurysmal bone cyst.

At biopsy, the presence of aneurysmal bone cyst-like structures in active fibrous dysplastic tissue was confirmed. Nine instances of aneurysmal bone cysts in cases of fibrous dysplasia have been previously reported: these included four in the jaws, one in a rib, one in the tibia and three not specified (Jaffe 1962; Clough and Price 1968; Buraczewski and Dabska 1971; Oliver 1973; Bonakdarpour, Levy and Aegerter 1978; El Deeb, Sedano and Waite 1980). To our knowledge this is the first report of a bone cyst arising in the femur.

At first sight this case seems to support the widely-accepted “precursor” theory of the aetiology of aneurysmal bone cysts (Jaffe 1950; Lichtenstein 1950, 1953;
Donaldson 1962); this theory states that haemodynamic changes take place in a pre-existing lesion, and these contribute to the formation of arteriovenous fistulae; bone expansion then follows from the raised intra-osseous vascular pressure. However, Ruiter, van Rijssel and van der Velde (1977) stated that the presence or absence of precursor lesions seems related to histodiagnostic selection and depends on how lesions with areas resembling aneurysmal bone cysts are classified. In this light it is interesting to note that, in our patient, a subsequent review of the biopsies taken at earlier operations revealed aneurysmal bone cyst-like structures in multiple sites. Our case thus seems to be one of active fibrous dysplasia in which haemodynamic changes gave rise to an expansive development of already existing aneurysmal bone cyst-like structures; this, rather than an aneurysmal bone cyst arising from a pre-existing lesion, seems the likely sequence of events.

REFERENCES


