DESMOPLASTIC FIBROMA OF BONE
A REPORT OF SIX CASES

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The clinical and pathological features of six cases of desmoplastic fibroma of bone are presented. Desmoplastic fibroma is rarely seen as a primary tumour of bone; when it does occur the sites of predilection are the long bones, but other sites such as the scapula and os calcis can be involved. Radiographically the lesion tends to expand the bone from within; it is well-demarcated and lytic, often with a trabeculated soap-bubble appearance. The cellular structure and the morphological arrangement are similar to those of aggressive fibromatosis of soft tissues.

Differential diagnosis from malignant spindle-cell lesions of bone is important because the treatment of choice for desmoplastic fibroma of bone is simply excision with a thin layer of healthy tissue.

Only about 60 cases of this rare tumour have been reported (Jaffe 1958; Rabhan and Rosai 1968; Nilsonne and Göthlin 1969; Dahlin 1978; Taguchi and Kaneda 1980). The purpose of this paper is to report an additional six cases.

MATERIALS AND METHODS

Only those primary intra-osseous lesions which were composed entirely of differentiated fibroblasts and collagen have been studied. We excluded similar lesions which appeared to originate in the parosteal soft tissues and secondarily invaded the bone, and those originating at the surface of the bone. We also excluded fibroblastic tumours with some cellular atypia which were considered to be well-differentiated, low-grade fibrosarcoma.

Clinical findings. The clinical details of the patients and the tumours are shown in Table I. They are consistent with those in previous reports.

In one case the tumour was located in the mid-shaft, which is distinctly unusual, as is the location in the fibula. The symptoms (swelling of the bone sometimes accompanied by pain), were those of a slow-growing, indolent lesion; often these symptoms had been present for long periods of time, over two years in three of the patients. In none of our patients was a pathological fracture seen.

Radiographic findings. In two patients the tumour, when diagnosed, had reached an enormous size (Fig. 1), involving a very large portion of the bone and markedly expanding the shaft. The following features were common to all our patients: an intra-osseous well-defined radiolucent lesion; expansion of the bone involving the whole circumference or only part of it (Figs 2 to 4); marked thinning of the cortex, which was regular and smooth, without any sign of recent periosteal reaction; a

Fig. 1
Case 1. Radiograph showing a large area of radiolucency with a coarse trabeculated soap-bubble appearance in the distal part of the femur. The cortex is thinned.

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Table I. Clinical findings in six cases of desmoplastic fibroma

<table>
<thead>
<tr>
<th>Cases</th>
<th>Sex</th>
<th>Age (years)</th>
<th>Location and size</th>
<th>Symptoms</th>
<th>Treatment</th>
<th>Complications</th>
<th>Follow-up (from diagnosis)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>24</td>
<td>Distal femur (8 x 15 x 9 cm)</td>
<td>Pain and swelling for 3 years</td>
<td>Wide excision</td>
<td>Infection; amputation 6 months later</td>
<td>No recurrence after 35 years</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>43</td>
<td>Scapula</td>
<td>Swelling for some months</td>
<td>Intrallesional excision</td>
<td>—</td>
<td>No recurrence after 23 years</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>32</td>
<td>Proximal fibula (5 x 17 x 6 5 cm)</td>
<td>Swelling for some years</td>
<td>Wide excision</td>
<td>Soft-tissue infection treated conservatively</td>
<td>No recurrence after 31 years</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>36</td>
<td>Calcaneus (2.5 x 4 x 6 cm)</td>
<td>Pain and swelling for 2 months</td>
<td>Intrallesional excision</td>
<td>—</td>
<td>No recurrence after 3 years</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>26</td>
<td>Distal fibula (4 x 10 x 3 cm)</td>
<td>Pain for 2 months swelling for 3 years</td>
<td>Wide excision</td>
<td>—</td>
<td>No recurrence after 1 1/2 years</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>17</td>
<td>Humerus (diaphysis) (5 x 5 x 3 cm)</td>
<td>Intermittent pain</td>
<td>Wide excision</td>
<td>—</td>
<td>No recurrence after 1 year</td>
</tr>
</tbody>
</table>

sclerotic margin (Figs 3, 4 and 5); and a delicate and diffuse trabeculation, giving a soap-bubble appearance (Figs 1 and 2). These features indicate a slowly growing tumour and should make one suspect the diagnosis pre-operatively.

Computerised axial tomography before operation can be useful in assessing the continuity of the cortex and the extent of invasion of the soft tissues.

**Macroscopic features.** The periosteum was intact but was usually thinner than normal. The lesions were a pale tan colour or whitish grey; they were solid, rubbery and tough in consistency. The cut surface sometimes had a fasciculated appearance. The margins of the lesion were rounded and blunt, the so-called pushing appearance.

**Microscopic features.** The lesions were composed of interlacing fascicles of mature, moderately cellular fibrous tissue (Fig. 6). In some cases the cells were sparse: a few spindle-shaped, slender cells were embedded in coarse strands of collagen (Fig. 7). Mitosis was very rare. The nuclei were dense with no nucleoli. Some cases showed more cellularity, the cells being generally flat and slender but sometimes plump and larger with finely dispersed nuclear chromatin and small round nucleoli; these cells were associated with a large amount of intercellular collagen fibres in bundles. In these instances also, mitosis was very rare (Figs 8 and 9). Pleomorphism, hyperchromatism and atypia were not features of our cases. Cells and collagen fibres were arranged in parallel fashion and in bundles; different bundles crossed each other, but the "herring bone" pattern of fibrosarcoma was not evident. Sometimes at the edges of the lesion one was able to see slight permeation of the bone trabeculae. In sections 0.5 μm thick, stained with toluidine blue and embedded in Epon, it was possible to see spindle cells with prominent nucleoli and processes intertwined with bundles of collagen (Fig. 10).

In one case electron microscopy was performed. Spindle cells were oriented side by side with bundles of collagen fibres that were sometimes seen in transverse section. Some cells showed rough endoplasmic reticulum.
Figure 6—Case 5. Prominent fascicles of dense collagenous tissue intermixed with spindle cells. The appearance is similar to that of a desmoid tumour of soft-tissue. (Haematoxylin and eosin, × 85.) Figure 7—Case 3. A few elongated spindle cells with inconspicuous nuclei are embedded in coarse strands of collagen. (Haematoxylin and eosin, × 85.) Figure 8—Case 5. The cells are sparsely separated from one another by abundant collagen. (Haematoxylin and eosin, × 210.) Figure 9—Case 1. There is more cellularity. As in Figure 8 the nuclei may be small and elongated, or large and plump. Mitoses are absent. (Haematoxylin and eosin, × 210.)

with a cisternal type of dilatation or with parallel anastomosing channels. Besides these fibroblastic features, occasionally filaments and dense bodies (Fig. 11) were identified at the periphery of the cell, associated with a well-developed Golgi apparatus.

TREATMENT AND RESULTS
The diagnosis can be ascertained with a frozen-section biopsy. This, supported by the gross appearance found at operation, the radiographic features and the clinical findings, should be sufficient to confirm the diagnosis so that definitive treatment can be undertaken in the same operative session as the biopsy.

Two of our cases (one in the scapula and one in the calcaneus) were treated by thorough curettage, which we called "intralesional excision". The cavity in the calcaneus was subsequently filled with autogenous cortical grafts.

The remaining four cases were treated by a wide segmental resection. Of these four tumours, the one in the proximal fibula was treated by resection and nothing more. The tumour in the distal femur required an...
endoprosthesis, but subsequently the limb was amputated because of infection. The tumour of the mid-shaft of the humerus was treated with a plate and an autogenous cortical graft. The tumour in the distal fibula required an autogenous cortical graft.

In no case was local recurrence observed during the follow-up, but in three cases this was less than three years.

DISCUSSION

Radiographically, desmoplastic fibroma can mimic haemangiomma because of its trabeculation. Haemangiomma is more painful and does not usually reach the size of a large and expanding desmoplastic fibroma. Other lesions which must be considered in the differential diagnosis are aneurysmal bone cyst, unicameral bone cyst, fibrous dysplasia, chondromyxoid fibroma and non-ossifying fibroma. As can be inferred from this differential diagnosis, the radiographic appearance is usually that of a benign lesion. Only rarely have primary malignant lesions such as adamantinoma, fibrosarcoma or metastatic carcinoma been suspected (Whitesides and Ackerman 1960; Nilsonne and Göthlin 1969).

Microscopically, the lesion is identical to the well-known desmoid tumour of the abdominal wall, a commoner aggressive growth of muscles and aponeuroses; it is also similar to the so-called "periosteal desmoid", a very small asymptomatic and innocuous defect found subperiosteally in the metaphysis, particularly of the distal femur of children and adolescents. The microscopic pattern of desmoplastic fibroma may also be mimicked by fibrous dysplasia, but only in areas where there is only fibrous tissue and the typical woven bone is lacking; adequate sampling eliminates such histological error.

The criteria for differentiation from a well-differentiated, low-grade fibrosarcoma have been described by Jaffe (1958); those tumours containing small fibroblasts with rather inconspicuous nuclei were called desmoplastic fibromata while those with relatively large and plump nuclei were considered to be well-differentiated fibrosarcomata. Certainly, on the basis of the microscopic details, it is possible to draw a presumptive line dividing what we call desmoplastic fibroma from what we call Grade I fibrosarcoma and this distinction can, in theory at least, be reasonably precise. Sometimes, however, the differentiation between the two is delicate and subjective, and there is then considerable possibility of error.

Electron microscopy studies (Lagacé et al. 1979; Goellner and Soule 1980) have led some workers to speculate that the greater number of myofibroblasts in desmoid fibromatosis (than in fibrosarcomata) may be a feature of their better prognosis (Stiller and Katenkamp 1975). It seems wise, however, to study more cases before attaching prognostic value to the number of these cells.

In our opinion, it would therefore be more honest and practical for therapeutic purposes to consider desmoplastic fibroma and Grade I fibrosarcoma under the same heading, that is as well-differentiated fibrous tumours having very slow but progressive growth and recurring locally when incompletely removed. The distinction between the two is relative and tentative rather than strict and definitive. The microscopic features which distinguish Grade I fibrosarcoma are the herringbone pattern, the richer cellularity, plumper nuclei, slight pleomorphism and hyperchromasia, and the mitotic activity which is greater than in desmoplastic fibroma.

The histological differential diagnosis may also include central low-grade osteosarcoma, but this is characterised by the production of osteoid and bone. In the jaw, the differentiation from a central fibroma can be difficult (Hinds, Kent and Fechner 1969).

It is interesting to note that in 12 recurrent cases of desmoplastic fibroma reported in the literature, the histology of the recurrence was the same as that of the original tumour (Sugiura 1976); the same is true of desmoid tumours of the abdominal wall. Dahlin (1978), however, has described a desmoplastic fibroma which increased markedly in grade with recurrence.

The treatment of desmoplastic fibroma is exclusively surgical; excision should be moderately wide. By that we mean an excision en bloc and complete, with a continuous, although thin, layer of healthy tissue around the tumour.

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REFERENCES


