MONOSTOTIC FIBROUS DYSPLASIA

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The term fibrous dysplasia of bone was first used by Lichtenstein in 1938 to describe a condition to which attention had been drawn by Hunter and Turnbull (1931). They stated: "Of much more common occurrence than the generalised disease is focal osteitis fibrosa. This is a condition affecting one or more bones; usually not disabling; of slow progress; and showing a tendency to become arrested. It occurs chiefly in adolescence, and is often symptomless until spontaneous fracture occurs . . . The figures for serum calcium and phosphorus are invariably normal." This is a description of fibrous dysplasia which has since hardly been improved.

The etiology of fibrous dysplasia is unknown, but it is now believed to be a developmental error in which primitive fibrous tissue proliferates within the bony medulla and encroaches upon the cortex from within, often producing expansion. The process may be localized to a small segment or may involve almost the entire shaft of the bone, and it may be monostotic, monomelic or polyostotic. In the latter form some patients exhibit a remarkable combination of skin pigmentation and endocrine disturbances. These cases, being dramatic, have predominated in the literature to give a false impression of their relative frequency. The single bone lesions unassociated with any of these other disturbances are much the most common form of fibrous dysplasia (Jaffe 1958).

This paper is a study of fifty patients with monostotic fibrous dysplasia, forty-six recorded in the Bristol bone tumour register and four from the Princess Elizabeth Orthopaedic Hospital at Exeter. The diagnosis in each instance was made after full clinical, radiological and histological findings.

**SITE**

Skeletal surveys were not carried out in all patients, so that there is no proof that the lesion recorded was the only one present. However, in no instance has a second lesion later come to light. The long bones were those most often affected, the femur and tibia being involved in equal numbers (Fig. 1). (In most previous reports the femur has been quoted as the most common site.) In none of this group of patients was the disease diagnosed in the scapula, vertebra or calvarium, though of course these areas are not exempt from this disease.

**AGE DISTRIBUTION**

As can be seen from the histogram (Fig. 2) the peak incidence was at ten to fifteen years of age when all sites are taken together. Fibrous dysplasia is thus primarily a disease of growing bone. If, however, the age distribution is considered in relation to site (excluding...
the tarsus, which was involved in two patients, and the pelvis in one) it is found that lesions in the long bones mostly appear in childhood (Fig. 3). In the jaws the disease seems to appear in the early twenties (Fig. 4). The condition here is usually obvious, so that this time of onset is probably true. Fibrous dysplasia in the rib does not give rise to urgent symptoms; in fact, it may be symptomless: this explains the late age of presentation of lesions at this site (Fig. 5).

**SEX DISTRIBUTION**

The cases in this series were almost equally distributed between the sexes (twenty-seven males: twenty-three females). This is in accordance with the findings of Harris, Dudley and Barry (1962). Previous observers have found a marked preponderance of females: Lichtenstein (1938) stated that in his material, and in that of others, the female/male ratio was at least 3:1.

**AGE/SEX RELATIONSHIP**

The number of patients is insufficient to allow statistical conclusions, but it seems likely that in the long bones there is no age difference between the time of onset of symptoms in either sex (Table 1). It might have been expected that the earlier bone maturity of girls would have caused their lesions to have presented before those in boys.
COURSE

It is relevant to the problem of treatment to note some factors in the progress of monostotic fibrous dysplasia as it affected this group of patients. It is generally believed that these lesions cease to be active with the onset of puberty. This may be true, but there were several examples in this series of patients presenting unusually late (Fig. 2). Several of these had had pathological fractures of a long bone (Fig. 6). Why should this happen at this late stage in patients who have presumably led a normal, active life?

Pregnancy appears sometimes to stimulate activity in the dysplastic tissue. Two patients suggested this. One was a woman of thirty, who was found on routine radiographs of the chest to have an expanded third left rib (Fig. 7). A further radiograph taken three years later,

TABLE 1

<table>
<thead>
<tr>
<th>Site</th>
<th>Sex</th>
<th>Number</th>
<th>Mean age (years)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Long bones</td>
<td>Males</td>
<td>16</td>
<td>13</td>
</tr>
<tr>
<td></td>
<td>Females</td>
<td>11</td>
<td>13.3</td>
</tr>
<tr>
<td>Ribs</td>
<td>Males</td>
<td>6</td>
<td>39.5</td>
</tr>
<tr>
<td></td>
<td>Females</td>
<td>2</td>
<td>26</td>
</tr>
<tr>
<td>Jaws</td>
<td>Males</td>
<td>5</td>
<td>16</td>
</tr>
<tr>
<td></td>
<td>Females</td>
<td>7</td>
<td>21</td>
</tr>
</tbody>
</table>

FIG. 6

Fibrous dysplasia with pathological fracture in the upper radius of labourer of 44 years.

three months after delivery, showed quite marked enlargement of the lesion (Fig. 8). The rib was excised, and showed typical fibrous dysplasia. The other patient was a woman who first complained of pain in the hip in the early months of her first pregnancy, when she was twenty-one. She had not suffered symptoms before. Fibrous dysplasia of the proximal right femur was treated by operation some time after delivery (Fig. 9).
Reactivation during pregnancy of previously diagnosed fibrous dysplasia has been reported on at least three occasions (Hunter and Turnbull 1931; Dockerty, Ghormley, Kennedy and Pugh 1945; Bonduelle and Claisse 1948).

No patient in this series suffered delayed union or non-union after fracture through a dysplastic area.

Excluding jaw lesions, those treated by the excision of an entire bone and one complicated by malignancy, radiological progression with or without treatment was observed in six patients with an average age of eleven years at presentation. Twelve patients showed no evidence of continued activity, the average age of this group at presentation being fifteen years. It is probable that lesions that come to light at an early age are those most likely to progress.

CLINICAL PRESENTATION

These patients sought advice on account of pain, swelling or pathological fracture (Table II). In three the disease was an incidental finding. In the long bones pathological fracture was the most frequent presentation. Pain with or without swelling is common as an initial symptom, but its basis is almost invariably an incomplete fracture which can be seen in a good radiograph (Fig. 10). This opinion is supported by the fact that pain is not so often present in rib lesions. This tendency to fracture in the long bones may be a factor in bringing the lesions to light at an earlier age in this site. The two tarsal lesions and the pelvic lesions
all presented with pain. Probably many patients with monostotic fibrous dysplasia do not have symptoms.

When a lesion is found, biopsy is required for accurate diagnosis. In the polyostotic form a diagnosis may often be made from the radiographs alone, but this is not so in monostotic fibrous dysplasia.

### TABLE II

**Presenting Symptoms in Forty-Seven Patients**

(Tarsus and Pelvis Excluded)

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Long bones</th>
<th>Ribs</th>
<th>Jaws</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain and swelling</td>
<td>4</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>Painless swelling</td>
<td>2</td>
<td>4</td>
<td>6</td>
</tr>
<tr>
<td>Pain alone</td>
<td>8</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Pathological fracture</td>
<td>12</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Incidental</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>27</strong></td>
<td><strong>8</strong></td>
<td><strong>12</strong></td>
</tr>
</tbody>
</table>

**TREATMENT**

In those patients with symptoms, surgery is required to remove abnormal tissue, to fix a fracture internally or to correct deformity. In the rib or fibula the involved segment of bone may be excised. At other sites, excluding the jaws, three types of surgical procedure have been carried out in this series: 1) curettage and packing with cancellous chip grafts; 2) subperiosteal excision and cancellous block graft; and 3) extraperiosteal excision and cancellous/cortical graft. The results in twenty-eight patients followed up for from two years to twenty-six years with an average of twelve years, are shown in Table III. Treatment was successful when
there was radiological evidence that the dysplastic tissue had been eradicated and replaced by viable bone, and when the patient remained symptom-free. "Failure" was denoted by persistence in whole or in part, or even radiological expansion of the lesion often with absorption of bone grafts. This may or may not have been accompanied by continuing or recurrent symptoms. It will be seen from Table III that the first two methods, curettage and subperiosteal excision, are not by any means always successful.

Fibrous dysplasia of the tibia (the late Mr John Bastow's patient). Figure 12—Initial radiographs. Figure 13—After subperiosteal excision and grafting. Figure 14—Two years later: recurrence of lesion. Figure 15—Satisfactory healing four years after extraperiosteal excision and cancellous cortical graft.
A probable cause of failure in many of these operations is that not all the abnormal tissue is excised. What remains is in an active phase which absorbs and replaces the cancellous grafts. It is sometimes extremely difficult to remove all this tissue. This is especially so when the medullary cavity has been replaced and the cortex eroded right out to subperiosteal level (Fig. 11). In this situation curettage or subperiosteal excision must inevitably leave behind abnormal material. If this is in an active state it will continue to grow and lead to the absorption of any implanted grafts. Therefore, in this particular situation complete removal of all dysplastic tissue means excising periosteum as well. This problem is well illustrated in Figures 12 to 15.

**SUMMARY**

1. Monostotic fibrous dysplasia in the long bones occurs most frequently in adolescence. In the jaws it is found mainly in early adult life. It presents later in the ribs, probably because it is often asymptomatic in this site.
2. The disease is equally distributed in both sexes.
3. Reactivation may occasionally occur in later life and in pregnancy.
4. Successful surgical treatment is by no means always easily achieved, and requires, in addition to the problems of fracture fixation and the correction of deformity, careful consideration of the age of the patient, the activity of the lesion and the extent to which it involves the cortical bone.

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**REFERENCES**


