CHONDROSARCOMA IN FIBROUS DYSPLASIA OF THE PELVIS
A CASE REPORT AND REVIEW OF THE LITERATURE

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Malignant transformation of fibrous dysplasia to chondrosarcoma is rare. We report a case in which malignancy developed in an area of fibrous dysplasia in the ilium. We believe this to be the second reported case at this site. Treatment was by excision of the hemipelvis including the ala of the sacrum. A review of the literature is presented.

Malignant change in fibrous dysplasia is rare. It has been estimated that it occurs in less than 1% of cases (Immenkamp 1975). The most common malignant tumour is osteosarcoma followed by fibrosarcoma and then chondrosarcoma. We report a case of chondrosarcoma arising in an area of fibrous dysplasia in the left iliac bone. The only other lesion of fibrous dysplasia in this patient was in the upper end of the left femur. Treatment was by partial resection of the left innominate bone and part of the left ala of the sacrum.

CASE REPORT

A 10-year-old boy presented in 1958 with pain in the left knee. The pain seemed to be referred from the hip, and radiographs showed fibrous dysplasia involving the left iliac bone and the proximal metaphysis of the left femur. This diagnosis was confirmed by biopsy of the femoral lesion (Fig. 1), which was bone grafted three weeks later.

Two years later the patient fell at school, sustaining a fracture of the shaft of the femur at the lower limit of the lesion. This fracture was treated by Kuntscher nailing, and six months later more bone graft was placed around the nail. The nail was removed two years after this and replaced by a Capener nail-plate. The femoral lesion eventually consolidated (Fig. 2), though with coxa vara and 1½ inches of shortening. Review was continued until 1971, when the 23-year-old patient was discharged from follow-up. Review of the radiographs from 1958 to 1971 revealed no significant change in the appearance of the fibrous dysplasia in the ilium.

In 1983, at the age of 35 years, the patient again developed pain in the left knee, and his general practitioner discovered a pelvic mass. This mass was huge but painless, occupying the iliac fossa, the gluteal region and the loin on the left side. It was hard, not tender, and fixed to the pelvis. The skin over the mass was mobile and showed a few dilated veins. The femoral artery could not be felt in the groin because of the encroaching tumour. There was no gap between the costal margin and the tumour mass.

The left hip joint had painfree flexion to 110° but abduction was limited to 10°. The left knee had a full painless range of movement. There were no abnormal neurological signs in the leg, and he had full power of the muscles of the left knee, ankle and foot. The dorsalis pedis and the posterior tibial pulses were present. A radiograph is shown in Figure 3; a chest radiograph was clear.

CT scans (Figs 4 to 10) showed that the lesion did not involve the floor of the acetabulum, but that the left ala of the sacrum was affected; this was thought to be involvement by fibrous dysplasia and not by tumour

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0301–620X/84/5133 $2.00

Fig. 1
Micrograph of the original biopsy of the upper left femur in 1958. The pathologist’s report stated: “The lesion is a loose, partly fibrous and partly vascular mass and the vessels are mainly lymphatic. In one fragment of tissue there is osteoblastic activity of the kind seen in fibrous dysplasia. There is one small island of cartilage, but it could very well be reactive. It is an uncommon but well-recognised variant of fibrous dysplasia for cartilage to be present.”
Figure 2—Fibrous dysplasia of the ilium and the upper femur. The femur has consolidated after biopsy, nailing and bone grafting.

Figure 3—Chondrosarcomatous change of the lesion in the ilium.

CT scans showing the extent of the lesion, the lack of involvement of the acetabulum and the changes in the ala of the sacrum.
tissue. The diagnosis of chondrosarcoma was based on the characteristic radiographic appearance, and it was decided to perform a left hemipelvectomy, including the left ala of the sacrum, but preserving the left lower limb.

**Operation.** The patient was placed on his right side on a table which was broken as for a kidney operation to create a gap between the tumour mass and the costal margin. An inverted U-incision, started upwards from the mid-inguinal point, curved above the swelling, then back to the paraspinal muscles and ended over the back of the sacrum. The tumour was then cleared on all its aspects except the medial one.

The sciatic nerve was exposed and the short rotator and abductor muscles cut to allow dislocation of the hip. The obturator foramen and the obturator nerve were exposed below the acetabulum and the superior pubic ramus was divided with a saw. The ischium could then be divided at the lesser sciatic notch, taking care to avoid injury to the pudendal nerve and vessels.

The dissection of the nerve roots from within the sacrum was difficult. The posterior sacral foramina were exposed and the bars between the foramina were excised using rongeurs. The superior surface of the ala of the sacrum was divided while protecting the lumbosacral nerve trunk. With the anterior primary rami of the sacral nerves exposed, the transverse bars between the sacral foramina were divided anteriorly and the mass excised *in toto*. Biopsy specimens from the remaining sacrum were taken for histological examination, though macroscopically it was clear that the tumour did not involve this bone. At the end of the operation the femoral nerve, the sciatic nerve, the lateral cutaneous nerve of the thigh, the obturator nerve and the femoral vessels were seen to be intact, but it was discovered that the pudendal nerve had been divided.

The removal of the mass left a large defect in relation to the abdominal wall, particularly after excision of the gluteal muscles. The defects were partially closed by suturing the abdominal muscles to the paraspinal muscles and to the adductors and flexors of the hip. The hip was found to be completely flail, but the head of the femur could not be pushed above the level of the sacrum.

After operation the leg was supported by pillows. A lateral popliteal nerve paresis eventually recovered and the wound healed satisfactorily after some delay. After six weeks of bed rest, the patient sat out and had good knee control. About two months after operation he stood and began to put some weight on the leg, using his original shoe-raise and a pair of crutches. At first the leg was flail but the hip gradually became more stable and could eventually take considerable weight. Walking with crutches is now satisfactory. The postoperative radiograph shows the extent of the resection (Fig. 11). Histology of the tumour (Fig. 12) was reported to show a low-grade chondrosarcoma.

**DISCUSSION**

Malignant transformation in fibrous dysplasia is rare; only 69 cases have been reported. This total includes 37 cases of osteosarcoma, 21 of fibrosarcoma and 11 of chondrosarcoma. The references for these cases are listed in Table I. In these collected cases the average age at the diagnosis of malignant change to osteosarcoma was 33 years, to fibrosarcoma 37 years and to chondrosarcoma 30 years. There was no sex predominance, with 33 men and 36 women. The sites at which malignant change occurred are listed in Table II.

Of the reported cases, 38 occurred in patients with polyostotic fibrous dysplasia and 29 in monostotic disease, while the status of the other two cases is not clear from the references. Twenty-one patients had received radiotherapy as treatment for fibrous dysplasia.

In the group developing *osteosarcoma* 24 of the 37 patients had polyostotic fibrous dysplasia; 15 had received radiotherapy, 13 of whom had polyostotic disease. These figures suggest an increased probability for malignant transformation to osteosarcoma after the treatment of polyostotic fibrous dysplasia by radiation.
Table I. Cases listed in the literature

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<thead>
<tr>
<th>Osteosarcoma</th>
<th>Fibrosarcoma</th>
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<tr>
<td>Abelanet et al. 1974</td>
<td>Anastavos et al. 1977</td>
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<td>Balcells-Gorina et al. 1963</td>
<td>Bell and Hinds 1967</td>
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<td>Belloni and Zanetti 1949</td>
<td>De Marchi 1956</td>
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<td>Coley and Stewart 1945</td>
<td>Euler 1956</td>
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<td>Dabska and Buraczewski 1972</td>
<td>Gimes et al. 1970</td>
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<td>Dustin and Ley 1950</td>
<td>Gross and Montgomery 1967</td>
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<td>Harris et al. 1962</td>
<td>Hall et al. 1955</td>
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<td>Huvos et al. 1972</td>
<td>Harris et al. 1962</td>
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<td>Jäger 1962</td>
<td>Hobbs et al. 1956</td>
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<td>Kiehn et al. 1961</td>
<td>Immenkamp 1975</td>
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<td>Perkinson and Higinbotham 1955</td>
<td>Johnson et al. 1979</td>
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<td>Pons et al. 1974</td>
<td>Mogensen 1958</td>
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<td>Pracke et al. 1968</td>
<td>Pons et al. 1974</td>
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<td>Riddell 1964</td>
<td>Schwartz and Alpert 1964</td>
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<td>Rozet et al. 1967</td>
<td>Tanner et al. 1961</td>
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<td>Sabanas et al. 1956</td>
<td>Turbinkov and Skoblin 1958</td>
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<td>Schwartz and Alpert 1964</td>
<td>Wanke 1927</td>
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<td>Sethi et al. 1962</td>
<td>Chondrosarcoma</td>
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<td>Slow 1971</td>
<td>Anastavos et al. 1977</td>
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<td>Sutro 1951</td>
<td>Beiji-Thiioleta et al. 1982</td>
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<td>Tanne et al. 1961</td>
<td>Cabitza 1951</td>
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<td>Vakhurkima 1958</td>
<td>Dabska and Buraczewski 1972</td>
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<td>Van Horn et al. 1963</td>
<td>De Smet et al. 1981</td>
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<td>Yannopulos et al. 1964</td>
<td>Feintuch 1973</td>
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<td>Hellen 1953</td>
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<td>Huvos et al. 1972</td>
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<td>Jaffe 1958</td>
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<td>Unni and Dahlin 1979</td>
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The group developing fibrosarcoma included 21 patients, 12 with monostotic fibrous dysplasia, 8 with polyostotic disease and one of unreported type. Only five patients had received radiotherapy and three of these originally had monostotic involvement.

Of 11 patients in the chondrosarcoma group, six had polyostotic disease, four had monostotic involvement and one type was not reported. Only one patient in this group had received radiotherapy (Feintuch 1973). The bones reported to be involved by chondrosarcomatous transformation were the femur in four cases, the pelvis in four (ilium two, ischium one and pubis one), and one case each involving the humerus, the maxilla, and the metatarsus.

Unni and Dahlin (1979) and Pelzmann, Nagel and Salyer (1980) reported that there may be large amounts of atypical cartilage in areas of fibrous dysplasia without definite chondrosarcomatous change. This could well lead to a mistaken diagnosis of malignancy. Neoplastic change in areas of fibrous dysplasia has the same prognosis as the same malignancy arising elsewhere, and should have the same treatment.

Partial or complete resection of the hemipelvis is a less mutilating procedure than hindquarter amputation, avoiding the problems of a hindquarter prosthesis and the psychological trauma of losing one quarter of the body. Enneking (1966) and Enneking and Dunham (1978) described partial and complete resections of the innominate bone for malignancy; they believe that involvement of the sciatic nerve is not a contra-indication, since the resection can include this nerve.

Steel (1978) reported five such resections on patients with chondrosarcoma involving different parts of the innominate bone. None of his patients had local recurrence or metastasis after follow-up for three to six years. It is vital to know the exact extent of the lesion before such an operation. Computerised tomography is effective and accurate in this respect. Involvement of the entire hemipelvis does not contra-indicate excision, though this becomes more difficult (Steel 1978). In the case we report, resection of the ala of the sacrum was necessary, with care to preserve the sciatic nerve. A biopsy from the retained part of the sacrum was free from involvement.

Sweetnam (1983) used the term hemipelvectomy to describe excision of the innominate bone while preserving the lower limb. He confirmed that the operation is no less radical than hindquarter amputation in terms of the proximal level of bone division, since bone resection can be at or, as in our case, beyond the sacro-iliac joint.

The authors would like to thank Dr Roger Drury, Consultant Pathologist, Plymouth General Hospital, Professor P. Byers, Professor of Morbid Anatomy, Royal National Orthopaedic Hospital and Dr Peter Antony, Consultant Pathologist, Royal Devon and Exeter Hospital, for reporting on the histological sections. We also wish to thank Mrs C. Postle-Hacon for typing the manuscript.

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VOL. 66-B. No. 5. NOVEMBER 1984
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THE JOURNAL OF BONE AND JOINT SURGERY