Locally Aggressive Fibrous Histioctytoma of Bone
A Case Report

E. W. NUNNERY, L. B. KAHN, W. B. GUILFORD

SUMMARY
A fibrohistiocytic tumour involving the distal diaphysis of the left femur in a 44-year-old woman is described. The lesion had a completely benign appearance cytologically but behaved in a locally aggressive manner in that it eroded the adjacent cortical bone and even extended into the soft tissue. To the best of our knowledge, no other similar case has been documented. A lesion of the humerus has been described as an atypical fibrous histiocytoma on the basis of prominent nuclear pleomorphism; the patient involved was alive 5 years after a disarticulation. The differential diagnosis of such a locally aggressive fibrous histiocytoma of bone would include a metaphyseal fibrous defect, benign fibroxanthoma, and malignant fibrous histiocytoma. We believe that the differentiation of a locally aggressive fibrous histiocytoma from the malignant variety may be important in view of the probable differing therapeutic and prognostic implications.


The concept of benign and malignant fibrous histiocytomas (xanthomas) of soft tissue was first introduced by O'Brien and Stout. Since then numerous benign and malignant fibrous histiocytes (MFH) arising in various sites have been documented. There have also been reports of lesions of the skin and soft tissue designated as atypical fibroxanthomas. More recently MFH has been described within bone. We have been able to find a total of 113 examples of such intra-osseous lesions in the literature, but only 6 cases of benign fibroxanthoma have been mentioned. A single case reported as atypical fibrous histiocytoma of bone has recently appeared. Dahlin et al. have alluded to 7 cases of benign or atypical fibrous histiocytomas in an article dealing with MFH of bone, but no further details were given. The purpose of this article is to document a case of fibrous histiocytoma of bone which behaved in a locally aggressive manner. We have been unable to find a report of any similar lesion.

CASE REPORT
A 44-year-old woman was admitted to the North Carolina Memorial Hospital with the complaint of acute onset of pain in the left knee. The pain progressively worsened and was associated with a rapidly progressing swelling which prevented her from walking. Sixteen years previously she had injured her knee in a motor vehicle accident and had since suffered recurrent episodes of mild knee pain. Physical examination revealed a swollen left knee with a tender area over the anterior aspect of the lateral femoral condyle and a palpable mass 4 cm above the superior pole of the patella. The patient was unable to bear weight on her left leg and she held the left knee in 30° of flexion. Results of routine laboratory tests were within normal limits. Preliminary radiographs of the left knee and distal femur revealed a lytic expansile lesion measuring 6 × 4 cm approximately 14 cm proximal to the knee joint. The superior margin of the tumour was ill defined with no recognizable transition zone. Marked invasion and erosion of cortical bone were present from the endosteal surfaces, but no periosteal new bone was observed. Several small linear areas of sclerotic bone were noted at the inferior aspect of the tumour which were thought to represent either reaction of bone to the lesion or possibly an area of a previous bone infarct (Fig. 1). The overall appearance of the lesion suggested a primary malignant process with no specific features to indicate a definite pathological diagnosis. A bone scan revealed marked activity in the femoral lesion but no metastatic lesions (Fig. 2). Tomography confirmed the...
findings of the plain film examination and suggested an area of deep cortical erosion with possible penetration to the extra-osseous soft tissues (Fig. 1). A biopsy of the lytic lesion of the left femur was performed which was histologically interpreted as 'malignant fibrous histiocytoma with areas having a benign appearance'. The patient was discharged and readmitted at a later date for an above-knee amputation.

**Pathological Features**

**Macroscopical features.** The amputation specimen revealed two well-healed scars, a 10-cm scar on the anteromedial aspect of the thigh and a 9-cm scar on the posterolateral aspect of the thigh. A longitudinal section of the femur showed the presence of a tumour measuring 5 cm in length and 3 cm in width located 10 cm from the knee joint and 7 cm from the margin of resection. It was ovoid in shape and greyish-white in colour with a soft, focal, central, reddish area. The tumour occupied the entire width of the medullary cavity and caused slight expansion of the bone. It had an irregular peripheral margin and had caused focal erosion of the cortex. In one area, there was complete disruption of the cortex by the tumour which had extended into the surrounding soft tissue.

**Microscopical features.** The tumour was composed of an admixture of foamy histiocytes and spindle-shaped cells arranged in a storiform pattern (Figs 3 and 4). The spindle cells were uniform with minimal cellular atypia (Fig. 3). No mitoses were seen in the 25 sections of tumour examined. Several large areas were composed of sheets of foamy histiocytes which resembled a pure xanthoma (Fig. 4). A few osteoclast-type giant cells were also present (Fig. 3). At the periphery, the tumour had infiltrated between spicules of lamellar bone causing this to erode (Fig. 5). In one area, the tumour had completely disrupted the cortex and invaded the extra-osseous soft tissue (Fig. 6). There was an associated mild periosteal reaction.
whether mitotic figures were observed at all. We believe the presence of mitotic activity and nuclear pleomorphism in a fibrohistiocytic tumour of bone would establish its malignant characteristics.

O'Brien and Stout 3 stressed the difficulty in defining reliable histological criteria that would predict the biological behaviour of various fibrohistiocytic tumours. As opposed to the osseous lesions, there is a fair amount of literature dealing with atypical and locally aggressive fibrohistiocytic lesions of skin and soft tissue. The lesion described as atypical fibroxanthoma of the skin is a dermal tumour with a frankly malignant histological appearance. There is usually severe cellular pleomorphism, much mitotic activity including atypical forms, and bizarre giant cells.4,5 However, nearly all these lesions behave in a clinically benign fashion and are curable by simple local excision.6 The tumour labelled as atypical fibroxanthoma of the deep soft tissues by Soule and Enriquez5 is located in the superficial or deep soft tissues and is composed of spindle-shaped cells arranged in a storiform pattern, foam cells, and giant cells. Mitoses are present but not numerous. Although these tumours have a much less disturbing histological appearance than those in the dermis, they are usually larger and biologically more aggressive because recurrences are not uncommon. These lesions may resemble dermatofibrosarcoma protuberans or giant-cell tumours of the tendon sheath if benign osteoclast-type giant cells predominate. Of 18 patients with atypical fibroxanthomas of soft tissue, 8 developed recurrences but all were free of metastases. Thirteen of the patients had been followed up for more than 5 years. Of 2 recorded deaths, only 1 could definitely be ascribed to recurrence.

DISCUSSION

The diagnosis of locally aggressive fibrous histiocytoma in this patient was based upon the presence of a locally destructive fibrohistiocytic lesion with a storiform growth pattern. There was no cytological anaplasia and a complete absence of mitotic activity in the many sections examined. A few osteoclast-type giant cells were seen. The original histological diagnosis of malignancy was probably based on the presence of foci of apparent increased cellularity in some fairly thick sections in a lesion showing disturbing radiological features. However, a review of well-prepared 5-μm sections of the tumour showed no disturbing foci of hypercellularity. Despite this completely benign histological appearance, the tumour had eroded the cortex and had even invaded extra-osseous soft tissue. In a single previously described case of atypical fibrous histiocytoma of bone, the tumour was also composed of spindle-shaped cells, arranged in a storiform pattern, foamy histiocytes, and giant cells.6 The authors considered this lesion to be atypical rather than malignant on the basis of prominent nuclear pleomorphism and an absence of atypical mitoses. However, they failed to indicate either in the text or on the photomicrographs...
of a retroperitoneal tumour. The differentiation between the atypical and the frankly malignant variety of tumour may be difficult, but minimal cellular atypia, low mitotic activity and size of the tumour are probably the most reliable criteria.

In the differential diagnosis of locally aggressive fibrous histiocytoma of bone, metaphyseal fibrous defect (fibrous cortical defect, non-ossifying fibroma), benign fibroxanthoma, and malignant fibrous histiocytoma must be considered. Metaphyseal fibrous defect is the most frequently encountered fibrohistiocytic lesion and generally occurs in the metaphyseal region of the long bones in children. The radiographic accuracy of diagnoses approaches 100% and features include an eccentrically located tumour with narrow sclerotic margins. Spjut et al. have mentioned 6 cases of benign fibroxanthoma of bone. The lesions were characterized histologically by xanthoma cells in a fibrous stroma with occasional giant cells. Spjut et al. questioned the existence of this lesion as a distinct entity, and suggested that it may represent a secondary degenerative or retrogressive change in an unrecognized pre-existing lesion.

We have recently seen a case of fibrous dysplasia of the left femur in a 76-year-old man who presented with an acute onset of pain in the left hip and posterior buttck. Radiological examination revealed a radiolucent area contained within wide, well-defined sclerotic margins. The appearance of this lesion, particularly the sclerotic 'rind', was typical of fibrous dysplasia; however, there were large focal areas composed of fibroblasts and lipid-laden macrophages producing a fibroxanthomatous appearance (Fig. 7). The diagnosis of malignant fibrous histiocytoma of bone is based upon the same histological criteria established for similar lesions occurring in soft tissue, viz. spindle-shaped cells arranged in a storiform pattern, cells with foamy or ground-glass cytoplasm, bizarre multinucleated giant cells, and prominent mitotic activity including abnormal mitoses. Benign-looking osteoclast-type giant cells may be present in moderate numbers. Dahlin et al. questioned the existence of this tumour in bone in their study of 1 120 bone tumours designated as either fibrosarcomas or osteosarcomas. They stated that the features mentioned above may be present as a component of other well-recognized osseous sarcomas but concluded that in 35 of these osseous lesions, no other components could be identified and were therefore acceptable examples of MFH of bone. The radiographical appearance of MFH is typical of an aggressive osteolytic malignant tumour. Most commonly, these tumours are metaphyseal in origin, although reports confirm their occurrence in epiphyseal, diaphyseal and axial skeletal locations. Margination is usually poor due to extensive permeation of bone and the notable lack of marrow and periosteal response, at times leading to radiographic underestimation of tumour size. Cortical erosion and permeation also characterize the aggressive nature of these tumours with frequent demonstration of soft-tissue extension from a primary osseous lesion. A less common feature is the presence of bony expansion in addition to the more typical malignant appearances. Calcification does occur in malignant fibrous histiocytoma, although it is more often seen in larger soft-tissue lesions than in those primary in bone.

The previously described patient with an atypical fibrous histiocytoma in the humerus was treated by disarticulation of the upper limb at the shoulder joint. Five years after treatment the patient was free of metastases. Our patient has been followed up for 17 months without recurrence or metastases. One of the 7 patients with benign or atypical fibrous histiocytoma referred to by Dahlin et al. has developed a pulmonary metastasis although details of histological features and therapeutic modalities are not known. The differentiation of a locally aggressive fibrohistiocytic tumour from the frankly malignant form arising in bone may be helpful in aiding the clinician both with regard to therapy and prognosis. More conservative treatment such as en bloc resection may suffice for the former, whereas the malignant variety is highly lethal and requires more radical management.

REFERENCES

Endoscopic Retrograde Cholangiopancreatography in the Management of Traumatic Pancreatic Pseudocysts

A Report of 2 Cases

L. OU TIM, P. M. THOMPSON, I. SEGAL, H. H. LAWSON

SUMMARY

The results of endoscopic retrograde cholangiopancreatography (ERCP) in 2 patients with traumatic pancreatic pseudocysts are described. As a pre-operative procedure, this investigation provided useful information on the exact site of duct disruption. In both patients, the pancreatic pseudocysts were drained via a posterior cyst gastrostomy, and they have remained well since surgery. Follow-up ERCP at 6 and 12 months demonstrated complete stenosis at the site of duct disruption. The value of ERCP in the pre-operative and follow-up management of traumatic pancreatic pseudocysts is discussed.


Blunt injury to the pancreas is uncommon and should be suspected in any patient with a history of forcible compression of the upper abdomen. This may occur as a result of kicks and punches in fights, steering wheel and seat-belt injuries in motor accidents, and falls on the handlebars of bicycles and tricycles in children.

Pseudocyst formation is a common complication of pancreatic injury. Indeed, pancreatic injury is often not suspected until a pseudocyst develops a few weeks or months later. The diagnosis is dependent on the presence of a tender abdominal mass, with abnormal elevation of the serum amylase level, and leucocytosis. Further diagnostic help may be obtained by barium meal investigation, abdominal ultrasound examination, and, in some cases, by arteriography.

The treatment of choice of traumatic pancreatic pseudocysts is surgical, viz. internal drainage, or pancreatic resection. The pre-operative demonstration of the exact site of the pancreatic duct disruption would provide useful additional information for the selection of the appropriate surgical therapy. Before the introduction of endoscopic retrograde cholangiopancreatography (ERCP), this was only possible by means of intra-operative pancreateography.

In this report the utilization of ERCP in the pre-operative diagnosis, and the follow-up management of 2 patients with pancreatic pseudocyst formation after abdominal trauma are described.