MALIGNANT FIBROUS HISTIOCYTOMA
AN UNUSUAL LESION OF INTEREST TO THE ORTHOPAEDIC SURGEON

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Malignant fibrous histiocytoma is an unusual musculoskeletal tumour which typically presents in the fifth and sixth decades. Extensive radiographic and laboratory investigations are seldom helpful in diagnosis, although scanning techniques and angiography may assist in determining the extent of the tumour. Wide local excision has been the most common form of treatment for this lesion, after open biopsy and tissue culture has confirmed the diagnosis. Recurrence rates, however, often exceed 30 per cent. Illustrative cases seen at the Montreal General Hospital are presented and their management discussed.

Malignant fibrous histiocytoma has received considerable attention in the oncological literature in recent years but few articles dealing with this lesion have appeared in the orthopaedic literature, despite the fact that the location and nature of the tumour would most probably bring the patient to the orthopaedic surgeon. In the vast majority of reported cases the patients have presented with a musculoskeletal mass, usually involving the soft tissues of the limbs or trunk.

It is the purpose of this paper to describe three cases of malignant fibrous histiocytoma of soft tissue and to review the characteristics of this rare but clinically significant lesion.

CASE REPORTS

Case 1. A seventy-five-year-old woman was admitted to Montreal General Hospital for investigation of a lump over the anterior aspect of her left knee; it had become tender and painful, limiting movement of the knee. On examination the mass was found to be well defined; it measured 8 by 10 centimetres and was adherent to the skin and to the deep tissues. All laboratory findings were within normal limits. Radiography revealed a sharply defined, homogeneous soft-tissue mass which contained no calcification; no bony abnormality was detected (Fig. 1). An angiogram revealed that the lesion was hypervascular with evident disruption of small vessels, pooling of contrast material, arteriovenous shunting, and large venous drainage channels (Fig. 2). Bone scanning with a polyphosphate demonstrated an accumulation of tracer over the left knee (Fig. 3). Scans of brain, liver and spleen, as well as radiographs of the chest, gave normal results.

An excision-biopsy was carried out. No obvious demarcation of the tumour was noted at the time of operation, nor was there any apparent extension of the tumour mass within the knee joint. The pathological diagnosis was malignant fibrous histiocytoma.

The patient's recovery from operation was uneventful. Radiotherapy was begun when sutures were removed. A follow-up examination two years after excision revealed no evidence of residual tumour.

Fig. 1

Fig. 2

Fig. 3

Case 1. A soft-tissue mass. Figure 1—Radiograph showing that the density of the mass over the anterior aspect of the left knee was sharply defined, homogeneous and contained no calcification. Figure 2—Angiogram revealing hypervascularity of the soft-tissue mass, with disruption of the small vessels and pooling of the contrast material. Figure 3—Polyphosphate bone scan revealing accumulation of the tracer in the soft-tissue mass.

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Case 2. A fifty-year-old woman was admitted for investigation of a lump on her left shoulder; it had been present for two years, but had recently increased in size. The patient had a history of infiltrating duct carcinoma of the left breast for which she had undergone local excision and radiotherapy sixteen years earlier. Physical examination revealed radiation scars over the left shoulder and chest. An erythematous and tender mass, measuring 8 by 10 centimetres was present on the left shoulder. No lymphadenopathy was noted. Results of all laboratory investigations were within normal limits. Radiological examination of the left shoulder revealed a soft-tissue mass with punctate calcification; the chest radiograph was normal. A bone scan showed increased uptake of polyphosphatc in a diffuse pattern over the left shoulder.

An excision-biopsy was attempted, but the entire mass could not be removed because it was found to extend deep along the scapula. Histological sections demonstrated an obvious malignant fibrous histiocytoma.

A left forequarter amputation was performed one month after the biopsy. Pathological examination confirmed the complete excision of the mass, although in one area of the tumour focal invasion of thin-walled blood vessels was noted.

After operation, the patient was well for five months before developing a dry hacking cough and dull parasternal pain. On her readmission to hospital, a radiograph showed several large masses in the lung (Fig. 4). Chemotherapy with vincristine, Adriamycin, Cytoxan and dacarbazine was begun shortly thereafter. Thirteen months from the start of chemotherapy, the clinical response of the patient was excellent; radiographs showed that the lung fields had cleared (Fig. 5).

Case 3. This seventy-year-old man presented with a lump, measuring 10 by 10 centimetres, over his right scapula; it had been present for ten years but had recently increased in size and become painful. The mass was fixed to deep tissues but not to the overlying skin. Results of all laboratory investigations were normal. Radiography revealed no active disease of the chest, but there was radiological evidence of a dense area of soft tissue over the right scapula. No areas of calcification were seen. Dynamic perfusion of the right scapula showed no increase in the vascularity of the tumour.

At open biopsy, the mass was found to be encapsulated, greyish-white in colour and firm. It was located over the medial aspect of the right scapula, and extended up the proximal aspect of the levator scapulae. The pathological diagnosis was malignant fibrous histiocytoma.

A wide local excision was carried out. The patient made an uneventful recovery, and there was no sign of recurrence eighteen months later.

PATHOLOGY

The tumours varied in size from 3 to 11 centimetres in diameter. All were circumscribed, exhibiting pseudo-encapsulation. Section of the gross specimen revealed a firm, white, glistening surface with small areas of necrosis (Fig. 6).

Histologically, the lesions were composed of interlacing bands of fibroblasts, bizarre histiocytes and multinucleated giant cells. A characteristic storiform pattern was seen in all cases as well as normal and abnormal mitoses (Fig. 7).
yet been unable to determine whether these populations represent two distinct cell lines or two pathways of differentiation of a single mesenchymal cell line transformed by the neoplastic stimulus (Fu et al. 1975).

**Clinical characteristics.** Malignant fibrous histiocytoma has been reported in patients aged from eighteen months to eighty-two years, with a mean age of occurrence in the fifth decade. Males are affected more often than females in a ratio of 1.5:1 to 2.1:1 (O'Brien and Stout 1964; Kempson and Kyriakos 1972; Soule and Enriquez 1972). The patient typically presents with a soft-tissue mass in a limb (Kempson and Kyriakos 1972; Soule and Enriquez 1972).

The aetiology of these tumours remains unknown. Often the soft-tissue mass has been present for some time, even years, before pain and a rapid increase in size denote a change in character of what was previously a very slow-growing lesion. There have been occasional reports of recent injury to the involved area.

Radiology most often reveals a soft-tissue mass which is ill-defined and lacking in any distinguishing features. Punctate calcification may be present; vascularity may be increased. The uptake of technetium-labelled polyphosphate is increased, as it is in many soft-tissue tumours (Poulose et al. 1975). Angiography plus scanning may help to define the extent of the lesion.

**Treatment and prognosis.** The operative treatment of soft-tissue sarcomata in general, and of malignant fibrous histiocytoma in particular, has traditionally been by local or en bloc excision of the tumour where possible. Amputation is carried out where more conservative means have failed to eradicate the tumour. It is generally agreed that the prognosis of these lesions in the absence of metastases is dependent upon local control of the tumour (Bowden and Booher 1958; Cantin et al. 1968; Simon and Enneking 1976). Because these tumours have a tendency to encapsulate and to spread along fascial planes, precise definition of the mass is difficult and this has led to a high recording of local recurrence. Distant metastases occur most commonly by the haematogenous route, probably from emboli originating in the vascular areas of the tumour, or from operative manipulation. Regional lymphatic spread is less common (Simon and Enneking 1976) but has been shown to occur with the primary tumour of bone (Spanier, Enneking and Enriquez 1975).

Kempson and Kyriakos (1972) have presented follow-up information for twenty-two patients who had undergone local excision: nine had developed a recurrence from six weeks to three years later, the average time being nine months, with the largest tumours being the more likely to recur.

In a comparison of those tumours which had metastasised with those which had not, the authors found no difference in the degree of pleomorphism, the number of giant cells, the presence of necrosis or the number of mitoses. All the tumours which metastasised

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**DISCUSSION**

The term "fibrous histiocytoma" is generally applied to a group of lesions which appear to be of histiocytic origin. These lesions often pose problems in diagnosis because of the wide variety of histological patterns which may be present.

**Nomenclature.** For many years, these soft-tissue tumours were considered to be completely benign. They were given various names: dermatofibroma, dermatofibrosarcoma protuberans, nevoid histiocytoma, juvenile xanthogranuloma, sclerosing haemangioma of soft tissue, giant-cell tumour and giant-cell tumour of tendon sheath, fibrous histiocytoma, xanthofibroma, villonodular synovitis or sclerosing haemangioma (Feldman and Norman 1972; Soule and Enriquez 1972).

**Cell of origin.** O'Brien and Stout (1964) were the first to establish the malignant potential of these tumours and relate it to the variable neoplastic potential of the tissue histiocyte. Subsequent tissue culture confirmed their findings. The histiocyte, however, may behave as a facultative fibroblast giving rise to a fibroblastic element in the tumours (Ozzello, Stout and Murray 1963; Merkow et al. 1971). More recently, researchers using the electron microscope have again emphasised the bimodal cell population of these tumours, but have as
were noted to have infiltrating borders; the same findings were reported for the locally recurrent tumours.

Evaluation of recent treatment of these lesions has led to a more optimistic outlook. Radical-dose radiotherapy combined with limited operative treatment has been effective in controlling soft-tissue sarcomata (Suit, Russell and Martin 1975). Furthermore, soft-tissue sarcomata in general could have a significantly lower recurrence rate when treated by adequate radical resection, bearing in mind their tendency to spread along fascial planes (Simon and Enneking 1976).

In the treatment of recurrences and metastatic disease, responses to chemotherapy and radiotherapy are often unpredictable. It has recently been suggested, in a study of primary malignant fibrous histiocytoma of bone, that the response of metastases to chemotherapy or radiotherapy will depend on the nature of the metastasising component; that is, primarily histiocytic lesions might be more radiosensitive than fibroblastic ones. It is possible, therefore, that biopsies of metastatic deposits could influence the choice of therapy (Spanier et al. 1975).

In summary, malignant fibrous histiocytoma is an unusual lesion of soft tissue, with a specific bimodal histological pattern which often leads to confusion in diagnosis. As this type of tumour becomes better recognised by the pathologists, it will probably appear more frequently to the orthopaedic surgeon as a diagnostic and therapeutic challenge.

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REFERENCES


