Acta Medica Okayama

Volume 32, *Issue* 5 1978 *Article* 4

OCTOBER 1978

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KEYWORDS: malignant fibrous histiocytoma, bone tumor

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*PMID: 215002 [PubMed - indexed for MEDLINE]

Acta Med. Okayama 32, (5), 343—354 (1978)

MALIGNANT FIBROUS HISTIOCYTOMA WITH SKELETAL INVOLVEMENT

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Received May 10, 1978

Abstract. Malignant fibrous histiocytoma of soft part is rather common but malignant fibrous histiocytoma of the bone is rarely encountered clinically. Authors present five cases of malignant fibrous histiocytoma with skeletal involvement and discuss their clinical course, x-ray findings and histological features. This tumor has marked tendency for local recurrence and metastasis. Other bone tumors such as giant cell tumor, aneurysmal bone cyst, non ossifying fibroma, osteosarcoma, fibrosarcoma of bone and metastatic cancer can be excluded by several characteristic findings observed in x-rays as well as histopathological features. All information on the patient should be carefully analysed, because it is difficult to decide whether bone involvement is primary or secondary. Four out of five cases definitely originated within the bone.

Key words : malignant fibrous histiocytoma, bone tumor

Soft part tumors which originated from tissue histiocytes had various characteristic histological features, therefore there have been used several names depending on their location as well as histopathological findings. Stout and his co-workers (1) investigated these tumors and simplified their nomenclature in 1967 and established fibrous histiocytoma as an entity which was generally accepted as a benign lesion.

Malignant fibrous histiocytoma of soft parts was reported first by O'Brien and Stout (2) in 1964. Later Guccion and Enzinger (3), Kempson and Kyriakos (4), and Soule and Enriquez (5) reported similar tumors. Since Feldman and Norman (6) reported nine cases of introsseous malignant histiocytoma, it is thought that this tumor could originate not only from soft part but also from bones. The authors (7) reported two cases of malignant fibrous histiocytoma with skeletal involvement in 1975 but at that time much attention was given to the behavior of this tumor.

The purpose of this paper is to analyse five cases of malignant fibrous histiocytoma with skeletal involvement clinically, radiologically and histopathologically.

MATERIALS AND METHODS

From 1971 to 1976 five cases of malignant fibrous histiocytoma with skeletal involvement were treated in the Department of Orthopedic Surgery, Okayama University Hospital. The clinical, radiological and pathological findings were as follows.

CLINICAL AND PATHOLOGICAL OBSERVATIONS

Clinical features. Age, sex, affected bone and location of the bone were shown in Table 1. Every patient came to the clinic complaining of slight spontaneous pain in the affected region and bone lesions were found by x-ray examination. The laboratory data showed no abnormalities. The age of the patients ranged from thirty-five to fifty years.

TABLE 1. LOCATIONS OF MALIGNANT FIBROUS HISTIOCYTOMA IN THE CASES

Case	Age	Sex	Affected bone	Location in the bone
1	50	Female	Left femur	Distal metaphysis
2	39	Female	Left femur	Greater trochanter
3	37	Male	Left femur	Distal metaphysis and epiphysis
4	39	Male	Left tibia	Diaphysis
5	35	Female	Left scapula	Neck of scapula

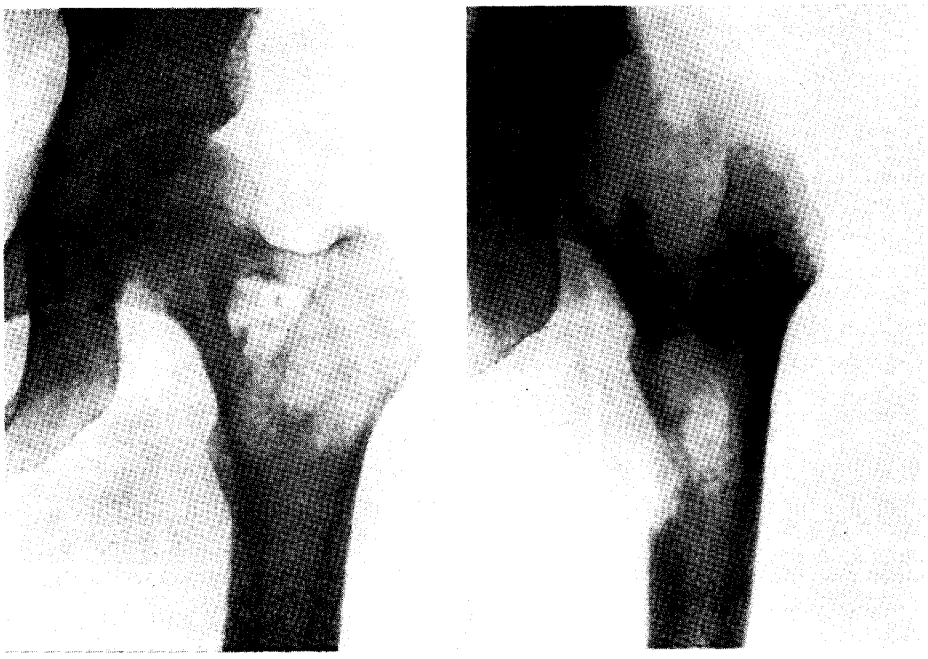
Two cases were evaluated as a benign lesion at the time of the first operation. Case 2 was diagnosed as a solitary bone cyst clinically as well as pathologically. Curettage and bone graft were performed. But during the year after this operation the bone lesion enlarged gradually and the disarticulation of the left hip joint was performed. Six months after the disarticulation, pulmonary metastasis was found. Case 3 was diagnosed as a pathologically benign lesion and curettage with bone graft was performed. Two months after the operation, hemarthrosis of the left knee joint was found. Since malignant tumor cells were recognized in the aspirated bloody fluid, above the knee (A/K) amputation was carried out. Case 1 and case 4 were amputated primarily and there was no local recurrence. Two years and four month after A/K amputation, pulmonary metastasis was found in case 1. Case 4 have had no metastasis and is quite healthy two years after below the knee (B.K) amputation. In case 5 the skeletal involvement seemed to be secondary at the time of partial scapulectomy. Local recurrence occurred one year and ten months after the first operation and at the same time a metastatic lesion was found in the left acetabulum. Both lesions were resected and the lesion of acetabulum was reconstructed by bone cement. Metastasis in both the lung and the lumbar vertebrae were recognized radiologically two years and three months after the first operation. The clinical course of these patients is shown in

TABLE 2. CLINICAL COURSE OF THE PATIENTS SHOWN IN TABLE 1

Case	First operation	Local recurrence	Second operation	Metastasis	Death	Autopsy
1	Left A/K amputation	(−)	(−)	Lung, 28 months	34 months	(+)
2	Curettage, bone graft	(+) 12 months	Disarticulation of left hip	Lung 18 months	24 months	(−)
3	Curettage, bone graft	(+) 2 months	Left A/K amputation	Lung 10 months	15 months	(+)
4	Left B/K amputation	(−)	(−)	(−)	Healthy 24 months	(−)
5	Left partial scapulectomy	(+) 22 months	Resection	Left acetabulum 22 months lung 27 months lumbar vertebrae 28 month	36 months	(−)

Table 2.

X ray findings. A localized intraosseous lesion was observed in preoperative x-ray pictures of cases 2, 3 and 4. These lesions seemed to have originated from bone. The lesion was commonly a monolocular radiolucent area, although several



a b

Fig. 1. case 2 a: x-ray picture before the first operation.
b: x-ray picture after one year after the onset.

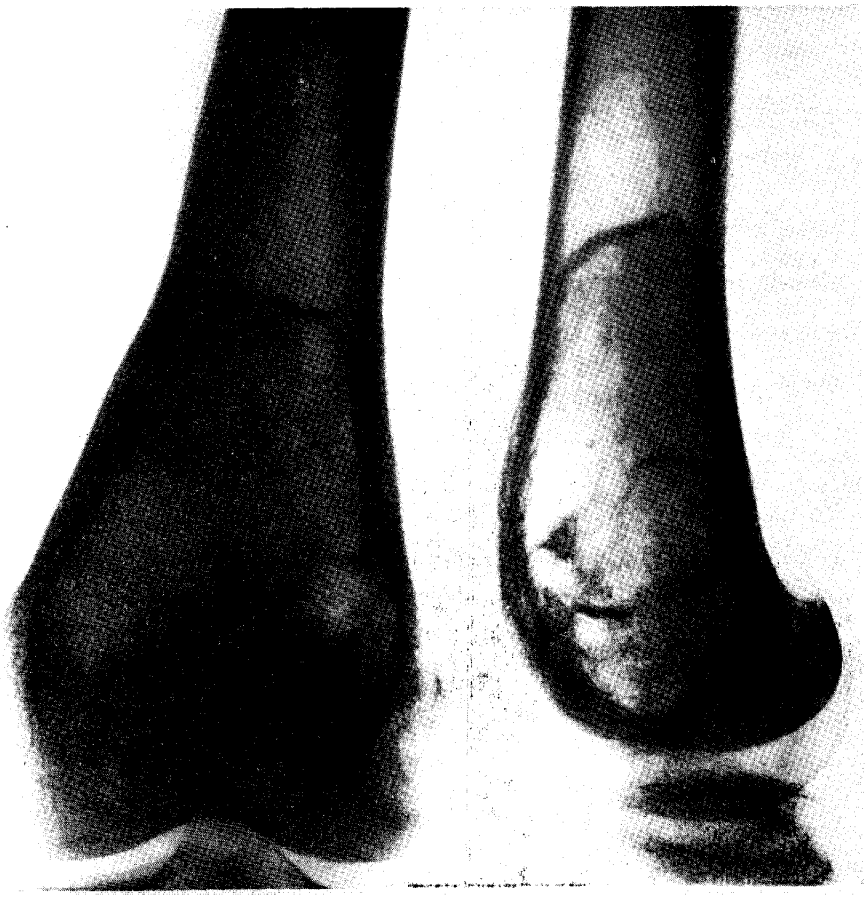


Fig. 2. case 3 x-ray picture at the onset.

septum-like shadows were recognized in the radiolucent area. Marginal sclerosing was observed partially at the border of the radiolucent area. These radiolucencies were located concentrically in the bone. No deformity of the bone contour was seen, even if the radiolucency became huge one (case 3). The location of the bone lesion had no definite rules, *i.e.* this lesion could appear in diaphysis (case 4), metaphysis (case 2) and from metaphysis to epiphysis (case 3). From these observations giant cell tumor, aneurysmal bone cyst and non ossifying fibroma could be ruled out. X-ray findings of case 2 during follow up examinations showed the pattern of progress of the bone lesion. The marginal sclerosing which was observed partially at the initial stage disappeared gradually. Then the radiolucency changed to osteolysis and finally the cortex was destroyed. This final x-ray picture was quite similar to the osteolysis which was seen very often

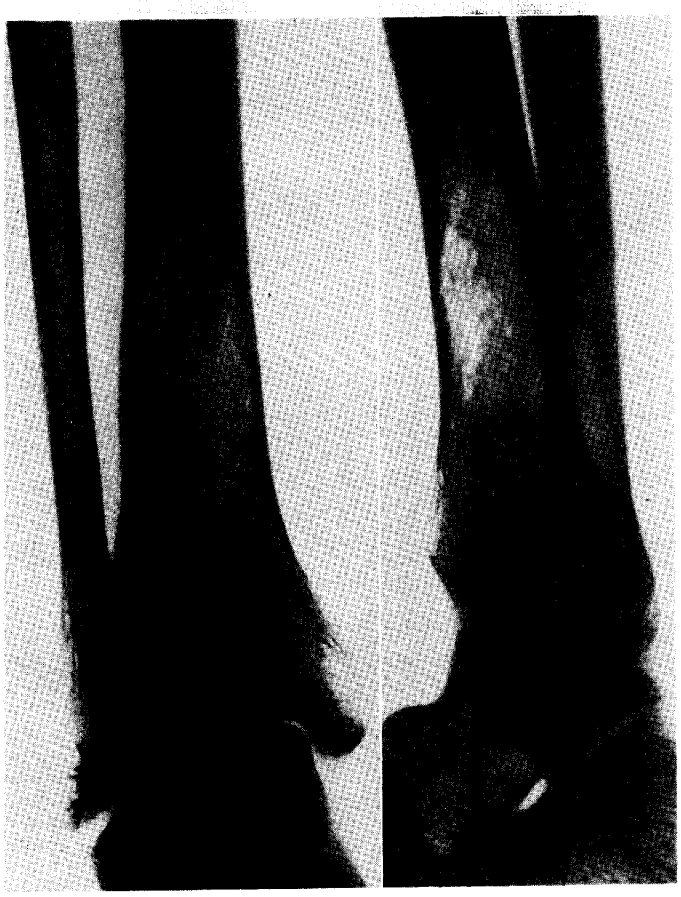


Fig. 3. case 4 x-ray picture at the onset.

in bone metastasis of cancer. X-ray picture of case 1 showed possibility of bone metastasis of cancer as the first choice for the above-mentioned reasons. But in the antero-posterior view, remnants of localized cystic radiolucency was partially recognized. The x-ray findings of case 5 show osteolysis, however, the border of osteolysis was quite irregular and marginal sclerosing was also observed. From the microscopic findings as well as macroscopic findings at the time of operation, it was concluded that this bone lesion was a secondary invasion of the tumor originating from soft parts. The x-ray picture of metastatic foci in acetabulum showed osteolysis with marginal sclerosing.

Summerizing these points, it is said that there are several characteristic x-ray findings of malignant fibruos histiocytoma of bone when the lesion is completely within bone. However, in the advanced stage characteristic findings disappeared and marked osteolysis was observed.

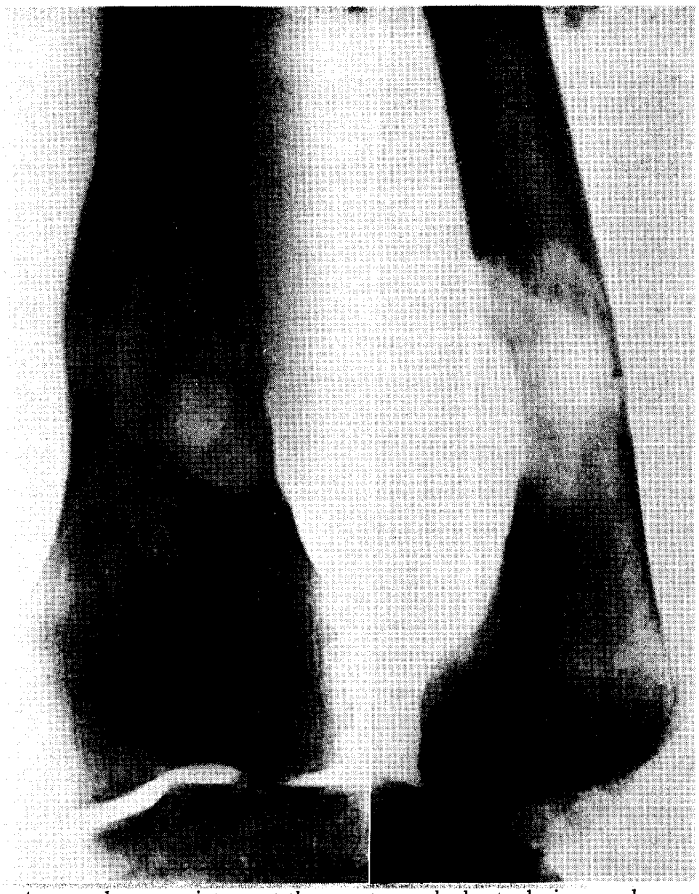


Fig. 4. case 1 x-ray picture at the onset, marked osteolysis was observed.

Fig. 5. case 3 x-ray picture at the onset, irregular osteolysis was observed in the neck of scapula.

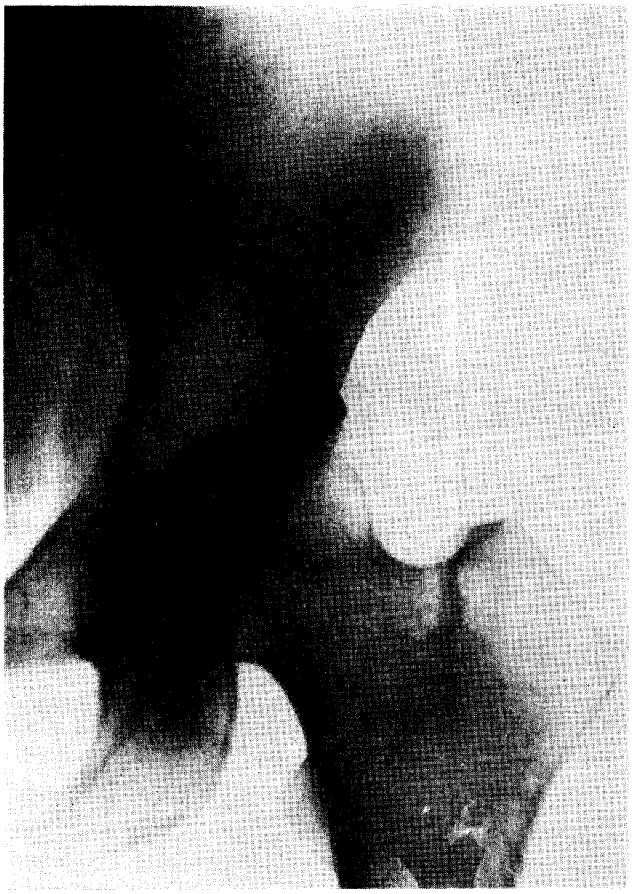


Fig. 6. case 5 metastatic lesion of the left acetabulum.

These characteristic findings are: a) There is no predilection site for the lesion in bone. The lesion could be found in diaphysis, metaphysis or epiphysis. b) The lesion was situated concentrically in bone and not eccentrically. c) The lesion is observed as a monolocular radiolucent area with partial marginal sclerosing but septum-like structure are rarely observed. d) No deformity of bone contour is observed even if the lesion is a huge one. e) In advanced stage the lesion shows marked osteolysis with irregular margin. f) No periosteal reaction was observed. g) When bone was invaded secondarily by the soft part tumor, the margin of bone destruction was irregular and accompanied by marginal sclerosing.

Pathological findings. Malignant fibrous histiocytoma had various marked histological features namely: a. Short spindle shaped tumor cells which have been called facultative fibroblasts arranged in storiform pattern and producing rich collagen fibers. b. The storiform pattern was observed even in the area

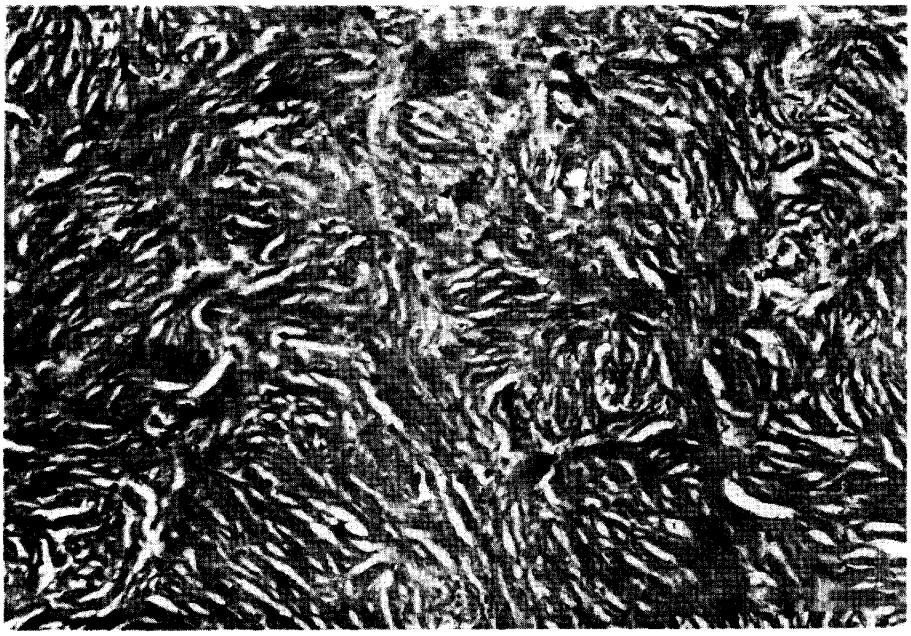


Fig. 7. Tumor cells arranged in typical storiform pattern and produced rich collagen fibers where cellularity was low. Collagen fibers showed marked hyalinization. (case 1, Hematoxylin-Eosin staining. ×200)

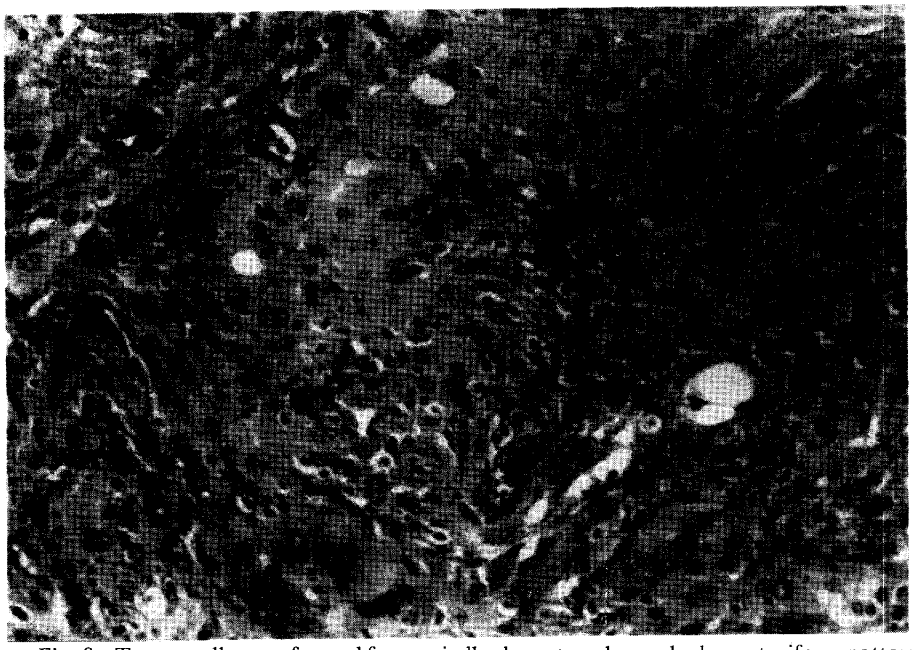


Fig. 8. Tumor cells transformed from spindle shape to polygonal where storiform pattern disappeared. In the center polygonal epithelioid cells with abundant cytoplasma were seen. There were intermingling histiocytic cells which showed phagocytic activity. (case 2, Hematoxylin-Eosin staining. ×800)

Fig. 9. Many multinucleated giant cells with bizarre nuclei and irregular in shape were observed and round cell infiltration which consisted mainly of lymphocytes was also seen. (case 3, Hematoxylin-Eosin staining. ×200)

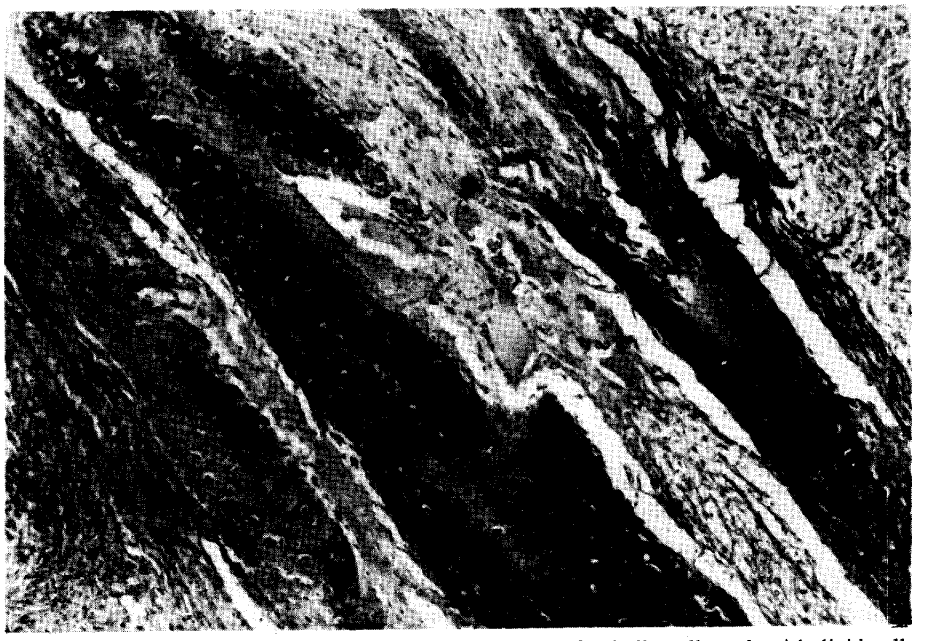


Fig. 10. Cortex of bone was destroyed by invasion of spindle cells and epithelioid cells. (case 4, Hematoxylin-Eosin staining. ×200)

where collagen fibers showed marked hyalinization. c. Polygonal tumor cells with abundant cytoplasm which were thought to be histiocytes varied in size. These cells and short spindle shaped tumor cells intermingling profusely. In these areas multinucleated giant cells were also observed.

These three main features occurred irregularly. In one high power field mitosis was observed in up to 10 cells and tumor cells showed marked pleomorphism. In the center of the tumor, there was relatively large necrotic foci, however, in the peripheral parts small necrotic foci were scattered. Around these necrotic foci round cell infiltration which consisted of mainly lymphcytes as well as small foamy cells were observed. Small fragments of bone and disrupted muscle fibers were found in the peripheral part of the tumor. Because these bone fragments showed lamellated structure without osteoblastic activity at the surface, it was considered that these bone tissues were not newly formed bones but a destroyed one. Touton type giant cells were observed in some cases. Silver impregnation showed that both spindle and polygonal tumor cells were surrounded by fine reticulin fibers. Moreover reticulin fibers were also storiform where tumor cells showed a storiform pattern.

DISCUSSION

Yumoto (8) has investigated cell components and histological features of histiocytic tumors originated from soft parts and further discussed their nomenclature. He stated that histiocytes, Touton type giant cells and storiform pattern were situated on each vertice of the equilateral triangle in his schema. The transitional process from histiocytes to Touton type giant cells was a xanthomatous change and the transitional process from histiocytes to storiform pattern was fibrogenesis. Therefore, specific names should be given in proportion to the dominancy of these three components (Fig. 11). All of our cases were considered to be situated at the right low vertice in Yumoto's schema.

Soule (5) summerized eleven items which were shown in Table 3 as the characteristic histological findings of histiocytic tumors. He emphasized that the anaplasia of stromal cells should be important for judgement of malignancy. As the evidence of malignancy, Rosar-Uribe (9) emphasized metastasis and Stout (1) indicated high frequency of mitosis, aggressive proliferation of tumor cells as well as metastatic foci.

In 1972 Feldman and Norman (6) investigated twenty cases of malignant histiocytoma and proved that nine of them originated from bone. In 1974 Mirra *et al.* (10) have reported four cases of malignant bone tumor associated with bone infarcts. Three of them were malignant fibrous histiocytoma. And they stated that the characteristic microscopic findings of the malignant fibrous histiocytoma of bone were: a. Bundles and whorls or spindle shaped, fibroblastlike cells

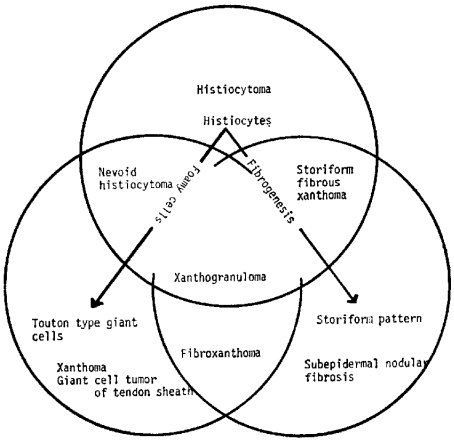


Fig. 11. Schema showing histological relationship between several types of fibrous histiocytoma (Yumoto, 1973).

TABLE 3. SYNOPSIS OF HISTOLOGIC FEATURES OF MALIGNANT FIBROUS HISTIOCYTOMA (SOULE & ENRIQUEZ, 1972) (5)

Histiocytic-like (epithelioid) cells
Spindle cells (facultative fibroblast)
Fibrogenesis
Storiform pattern
Multinucleated benign giant cells
Tumor giant cells (often bizarre)
Foam cells
Inflammatory cell (usually lymphocytes)
Anaplasia of stromal cells
Mitotic figures (normal and atypical)
"Granulomatous" features

constitute most of the lesion and are arranged primarily in storiform pattern; the cells and fibers often meet one another at right angles and sometimes take on a pinwheel pattern. The nuclei of the spindle cells display features of anaplasia. b. Foci of rounded cells may be found, with a foamy or vacuolated cytoplasm containing bizarre, pleomorphic, and multianaplastic nuclei with atypical (non-mirror image) mitotic figures. c. There is evidence of phagocytosed intracytoplsmic material including hemosiderin, hematin, and lipofuscin pigments. d. There also is evidence of patchy or extensive reticulin-fiber production.

However, it must be quite difficult to decide whether the lesion originated within the bone or not by histological findings alone, because these findings were generally observed in malignant fibrous histiocytoma of soft parts. Besides these authors eleven malignant fibrous histiocytoma of bone were reported by Spanier *et al.* (11) in 1975, one by Newland *et al.* (12) in 1975 and eighteen by Huvos (13) in 1976. In 1977 Dahlin *et al.* (14) reviewed one hundred and fifty eight fibrosarcomas of bone and nine hundred and sixty-two osteosarcomas in their files and stated that thirty-five tumors seemed to be malignant fibrous histiocytoma because these cases provided the histological spectrum which was characteristic of this lesion. They concluded that malignant fibrous histiocytoma was a valid designation for a bone tumor if thorough sampling proved that they were not being deceived by the histologic appearance by lesions which could turn out to be fibrosarcoma, osteosarcoma or even metastatic carcinoma when studied completely.

In order to establish the definite diagnosis of malignant fibrous histiocytoma

of bone, one should take as large a piece of tissue as possible from the affected part and find out the characteristic histological features which were mentioned in the above three items. Moreover it must be carefully decided radiologically and clinically whether the tumor originated in the bone or not.

REFERENCES

1. Stout, A. P. and Lattes, R.: Tumors of soft tissues, In *Atlas of Tumor Pathology*, Series 2, Fasc. 1, AFIP, Washington, D. C., pp. 38-52, 1967.
2. O'Brien, J. E. and Stout, A. P.: Malignant fibrous xanthoma. *Cancer* **17**, 1445-1455, 1964.
3. Guccion, J. G. and Enzinger, F. M.: Malignant giant cell tumor of soft parts, an analysis of 32 cases. *Cancer* **20**, 1518-1529, 1972.
4. Kempson, R. L. and Kyriakos, M.: Fibroxanthosarcoma of the soft tissue, a type of malignant fibrous histiocytoma. *Cancer* **29**, 961-976, 1972.
5. Soule, E. H. and Enriquez, P.: Atypical fibrous histiocytoma, malignant fibrous histiocytoma and epithelioid sarcoma. a comparative study of 65 tumors. *Cancer* **30**, 128-143, 1972.
6. Feldman, F. and Norman, D.: Intra- and extraosseous malignant histiocytoma (malignant fibrous xanthoma). *Radiology* **104**, 497-508, 1972.
7. Takechi, H. and Taguchi, K.: Malignant fibrous histiocytoma. *Seikeigeka* **26**, 335-342, 1975 (in Japanese).
8. Yumoto, T.: On the fibrous histiocytoma. *Rinshoseikeigeka* **8**, 698-713, 1973 (in Japanese).
9. Rosas-Uribe, A., Ring, A. M. and Rappaport, H.: Metastasizing retroperitoneal fibroxanthoma (malignant fibroxanthoma). *Cancer* **26**, 827-831, 1970.
10. Mirra, J. M., Bullough, P. G., Marcove, R. C., Jacob, B. and Huvos, A. G.: Malignant fibrous histiocytoma and osteosarcoma in association with bone infarcts. *J. Bone Jt. Surg.* **56A**, 932-940, 1974.
11. Spanier, S. S., Enneking, W. F. and Enriquez, P.: Primary malignant fibrous histiocytoma of bone. *Cancer* **36**, 2084-2098, 1975.
12. Newland, R. C., Harrison, M. A. and Wright, R. G.: Fibroxanthosarcoma of bone. *Pathology* **7**, 203-208, 1975.
13. Huvos, A. G.: Primary malignant fibrous histiocytoma of bone. *NY State J. Med.* **76**, 552-559, 1976.
14. Dahlin, D. C., Unni, K. K. and Matsuno, T.: Malignant (fibrous) histiocytoma of bone fact or fancy? *Cancer* **39**, 1508-1516, 1977.