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Intraosseous "Chordoid" Sarcoma, Chondroblastic or Lipoblastic Origin?

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Summary. An unusual sarcoma with myxoid features that apparently originated within the scapula and has been locally aggressive for 10 years is reported. Clinically and by light microscopy it was considered to be a cartilaginous tumor, possibly chordoid sarcoma. By histochemical techniques and electron microscopy it most closely resembles a tumor of brown fat.

Key words: Sarcoma – Hibernoma – Liposarcoma.

Introduction

A locally aggressive tumor involving the scapula and surrounding soft tissue was examined by light and electron microscopy and histochemical techniques. Although the light microscopic appearance was compatible with "chordoid sarcoma" (Stewart, 1948) several factors mitigate against that diagnosis, at least as it is currently used (Mehio and Ferenczy, 1978; Weiss, 1976). The current case is probably of lipoblastic origin, more like brown than yellow fat.

Case Report

A twenty-six year old black man was admitted to Presbyterian-University Hospital for resection of a right scapular tumor.

He had been well until ten years before admission, when he complained of pain in the right shoulder. Roentgenograms at that time showed a well circumscribed erosive lesion of the glenoid and coracoid regions of the scapula with two intra-osseous satellite lesions (Fig. 1). Biopsy was performed and a diagnosis of chondromyxoid fibroma was made. The tumor was considered to be primary in the bone. Seven months later excision of the mass was performed and a diagnosis

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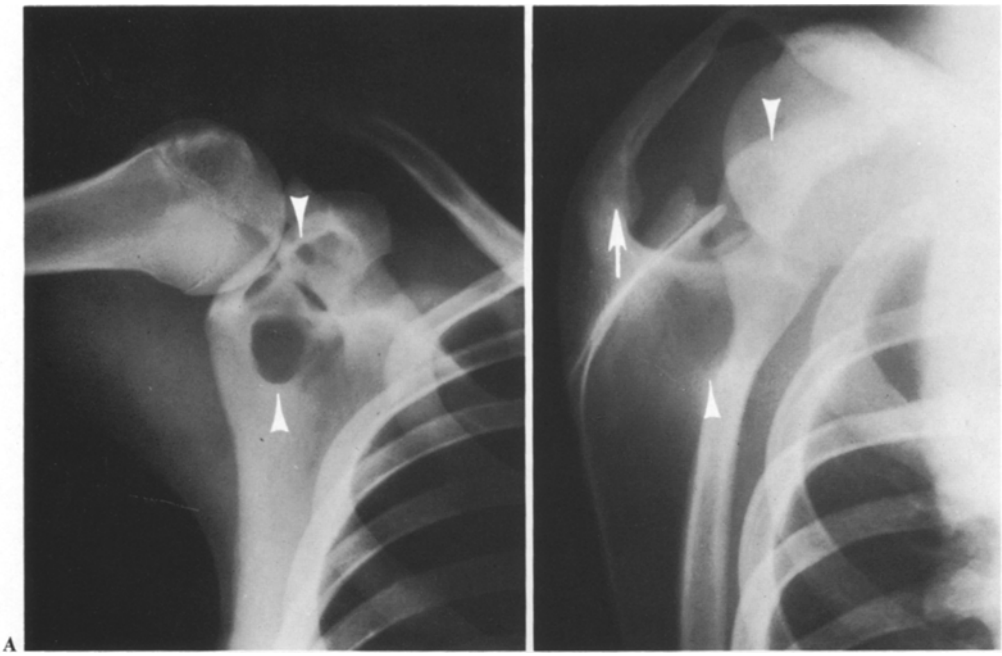

Fig. 1. A Anterior-posterior view. Smooth-walled intrascapular lucency (*arrowheads*) involving the glenoid region. **B** Rotated view demonstrating a large apparently primary lesion of the glenoid fossa extending into the coracoid (*arrowheads*) and an apparently separate lesion of the acromion (*arrow*)

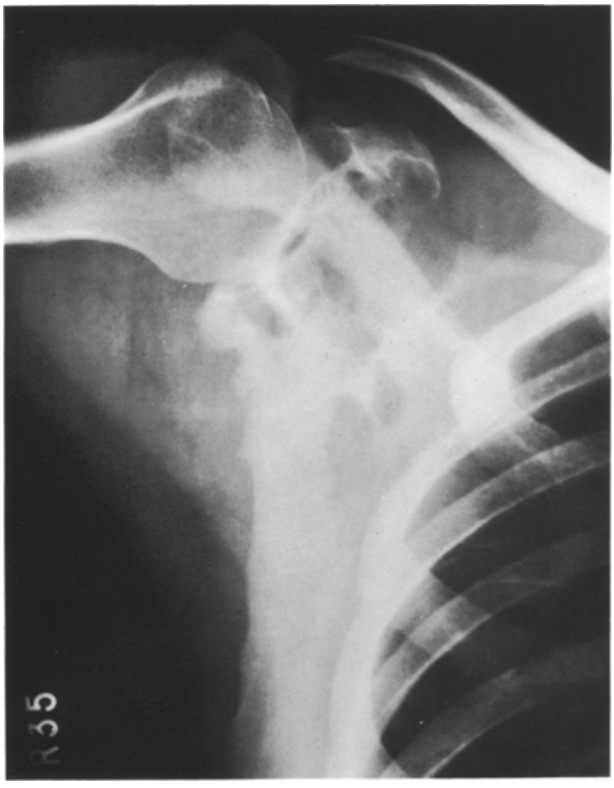

Fig. 2. X-rays upon current admission, showing multiple smooth-walled lucencies and extensive scalloping of the lateral margin of the scapula

of chondromyxosarcoma was made. Because of recurrent pain and radiologic evidence of recurrence, excision of recurrent tumor was performed five years after the first excision.

Once again the pain and a palpable mass recurred and progressed during the year before his most recent hospitalization. Admission physical examination revealed slight fullness in the soft tissues of the lateral aspect of the right trapezius insertion, tenderness in the supraclavicular area, the mid-point of the scapular spine and the intraspinous fossa, and atrophy of the infra-spinous muscle and the right upper extremity. There was a full range of motion with normal neurological and circulatory findings. Roentgenograms showed multiple lucencies with smooth sclerotic margins in the superior border and glenoid region of the right scapula. The largest was 5.0×5.5 cm. The lateral scapular border was scalloped and the superior border was not identifiable, suggesting soft tissue extension (Fig. 2).

At exploration, glistening, soft, yellow lobulated tumor involved the supraspinous muscle and extended completely through the scapula, impinging upon the subscapularis muscle. It extended into the infra-spinatous fossa. Excision with sacrifice of portions of the infra-spinatous and teres minor muscles was performed. The patient was discharged three days postoperatively. There is no evidence of distant metastases.

Materials and Methods

Tissue from the latest operation was received fresh. Sections of grossly recognizable tumor were processed with formalin fixation and paraffin embedding for light microscopy. Hematoxylin and eosin staining was performed on all blocks. Verhoeff-van Gieson, periodic-acid-Schiff with and without diastase, Jones Methenamine Silver, Fraser-Lendrum and toluidine blue were performed on selected blocks. Oil red 0 was performed on frozen tissue. Alcian blue (pH 1.0, 2.5), mucicarmine and colloidal iron were performed with and without hyaluronidase digestion. Immunoperoxidase staining for alpha-l-antitrypsin and alpha-fetoprotein were performed using the bridge technique.

Tissue from multiple areas was minced into 1 mm cubes, fixed in 4% paraformaldehyde, dehydrated and embedded in Epon-araldite. One micron sections were cut, stained with toluidine blue and examined by light microscopy. Thin sections were cut, stained with lead citrate and uranyl acetate and examined with an electron microscope. Microscopic slides and roentgenograms from the first resection were reviewed.

Pathology

The most recent operation yielded $15.0 \times 6.0 \times 3.0$ cm of tissue. Approximately 3/4 was soft, tan, lobulated and myxoid tumor with focal firmer yellow areas. The rest was muscle and fibrous tissue. There were no definable margins around the tumor. No bone or cartilage were apparent.

By light microscopy, the tumor was composed of moderate sized polygonal to spindled cells with round to oval nuclei without mitoses. Chromatin was not markedly increased. A small amount of slightly eosinophilic cytoplasm was present, with fine vacuolization. Cells were separated by large amounts of myxoid stroma with moderate Alcian blue, mucicarmine, and colloidal iron positivity which was abolished by hyaluronidase digestion. The Alcian blue reaction was the same at both pH's. Oil red 0 was negative.

Frequent large eosinophilic bodies were present within the stroma (Fig. 3). They stained strongly for fibrin and were PAS positive, diastase resistant. They did not stain with the Jones Methenamine Silver stain or the immunoperoxidase staining for alpha-l-antitrypsin and alpha-fetoprotein.

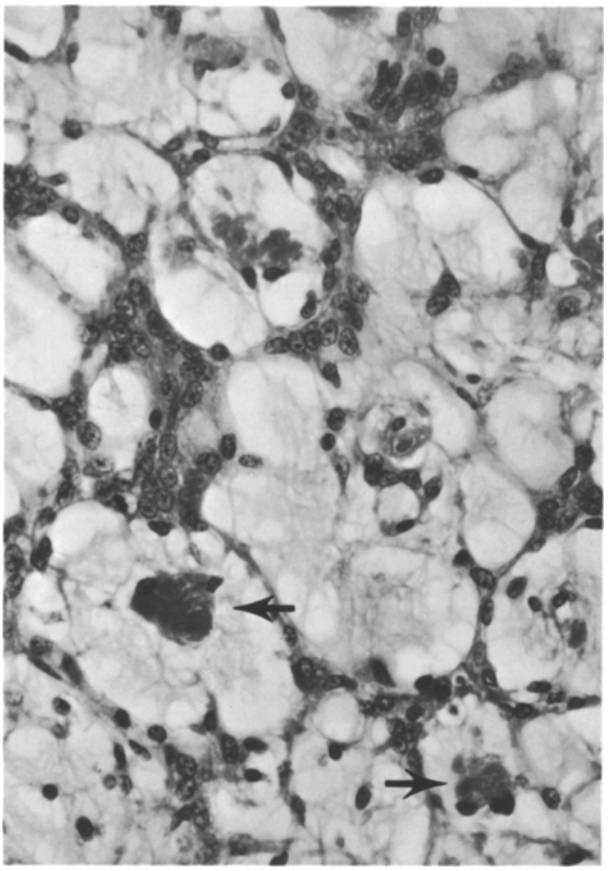

Fig. 3. A cellular area with a lacy network of cells and large extracellular spaces with frequent eosinophilic globules (*arrows*) (H & E, × 370)

The tumor was arranged in distinct lobules, some of which contained cells with a striking radial pattern of growth (Fig. 4). In other areas cells were in a spindled pattern, either solid or separated by stroma which yielded a lace-like pattern. No giant cells, cartilage, osteoid or calcification were seen. Margins of the tumor were infiltrating.

Ultrastructurally, the cells had irregularly shaped nuclei with chromatin marginated at the periphery and no nucleoli. The cytoplasm contained many polyribosomes, phagolysosomes, a few mitochondria, moderate amounts of rough endoplasmic reticulum (frequently dilated) and a finely fibrillar matrix (Fig. 5). Scattered lipid vacuoles were present, occasionally incorporated into lysosomes. There were long cytoplasmic processes with numerous tight junctions joining cells into a network (Fig. 6). In areas the processes coiled into a tight spiral.

Most cells were surrounded by a thick layer of flocculent basement membrane material, which revealed a well-defined basement membrane on perpendicular sections. The very extensive extracellular space contained large aggregates of fine fibrils arranged in bundles with sporadic condensations (Fig. 7). These

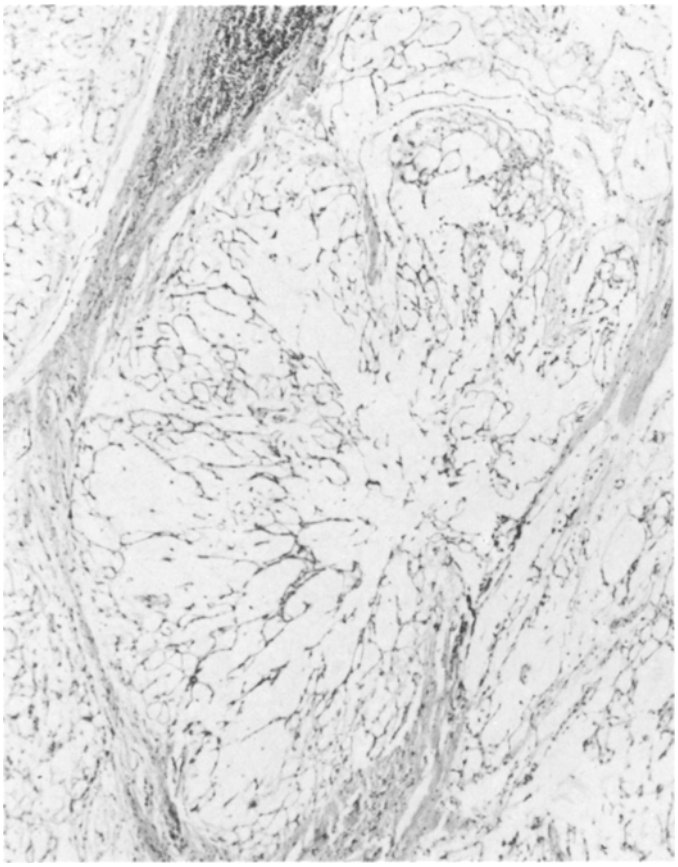

Fig. 4. There are prominent lobules with a strikingly radial growth pattern (H&E, ×41)

fibrils were much thinner than mature collagen fibers, which were also present in aggregates. They have the ultrastructural appearance of fibrin. They occasionally merged into the basement membrane material next to cells and were sometimes seen in the center of a group of cells without basement membrane. At times a bundle appeared to be within the cytoplasm of a tumor cell. These bundles are probably the ultrastructural correlate of the eosinophilic globules present on a light microscopic level.

Discussion

The tumor reported is unusual in several aspects. Although it is clearly of mesenchymal origin, the clinical and radiologic evidence indicate that it either arose within the bone or invaded it very early. This was one of the factors supporting the original diagnosis of a cartilaginous tumor, either chondromyxoid fibroma or chondrosarcoma.

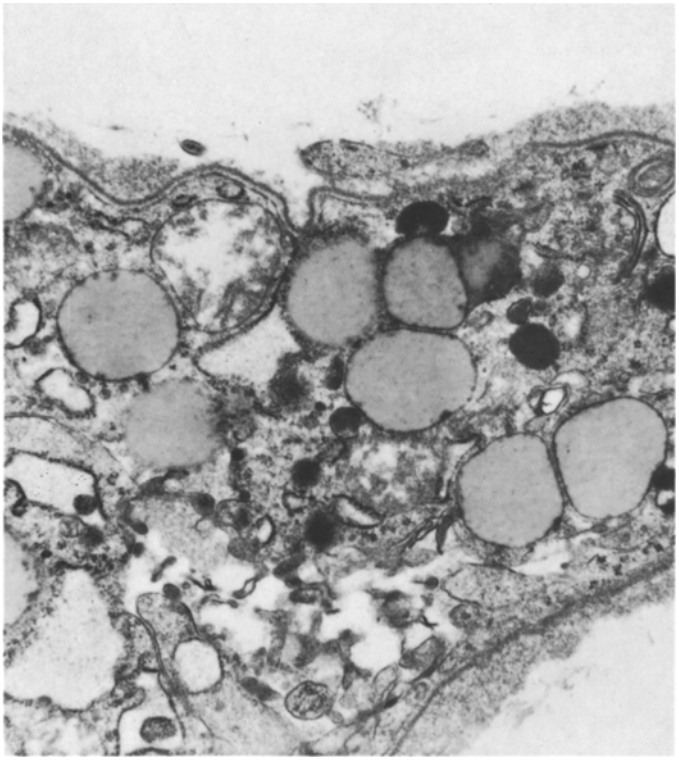

Fig. 5. Electron micrograph illustrating a portion of a cell. There are lipid vacuoles, one incorporated into a phagolysosome, lysosomes, polyribosomes, and dilated endoplasmic reticulum. A well defined basement membrane is apparent and is enmeshed in flocculent material ($\times$ 10,3)0)

Routine light microscopy showed no areas with identifiable chondrocytes within lacunae. Positive findings that are not common in chondroid tumors include the many vacuolated cells and frequent large eosinophilic bodies which stained positively for fibrin.

The light microscopic appearance was compatible with "chordoid sarcoma" particularly as described by Martin et al. (Martin, 1973). The tumor had distinct lobules, within some of which was a strikingly radial arrangement of cells with a lacy appearance (Figs. 3, 4). Several recent case reports have provided evidence that some "chordoid sarcomas" are of cartilaginous origin, and have suggested that all of them are variants of extraskeletal myxoid chondrosarcoma (Martin, 1973; Mehio and Ferenczy, 1978; Weiss, 1976). If it is accepted that the histochemical and ultrastructural findings in a case of chordoid sarcoma must indicate a cartilaginous origin, the current case is not a "chordoid sarcoma" despite fitting the original description of that entity as a soft tissue tumor, away from the midline, with the histologic appearance of chordoma (Stewart, 1948; Martin, 1973).

Histochemically, cartilage should contain Alcian blue, mucicarmine and colloidal iron positive material which is hyaluronidase resistant (Enzinger and

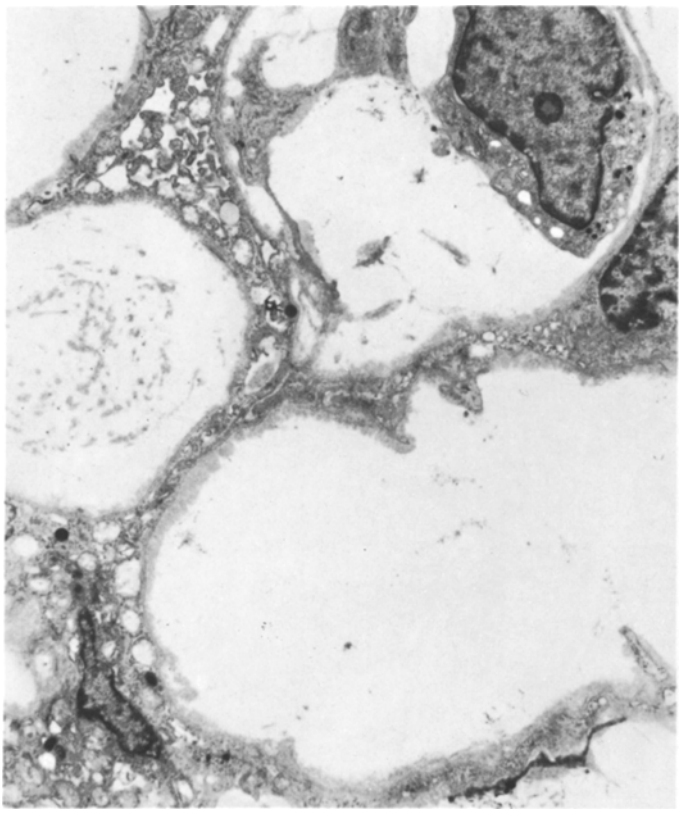

Fig. 6. Long cytoplasmic processes form a lacy network. The cells are surrounded by basement membrane and finely fibrillar material (×3,998)

Shiraki, 1972; Mehio and Ferenczy, 1978; Pearse, 1975). The presence of Alcian blue, colloidal iron and mucicarmine staining that is sensitive to hyaluronidase is similar to the findings of Kalderon and Fethiere (1973) in two liposarcomas and serves to distinguish the current case from chondrosarcoma. The negative oil red 0 stain in the current case is probably due to the low cell to stroma ratio and poor differentiation.

The ultrastructure of normal cartilage and cartilaginous tumors has been extensively studied (Hirohato and Marimoto, 1971; Anderson et al., 1963; Enzinger and Shiraki, 1972; Fu and Kay, 1974; Steiner, 1979). Chondroid cells typically have vesicular granular endoplasmic reticulum, intracytoplasmic glycogen, lipid bodies, and a collagen containing extracellular matrix with electron-dense granules. They may have a thick nuclear fibrous lamina, and rare tight junctions. Frequent junctions and prominent basement membrane have not been described. Although the above appearance is typical of cartilage, none of the findings are specific.

Ultrastructurally chordoma is described as having stellate cells with cell processes which spread along the processes of other cells and have moderate

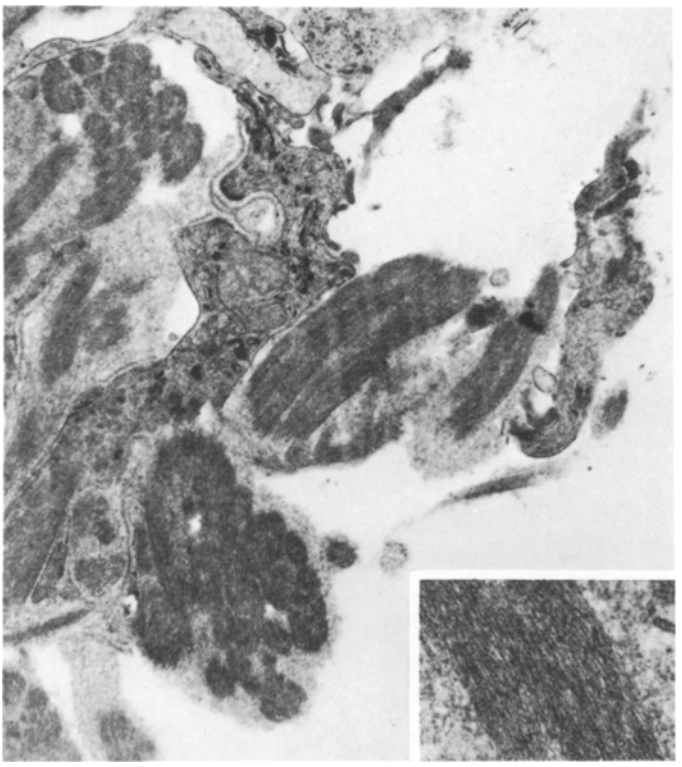

Fig. 7. Extracellular aggregates of fine fibrils (× 9,632). Inset detail of fibrils (× 19,870). This material is probably the correlate of the eosinophilic globules

to large numbers of vacuoles, rare desmosomes, pinocytosis and phagocytosis, cytoplasmic tonofilaments and no basement membrane (Spjut and Luse, 1964). Mitochondria stacked with rough endoplasmic reticulum may be present (Erlandson et al., 1968).

Recent reports on the ultrastructure of brown fat, hibernoma (Gould et al., 1976) and myxoid liposarcoma have emphasized the presence of desmosomes or gap junctions (Seemayer et al., 1975) at contact sites with other cells and well defined basement membrane. There are lipid vacuoles, many mitochondria, variable numbers of cytoplasmic and pinocytotic vacuoles, and a stroma with microfibrils and some collagen. Subplasmalemmal focal condensations may be prominent.

Although the current case has clinical and light microscopic features that are compatible with cartilaginous origin, the histochemical and ultrastructural findings favor fatty rather than cartilaginous differentiation. It has cytoplasm consistent with most mesenchymal tumors, with a scattering of mitochondria, some lipid vacuoles, and rough endoplasmic reticulum (Fig. 5), but lacks the thickened nuclear membrane and glycogen frequently found in cartilaginous tumors. The prominent basement membrane in the current case (Figs. 5, 6) is typically seen in brown fat but has not been described or illustrated in

cartilage or chordoma. The frequency of desmosomes is also more compatible with brown fat than cartilage. Although no definitive test exists for distinguishing one type of neoplastic mesenchymal cell from another, the weight of evidence suggests that the current case is of brown fat origin.

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Note Added in Proof

Since acceptance of this article a very similar tumor has been reported and the suggestion made that it was of synovial origin (Robertson, D., Hogg, G.: Chordoid sarcoma: Ultrastructural evidence supporting a synovial origin. Cancer **45**, 520–527 (1980)