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Chordoid sarcoma (extraskeletal myxoid chondrosarcoma)

A light and electron microscope study

Irving Dardick,[1] Réal Lgacé,[2] Maureen T. Carlier,[3] and Roland C. Jung[3]

[1] Canadian Tumour Reference Centre, Ottawa, Ontario,
[2] Department of Pathology, Hotel Dieu Hospital, Quebec,
[3] Department of Laboratory Medicine, Ottawa Civic Hospital, Ottawa, Ontario, Canada

Summary. Evaluation of a series of 12 chordoid sarcomas suggests that there is a wider range of histological features in this entity then previously appreciated. Six of the lesions had a typical tumor cell organization and a mixture of cellular and myxoid stromal components, while the remaining cases were atypical because of a more solid growth pattern. Four of the 12 cases, that included both typically myxoid and more cellular examples, had small foci with hyalinized stroma segragating individual or small groups of tumor cells with and without lacunar spaces. Two atypical cases revealed more extensive and obvious chondrocytic differentiation in recurrent or metastatic lesions and in one of these, the histological pattern was that of mesenchymal chondrosarcoma. Ultrastructural examination of three cases revealed fine structural features of both the tumor cell population and extracellular matrix compatible with chondrocytic differentiation.

Results of light and electron microscopy of this series of chordoid sarcoma add further support for categorizing this tumor with other malignant chondrocytic neoplasms. It is probable that chordoid sarcoma and extraskeletal myxoid chondrosarcoma represent the same entity and that this lesion has a close histogenetic relationship to mesenchymal chondrosarcoma.

Key words: Chordoid sarcoma – Chondrosarcoma – Electron microscopy

With its rare occurrence, particular histology and disputed histogenesis, the unusual and relatively uncommon soft tissue tumor, chordoid sarcoma, has stimulated considerable interest amongst pathologists since the designation and description of this neoplasm by Stewart (1948).

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Offprint requests to: I. Dardick, Canadian Tumour Reference Centre, Clinical Studies Unit Building, Ottawa Civic Hospital, 60 Ruskin Avenue, Ottawa, Ontario, Canada K1Y 4M9

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It is apparent from previous publications that the type of precursor tissue involved in the histogenesis of chordoid sarcoma is still not settled; a variety of histogenetic origins have been suggested including primitive mesenchyme (Martin et al. 1973), extramidline chordoid tissue (Dabska 1977), synovium (Robertson and Hogg 1980) and adipose (Bender et al. 1980) and chondrocytic (Enzinger and Shiraki 1972; Mehio and Ferenczy 1978; Pardo-Mindan et al. 1981; Smith et al. 1976) tissues. It is evident that the majority of these reports have suggested that a chondrocytic origin is more likely, although the exact nosologic position for chordoid sarcoma within the group of chondrosarcomas is unclear. In addition, the relationship of this entity to mesenchymal chondrosarcoma and, particularly, extraskeletal myxoid chondrosarcoma is also the subject of divergent opinion. Some authors suggest that chordoid sarcoma and extraskeletal myxoid chondrosarcoma represent the same entity (Allen 1980; Mehio and Ferenczy 1978; Pardo-Mindan et al. 1981), while others maintain they are distinct lesions (Hajdu 1979; Robertson and Hogg 1980). In the latter situation, however, no distinctive cytological or histological criteria have been established to distinguish these supposedly separate entities.

One of the difficulties in establishing the histogenesis of chordoid sarcoma has been the relatively undifferentiated features and organization of the tumor cells. In particular, there has been a lack of differentiation of hyaline cartilage in the majority of reported cases whether designated as chordoid sarcoma or extraskeletal myxoid chondrosarcoma. Tumor cells enclosed in lacunar-like spaces have been described in four reports (Enzinger and Shiraki 1972; Mehio and Ferenczy 1978; Smith et al. 1976; Tsuneyoshi et al. 1981), a few examples have had focal hyaline cartilage (Tsuneyoshi et al. 1981) and one case had a small focus of chondrosarcoma (Pardo-Mindan et al. 1981). Adding to the difficulties in classifying this lesion have been the absence of neoplasms with coexisting histological patterns of typical chordoid sarcoma and mesenchymal chondrosarcoma.

In reviewing a series of 12 cases of chordoid sarcoma (three of which had tissue available for electron microscopy), we have been impressed by the variation and multiplicity of morphological patterns in both primary and recurrent lesions. Features evident in some of these cases appear to add further support for a chondrogenetic derivation of chordoid sarcoma.

Methods and materials

Soft tissue tumors with histological features compatible with a diagnosis of chordoid sarcoma or extraskeletal myxoid chondrosarcoma were culled from the files of the Canadian Tumour Reference Centre, Ottawa, Canada and the Department of Pathology, Hotel Dieu Hospital, Quebec, Canada. In most cases, hematoxylin and eosin stained slides and paraffin blocks were available for review. In selected cases, one or more of the following special stains were also performed; periodic acid Schiff (with and without diastase), alcian blue (with and without hyaluronidase) at pH 1.0 and 2.5, mucicarmine, Masson's trichrome and Bielschowsky's reticulin.

Three cases had wet tissue, either formalin or glutaraldehyde fixed, available for ultrastructural examination. Small portions of these tissues were post-fixed with osmium tetroxide,

dehydrated in a graded series of alcohol and embedded in epon-araldite resin. Toluidine blue stained 1 μm sections were used to select appropriate areas for thin sectioning and the grids subsequently prepared were double stained with uranyl acetate and lead citrate. Sections were screened and photographed using either a Phillips 301 or Zeiss EM9A electron microscope.

Results

Clinical information

Table 1 provides details of clinical, therapeutic and follow-up data on all 12 cases. The ages range from 21 to 92 with a mean age of 46 years. There was a marked preponderance of females (3 to 1; female to male) and a predilection for the lower extremity (66%). One case occurred in an unusual location, the retroperitoneum adjacent to the right kidney.

Light microscopy

In all cases the tumors were circumscribed, and in some cases there was a complete or, at least, partial well-defined fibrous capsule. The majority of cases revealed a lobular growth pattern (Figs. 1 and 4) but in a few cases this characteristic was minimal.

In general, tumor cells were small with scanty amounts of acidophilic cytoplasm and round, oval or spindle-shaped nuclei (Fig. 2). There was considerable variation in cell shape and organization; patterns varied from single cells or small, loose clusters to short, linear, poorly anastomosing cords of polygonal (Fig. 2a) or spindle-shaped (Fig. 2b) tumor cells. Linear, radial arrangements or partially concentric arrays of cords or columns of cells were common peripherally; looser, more irregularly arranged cells separated by increased amounts of extracellular mucoid matrix were seen centrally in many lobules (Figs. 1 and 2a, b, d). Tumor cells could also be aligned in a more orderly arrangement of anastomosing cords of polygonal (Fig. 2c) or spindle and stellate cells (Fig. 2d). In the former case (Fig. 2c), pseudoacinar formations were common. Although one particular pattern usually predominated, it was not uncommon to note foci in which the basic cell type and organization varied.

During review of the histological features of this series of chordoid sarcomas, it became apparent that some examples contained focal areas in which tumor cells were compactly organized. In the more cellular areas of these tumors, the basic patterns described above persisted but cords and columns of cells were less separated by mucoid ground substance. This group was designated as having primarily myxoid histology (Table 1). Using a somewhat subjective evaluation, if a more cellular growth pattern predominated or at least accounted for 50% of available tissue sections, the case was designated as having a predominantly cellular histology (Table 1).

As an example of the lesions with cellular histology, Fig. 4a illustrates the increased cellularity of the variably sized and defined nodules separated by relatively acellular fibrous connective tissue strands. Although cellular

Table 1.

Cases	Age	Sex	Site	Size	Pre-op-duration	Treatment	Follow up	Result
A. *Primarily myxoid histology*								
1	60	M	Left popliteal fossa	$11 \times 8 \times 5$ cm	17 months	Amputation	19 years	Alive, no recurrence Squamous carcinoma, Vocal cord, May 1981
2	92	F	Metatarsal bone	Not given	Not given	Amputation	$4^1/_2$ years	Alive and well, November 1981
3	27	F	Right thigh	$10 \times 5 \times 4$ cm	4 weeks	Excision, radiation therapy to thigh and chemotherapy for lung metastases	4 years	Alive with complete remission, March 1982
4	30	F	Right inguino-femoral region	7 cm	6 days	Surgical excision	2 years	Alive and well, January 1982
5	75	M	Right popliteal fossa	Not given	2 months	Excision	None	–
6	32	F	Popliteal area	4 cm	7 months	Excision, recurrence recurrence at 2 years with amputation	6 years	Alive and well, January 1982
B. *Primarily cellular histology*								
7	55	F	Quadriceps tendon	$6 \times 5 \times 4$ cm	7 months	Cobalt therapy	15 months	Died, March 1963 Broncho-pneumonia
8	44	F	Left leg	4 cm	8 months	Radiotherapy to left groin. Resection left inguinal nodes. Partial excision recurrent tumor. Amputation	4 years	Died, 1977 No autopsy
9	40	M	Right deltoid muscle	$26 \times 15 \times 11$ cm	31 months	Forequarter amputation	43 months	Died; metastatic lesions lungs and lumbar vertebrae

Table 1 (continued)

Cases	Age	Sex	Site	Size	Pre-op-duration	Treatment	Follow up	Result
10	36	F	Left thigh	11 × 6 × 5 cm	8 weeks	Excision	1 year	Died, December 1978
11	21	F	Neck	4 × 2 × 2 cm	Few months	Surgical excision of mass	2 years	Alive, no recurrence October 1981
12	42	F	Retro-peritoneal	8 cm	Not stated	Excision; 6 recurrences in 5 years. (radiotherapy, 4,500 rads at 3rd recurrence)	6 years	Alive with residual tumor, September 1981. Laparotomy January 1981 for intestinal obstruction due to tumor

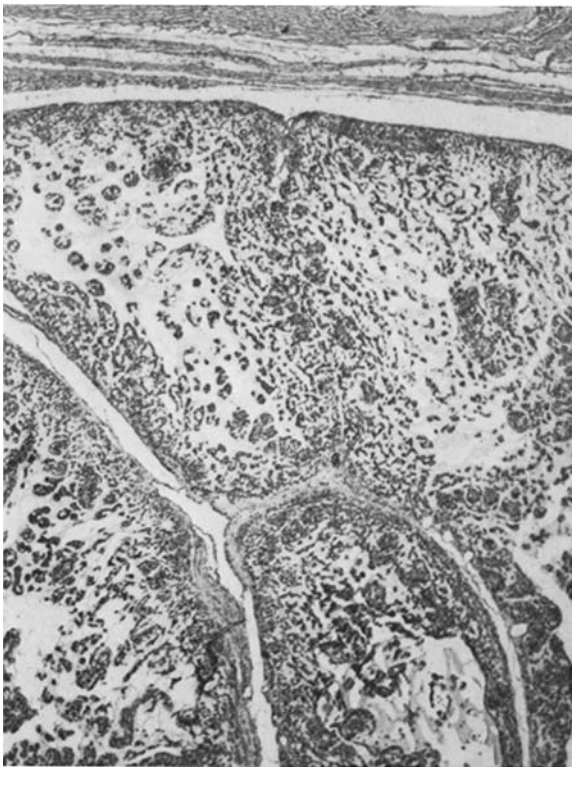

Fig. 1. Chordoid sarcoma with encapsulated, multiple nodules composed of short cords and small clusters of tumor cells increasingly separated centrally by mucinous stroma (× 30)

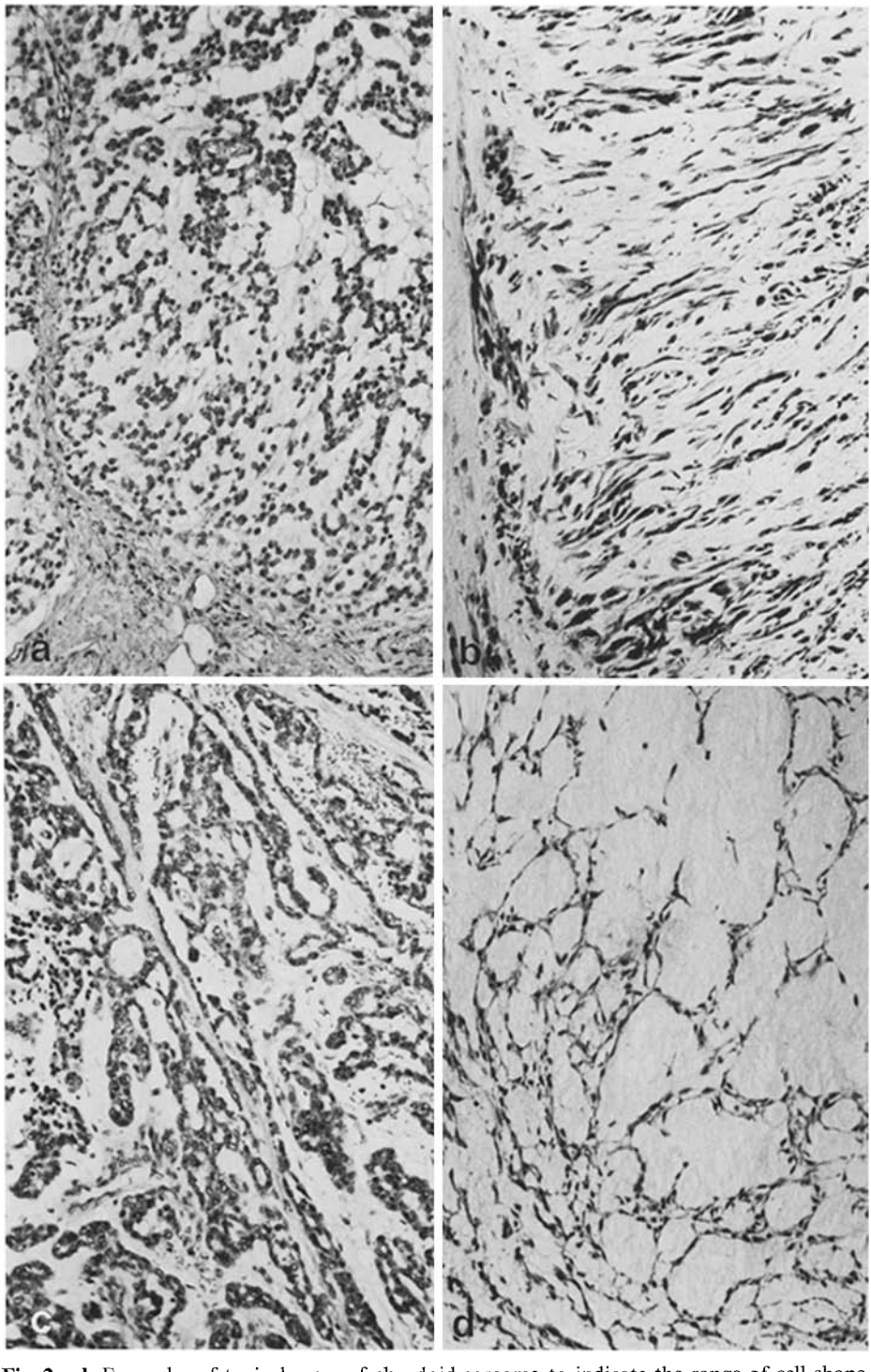

Fig. 2a–d. Examples of typical cases of chordoid sarcoma to indicate the range of cell shape and organization and the variation in amount and distribution of myxoid stroma. (**a**) Case 1. (**b**) Case 3. (**c**) Case 4. (**d**) Case 5 (All ×120)

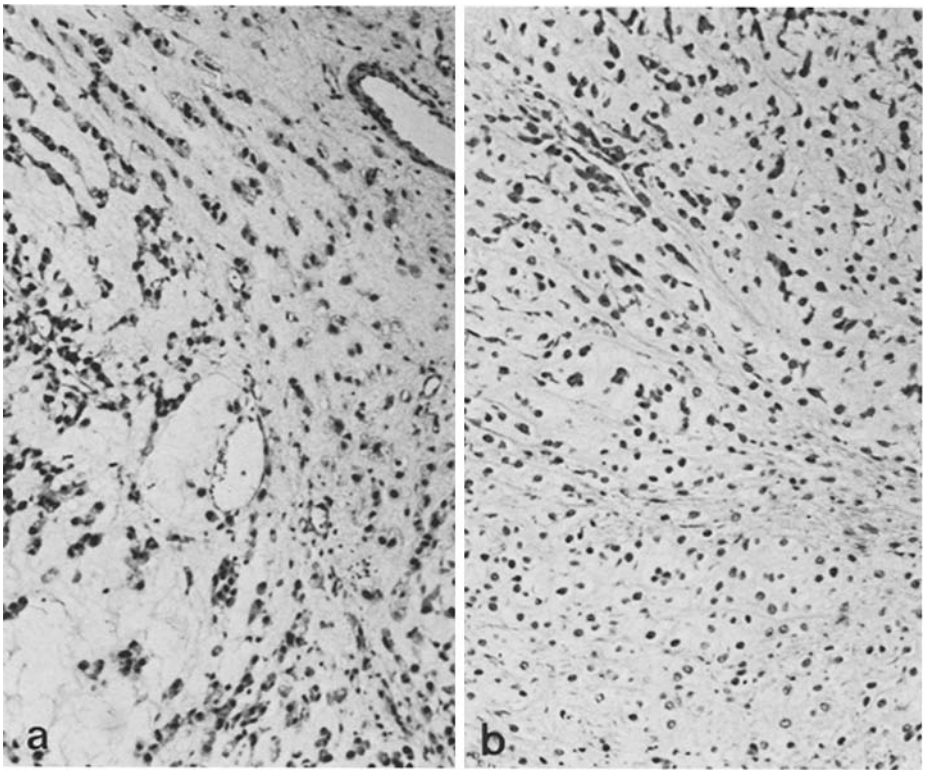

Fig. 3a, b. Examples of chordoid sarcoma with focal regions suggestive of chondrocytic differentiation. **a** Case 2. Hyaline stroma isolates single or small groups of tumor cells many of which are enclosed in a lacunar space. **b** Case 1. Region with histological features suggestive of immature cartilage. Note single, double or multiple cells within lacunae (**a** ×120), (**b** ×140)

areas often had tumor cells aligned in rows of minimally separated narrow columns, the more typical pattern of moderately dispersed, partially whorled, spindle and polygonal cells was also focally evident, particularly, at the periphery of lobules (Fig. 4b).

Two other cases in the cellular subgroup presented unusual findings. In one of these cases, the tumor at initial presentation was composed of cellular lobules often with a central vessel (Fig. 5a). Even in this low power micrograph, irregular anastomosing columns of tumor cells, often with a radial orientation, are apparent and produce a pattern compatible with chordoid sarcoma. Tumor cells in an inguinal lymph node metastasis, resected five months later, were compactly disposed in poorly defined lobules without evidence of the histological patterns evident in Fig. 5a. Both in the lymph node metastasis and a local recurrence partially excised two years later, tumor cells were slightly larger than in the original lesion and had moderate amounts of clear cytoplasm. In the case of the local recurrence (Fig. 5b), a striking development was the formation of multiple foci of chondrosarcoma within diffuse areas of compact dark staining cells. This

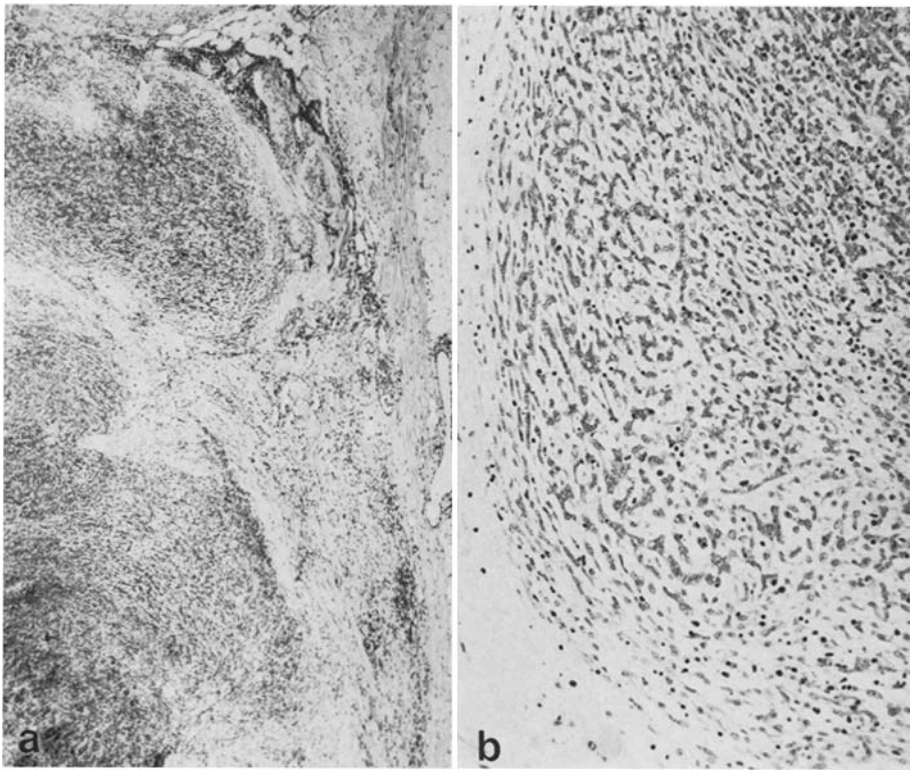

Fig. 4. Case 11. **a** A chordoid sarcoma in which tumor nodules have increased cellularity and a more homogeneous distribution of tumor cells. **b** Despite reduced extracellular materials, short columns of cuboidal and spindly tumor cells continue to have a somewhat lamellar and orderly arrangement, particularly, at the periphery of lobules (**a** ×30), (**b** ×120)

feature and the ability to trace the gradual differentiation of cartilaginous cells from undifferentiated cells at the margin of chondrocytic foci are characteristics of mesenchymal chondrosarcoma (Hajdu 1979). Residual tumor from the amputation specimen again had an undifferentiated character without evidence of chondroid differentiation. The second example was unique in that it originated in the retroperitoneum. The primary tumor was formed of lobules (Fig. 6a) containing nests of small tumor cells separated by minimal amounts of matrix which was alcian blue positive (pH 1.0) and partially sensitive to hyaluronidase indicating a content of hyaluronic acid and chondroitin sulphates. Samples of recurrent tumor resected a year later (Fig. 6b) continued to have a few scattered regions in which the lobules contained short cords and nests of tumor cells with slightly increased matrix. However, the bulk of the lesion (Fig. 6c) was formed of diffuse sheets of considerably separated tumor cells. Such cells were often binucleated and generally individually enclosed in a well delineated compartment producing a likeness to fetal cartilage. A subsequent recurrence (Fig. 6d) showed only histological features noted in the primary tumor (Fig. 6a).

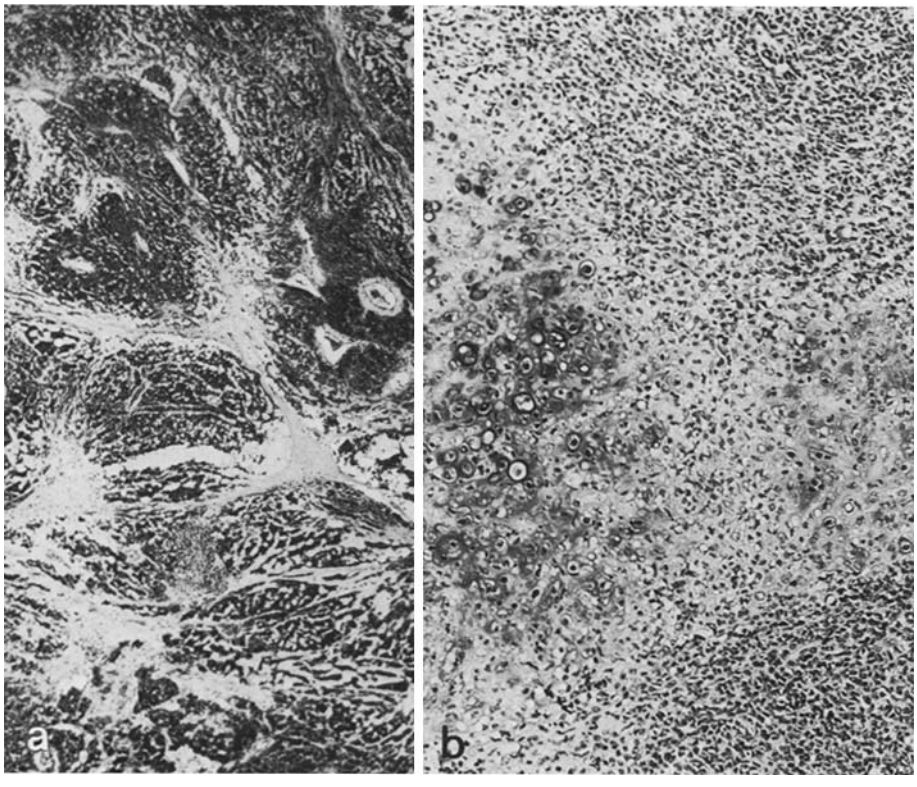

Fig. 5. Case 8. **a** An example of chordoid sarcoma with primarily cellular histology in which the primary lesion has numerous nodules composed of compact cells often with a centrally situated blood vessel. **b** A field from the local recurrence resected two years later shows diffuse areas of small, undifferentiated tumor cells gradually differentiating into malignant cartilage, i.e., mesenchymal chondrosarcoma (**a** ×30, **b** ×120)

In three of the six cases with myxoid histology and one case which was predominantly cellular, a careful search of sections revealed one or two foci with either increased amounts of hyaline, homogeneous intercellular matrix isolating occasional tumor cells in lacunae (Fig. 3a) or compact groups of tumor cells (Fig. 3b) in which single cells or small clusters of cells were enclosed in lacunar-like spaces. None of the predominantly myxoid cases showed evidence of fully developed hyaline cartilage. As detailed above an additional two cases with a predominantly cellular pattern displayed evidence of cartilaginous differentiation in recurrences. In all cases, stromal regions stained moderately to intensely with alcian blue at pH 2.5 and this histochemical result was reduced or abolished by pretreatment with hyaluronidase.

Electron microscopy

Among the three cases with material available for electron microscopic examination, two had been designated as typical examples (Cases 4 and 5)

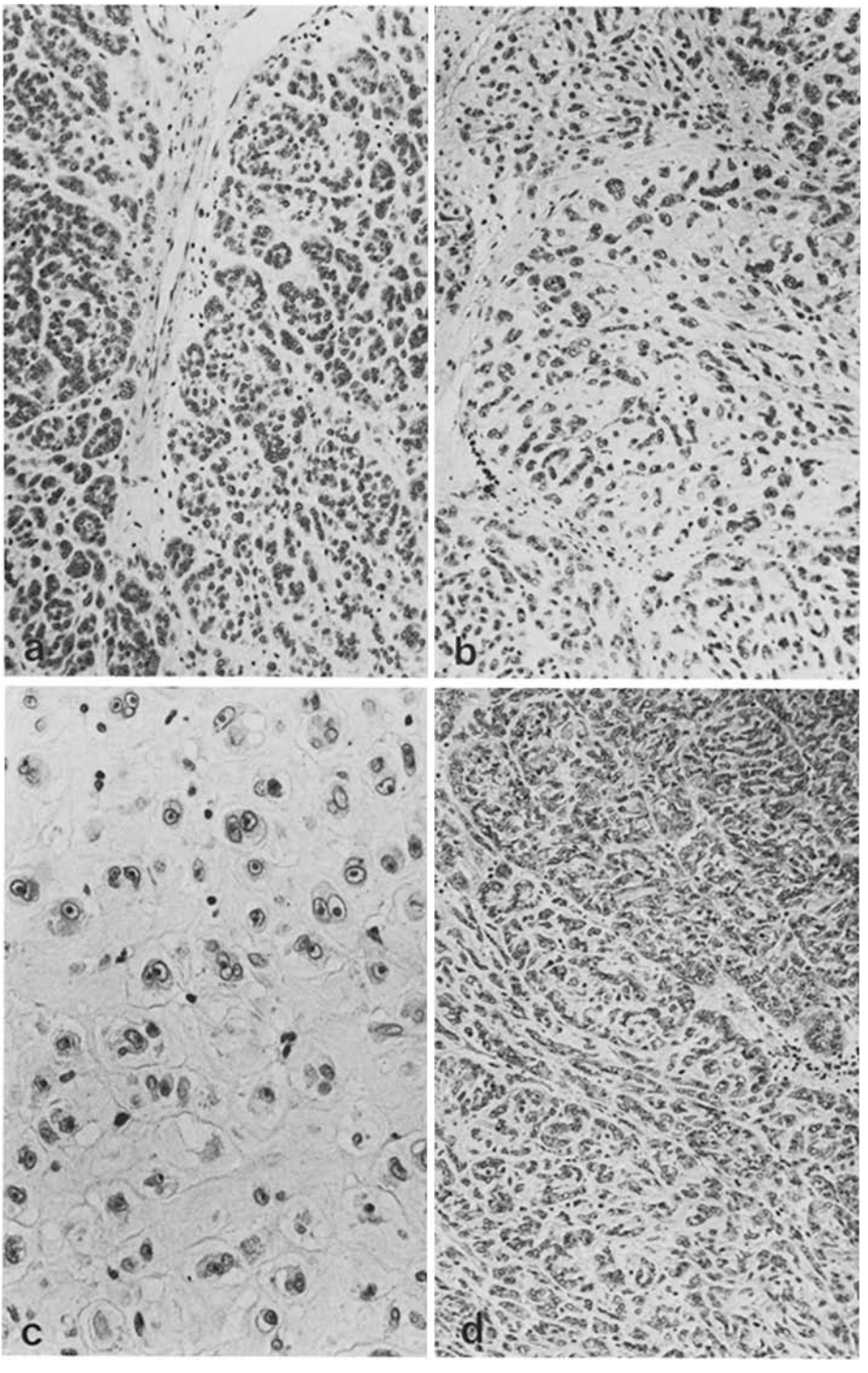

Fig. 6a–d. Case 12. A retroperitoneal chordoid sarcoma. **a** Lobules in the primary tumor are formed of nests of small tumor cells separated by minimal matrix. **b** Recurrent tumor resected one year later has scattered regions in which clusters and cords of cells are isolated by increased matrix. **c** The bulk of the above recurrence is formed by compartmentalized tumor cells, many of which are binucleated and partially surrounded by a clear space. **d** Intra-abdominal recurrent tumor, a further year later, has only histological patterns evident in the primary tumor. (**a, b, d** $\times 120$), (**c** $\times 300$)

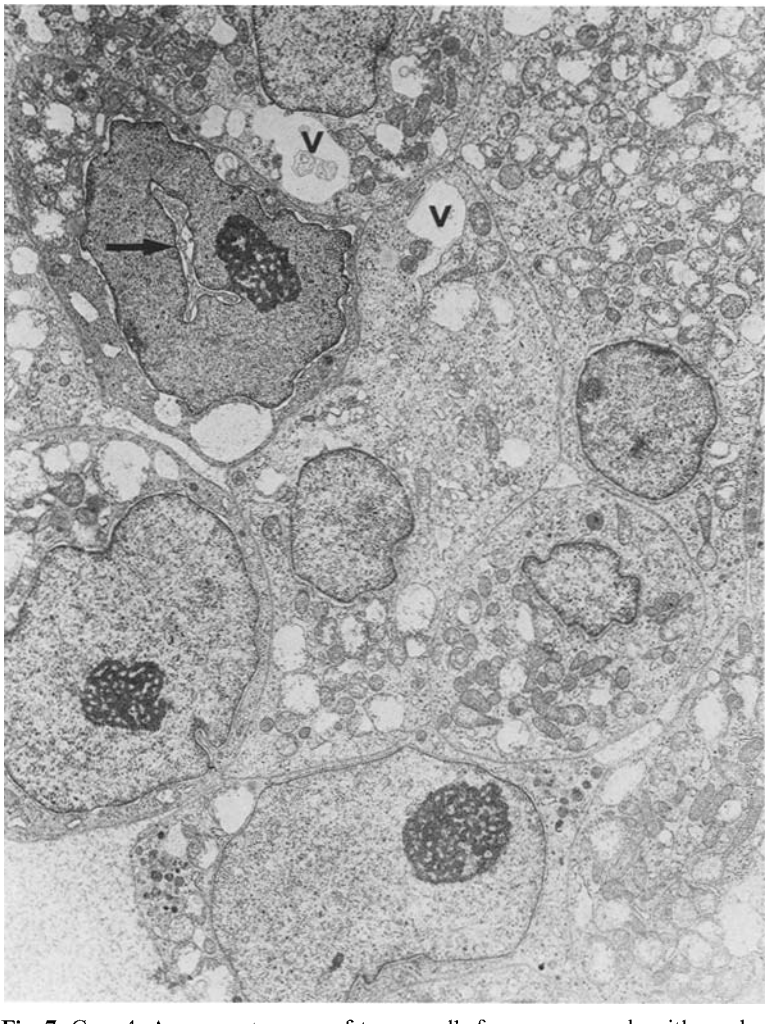

Fig. 7. Case 4. A compact group of tumor cells from an example with predominantly myxoid histology. Tumor cells have an irregular contour of the nuclear membrane with formation of pseudoinclusions (*arrow*) and a prominent nucleolus. The cytoplasm displays numerous mitochondria, polyribosomes and a few vacuoles (*V*) (× 4,200)

and one as an example of the more cellular type of chondroid sarcoma (Case 12) at the histopathological level. However, the ultrastructural findings of the tumor cells and tumor matrix are quite similar in the three cases examined and, therefore, will be described together.

The great majority of tumor cells contacted one another and were grouped either in small lobules, in sheets or in pseudoalveolar arrangements (Fig. 7). A limited number of tumor cells lay singly within the matrix. As observed by light microscopy, there is a considerable variation in cell shape with round, polygonal and spindle shaped tumor cells. The nuclear envelope had an irregular contour with indentations of variable depth that gave rise

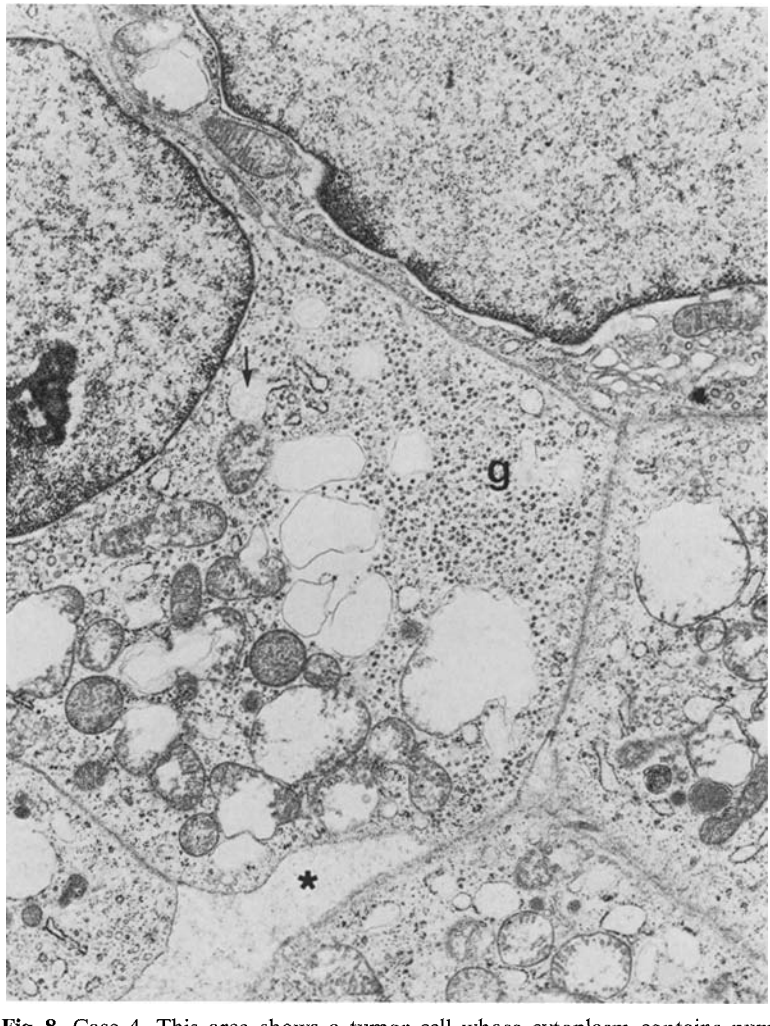

Fig. 8. Case 4. This area shows a tumor cell whose cytoplasm contains numerous glycogen particles (*g*) and artifactually ballooned mitochondria. Cisternae of rough endoplasmic reticulum (*arrows*) contain a finely granular product similar to the extracellular matrix (*asterisk*) (× 9,800)

to pseudonuclear inclusions (Fig. 7). Prominent nucleoli were seen in the vast majority of tumor cells. Chromatin was finely granular and uniformly dispersed within the nucleoplasm, although clumps of heterochromatin were present against the inner nuclear membrane in a few cells.

Areas of membrane specialization, such as micropinocytotic vesicles and rudimentary intercellular junctions were occasionally seen. Moderate to large numbers of mitochondria, a few of which contained intramatrical, paracrystalline structures and round osmiophilic inclusions, were frequently seen (Figs. 7 and 8). Free and bound ribosomes and focal aggregates of microfibrils were also observed. Rough endoplasmic reticulum occurred as

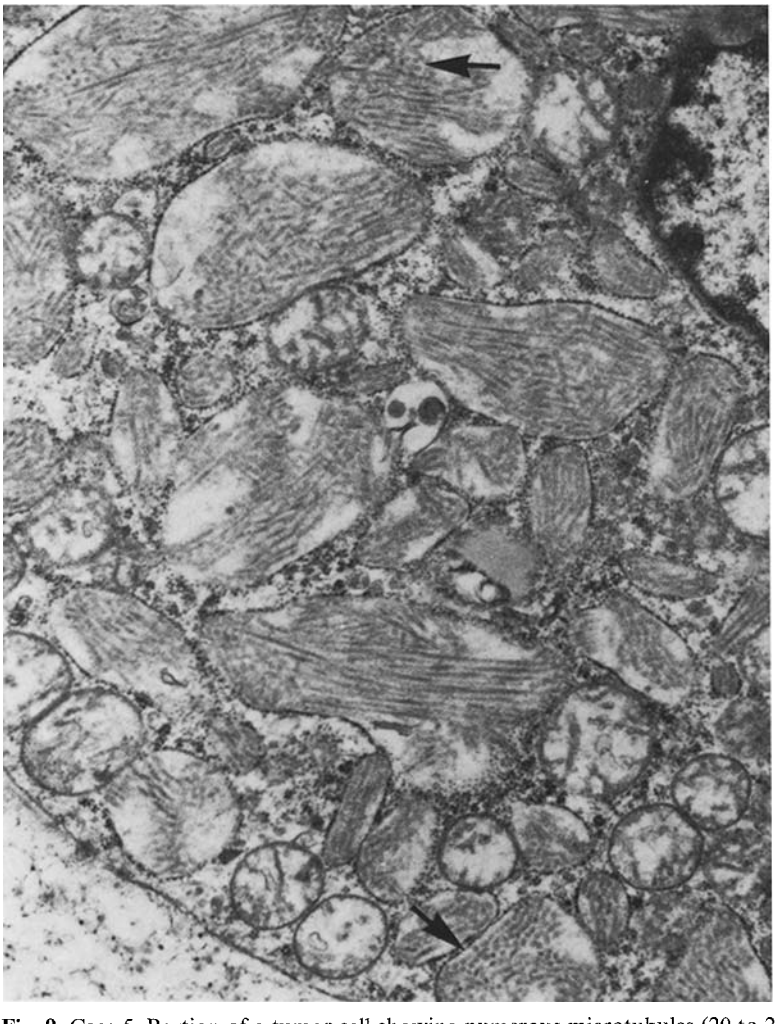

Fig. 9. Case 5. Portion of a tumor cell showing numerous microtubules (20 to 22 nm diameter) within the rough endoplasmic reticulum, some of which are cut in cross-section (*arrows*) (×19,000)

short segments, often dilated and containing a finely granular, weakly osmiophilic material similar to the granular part of the extracellular matrix (Fig. 8). Glycogenic particles were often grouped within the cytoplasm but do not show any particular relationship to other cellular components (Fig. 8). An unusual finding was the presence of numerous microtubules within the cisternae of the rough endoplasmic reticulum in one case (Fig. 9).

Ultrastructural features of the extracellular matrix were particularly interesting. The composition of the matrix varied greatly depending on the area examined. However, in all three cases, it was possible to demonstrate granular amorphous and granulofibrillar materials along with collagen and

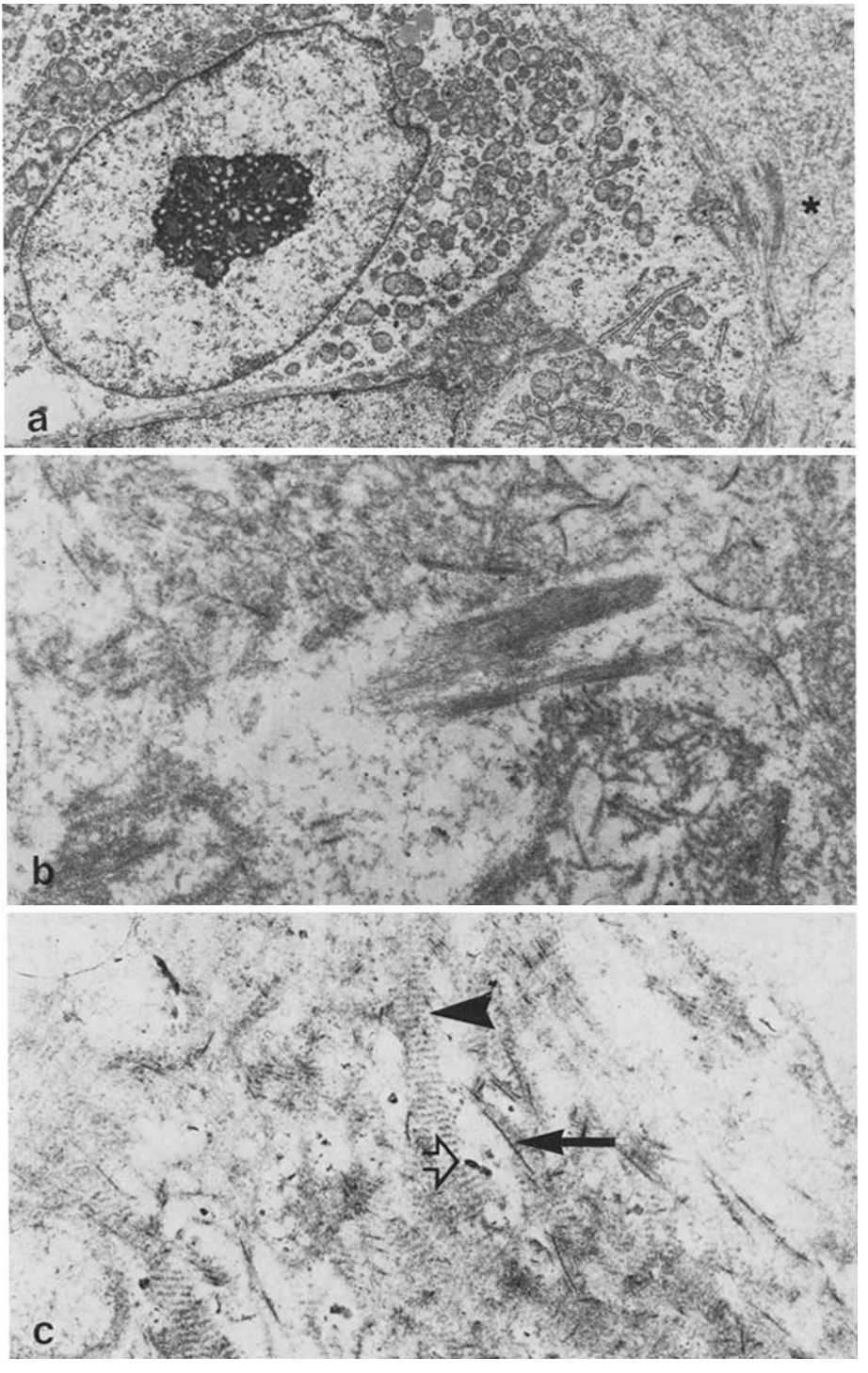

Fig. 10. **a** Case 12. Tumor cell margins are associated with clusters of microfibrils within the extracellular matrix (*asterisk*) (×4,200). **b** Case 4. Electron micrograph illustrating the general granulo-fibrillar nature of the matrix evident in all three cases examined ultrastructurally (×11,500). **c** Case 12. Matrix showing collagen fibers (*arrow*), broad band material corresponding to long-space collagen (*arrow-head*) and granular osmiophilic material (*open arrow*) (×11,500)

tropocollagen fibers (Fig. 10). Condensations of microfibrils varying from 50 to 500 Å in thickness were present along cell membranes and occasionally showed a tendency to encircle or separate tumor cells (Fig. 10a). Fine condensations of amorphous and rather osmiophilic granulofibrillar materials measuring about 100 to 300 Å in diameter are also interspersed between the fibrillar component (Fig. 10c). Relatively large fibers showing the typical 640 Å periodicity of collagen and broad-band material with a periodicity of about 1,000 Å, corresponding to long-spacing collagen, could be observed in all three cases examined ultrastructurally (Fig. 10).

Discussion

Due to its rare occurrence, a number of uncertainties concerning chordoid sarcoma continue to persist. These include its suggested chondrocytic histogenesis, the relationship of this tumor to extraskeletal myxoid chondrosarcoma and mesenchymal chondrosarcoma and an appreciation by pathologists of the range of histopathology consistent with this diagnostic category.

Although typical examples of chordoid sarcoma have rather characteristic morphological features, as the cases illustrated in this report indicate, there is considerable variation between tumors depending on the tumor cell arrangement and the amount and distribution of mucopolysaccharide matrix. However, histological patterns in chordoid sarcoma appear to be more complex than previously appreciated. In reviewing soft tissue tumors for possible inclusion in this study, six cases were found to have a considerable portion of the lesion formed by rather compact areas of relatively small, undifferentiated appearing tumor cells and limited amounts of mucoid and myxoid stroma. Many of these cases had presented considerable diagnostic problems and only careful reassessment revealed variable portions of the tumor with histological features consistent with chordoid sarcoma. The presence of a nodular growth pattern in the majority of these lesions and small, focal areas of chondrocytic differentiation frequently drew attention to this diagnostic possibility. Thus, the typical examples and, more importantly, the more cellular cases serve to expand the range of histopathology representative of chordoid sarcoma.

We share the opinion of a number of other authors (Allen 1980; Mehio and Ferenczy 1978; Pardo-Mindan et al. 1981; Tsuneyoshi et al. 1981; Weiss 1976) that chordoid sarcoma and extraskeletal myxoid chondrosarcoma are likely the same lesion. Similarities of clinical presentation, response to therapy and histopathology support this premise. Since there appears to be increasing support to classify this lesion with soft tissue tumors of cartilaginous derivation, perhaps the latter term is more appropriate. However, as is apparent in this report, a considerable proportion of these tumors are not predominantly myxoid. Despite this feature, even focal areas of typical histology and myxoid stroma in the more cellular variant of chordoid sarcoma indicate the potential for cartilaginous differentiation. Additional support for a chondroblastic origin for this tumor was evidenced during review of routine histological sections from the described cases. Although

usually not a prominent feature in most cases, 6 of the 12 cases contained foci indicative of some degree of chondroid, chondrocytic or chondrosarcomatous differentiation. This occurred in equal numbers of typical and atypical cases and varied in degree of differentiation from individual cells isolated in hyaline intercellular matrix, with or without lacunar spaces, to patterns reminiscent of fetal cartilage. No cases with myxoid histology exhibited fully developed hyaline cartilage but one example of the cellular variety contained multiple foci of well differentiated malignant cartilage in a local tumor recurrence. Tsuneyoshi et al. (1981) in reporting a series of 14 extraskeletal myxoid chondrosarcomas noted that all contained foci of chondroid differentiation and 4 cases had evidence of hyaline cartilage. The consistent histochemical finding that a proportion of the tumor glycosaminoglycan matrix consists of hyaluronic acid is further indication of the cartilaginous differentiation of chordoid sarcoma.

Previous studies of the ultrastructure of chordoid sarcoma (Mehio and Ferenczy 1978; Pardo-Mindan et al. 1981; Tsuneyoshi et al. 1981; Weiss 1976) also support the concept that this particular soft tissue neoplasm is either the same entity or at least represents a morphological variant of the tumor designated by Enzinger and Shiraki (1972) as extraskeletal myxoid chondrosarcoma. The ultrastructural findings of the cases examined in the present series support and reinforce this histogenetic concept. Electron microsopic features considered to be essential for the diagnosis of chondrocytic tumors, particularly the malignant ones, include the observation of numerous and often dilated portions of rough endoplasmic reticulum, whose content is often similar to a part of the matrix component, and the presence of lipid bodies and glycogen rosettes. In one case described in this report, numerous microtubules were noted within the rough endoplasmic reticulum and such a finding has also been reported in two cases of myxoid chondrosarcoma of soft tissues (Vernick et al. 1977; Wetzel and Reuhl 1980). However, this feature is not specific to tumors of cartilaginous origin (MacKay and Ayala 1980).

Although the above cytoplasmic fine structural features are suggestive of chondrocytic derivation for chordoid sarcoma, perhaps more significant indicators for such an origin are the tumor cell-matrix association and the quality of the intercellular matrix. Histochemical methods have demonstrated the acid mucopolysaccharide nature of the intercellular matrix with its predominance of hyaluronic acid and chondroitin sulphates (Martin et al. 1973; Mehio and Ferenczy 1978). Our electron microscopic observations, as well as those of others (Enzinger and Shiraki 1972; Fu and Kay 1974; Mehio and Ferenczy 1978; Pardo-Mindan et al. 1981; Smith et al. 1976; Steiner et al. 1973; Tsuneyoshi et al. 1981; Weiss 1976), strongly support these findings. The presence of a granular, poorly osmiophilic material and granular materials admixed with larger fibers such as collagen and long-spacing collagen are features characteristic of hyaline cartilage (Fu and Kay 1974; Ravel and Hay 1963; Schajowicz et al. 1974). Extensive sampling of our material available for electron microscopic examination established the chondroid nature of the matrix in all three cases. Even in regions of chordoid sarcomas with classical histological patterns and a predominantly

myxoid stroma, i.e., without frank evidence of chondrocytic differentiation, it was possible to demonstrate in varying degrees, at least one but more frequently two or three of the matrix components described above as indicative of cartilaginous development. Although not described in the case report of Mehio and Ferenczy (1978), long spacing collagen was observed in the case reported by Weiss (1976).

The relationship of chordoid sarcoma of soft tissues to mesenchymal chondrosarcoma remains an unsettled issue. It is noteworthy that Case 8 with histological patterns of chordoid sarcoma in the primary lesion had features diagnostic of mesenchymal chondrosarcoma in the local recurrence. It is significant that mesenchymal chondrosarcoma, in well differentiated cartilaginous areas, has similar ultrastructural characteristics both in the cellular and matrix components (Fu and Kay 1974; Steiner et al. 1973) to chordoid sarcoma and myxoid chondrosarcoma. In a case of mesenchymal chondrosarcoma of bone (Mandalenakis 1974), fibers with a periodicity of 217 Å and a resemblance to embryonal collagen have been described within endoplasmic reticulum. Using special techniques, this study (Mandalenakis 1974) also demonstrated intracellular mature collagen fibers and matrix-type glycoproteins within both the Golgi apparatus and the extracellular compartment. Such results suggest similar mechanisms of synthesis and secretion of matrix materials in chondrosarcomas, particularly when compared to autoradiographic studies in developing cartilage tissue (Leblond et al. 1955; Ravel and Hay 1963). Thus, both experimental and ultrastructural observations in myxoid chondrosarcoma, mesenchymal chondrosarcoma and chordoid sarcoma show many similar features suggesting a common histogenetic pattern for these three tumors.

Histological and ultrastructural evidence supporting a synovial (Hajdu 1979; Robertson and Hogg 1980) and a lipoblastic origin (Bender et al. 1980) for chordoid sarcoma have been reported. Although, it is neither our opinion nor apparently that of other authors, such reports suggest the possibility that chordoid sarcoma might represent a particular morphological pattern common to a variety of malignant soft tissue neoplasms. If one considers extraskeletal myxoid chondrosarcoma synonymous with chordoid sarcoma (Allen 1980), then comparison of clinical data in the present report with previous series (Allen 1980; Dabska 1977; Enzinger and Shiraki 1972; Martin et al. 1973) shows similarities of mean age and age distribution and predilection for the lower limb. But in contrast to the usual male preponderance (Allen 1980; Enzinger and Shiraki 1972), the current series showed a predilection for females. As is evident from the clinicopathological data in Table 1 and the study of Enzinger and Shiraki (1972), many patients with chordoid sarcoma survive for long periods of time even with local recurrence or metastases. Although the case series is too small for statistical analysis, it is noteworthy that examples of chordoid sarcoma with a more cellular type of histology appear to have a worse prognosis.

The tumor presenting in the retroperitoneum (Case 12) appears to be a very unusual occurrence with only one previous case having been reported intra-abdominally (Mehio and Ferenczy 1978). On the basis of its axial

distribution, chordoma had to be included in the differential diagnosis of this case. However, the absence of typical physaliferous cells and the ultrastructural features, particularly, the absence of intercellular lumen formation eliminated this diagnostic possibility (Erlandson et al. 1968; Lagacé and Delage 1975).

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