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Case Report

Chordoid Sarcoma of the Soft Tissue of the Nape of the Neck: A Case with a 20 Year Follow-Up

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Summary. A huge tumor mass arising from the soft tissue of the nape of the neck, which histologically resembled chordoma is reported. This case had a history of over 30 years after onset at around 37 years of age. With frequent recurrence and re-excision after en bloc radical resection, performed at 47 years of age, this female patient expired at the age of 66, 20 years after the first radical operation.

At operation, the mass was found to be firmly attached to the dorsal soft tissue. There was no connection with any bony structure, such as the cervical vertebrae or skull. Chordoma-like histological features were unchanged throughout the next 20 years. This unusual malignant neoplasm appears to correspond to the "chordoid tumor" or "chordoid sarcoma" of soft tissue, a term coined by F.W. Stewart in 1948. Alternatively the name "chondroid chordoma" used by Heffelfinger, Dahlin and others may be employed.

Key word: Chordoid sarcoma – Chordoid tumor of soft tissue.

This case was previously reported under the title of "chordoid tumor" arising in the nape of the neck in 1955 (Tanaka et al.). It has had a prolonged remission of 20 years duration after initial radical en bloc removal of a huge mass. The patient, a woman, died in 1975 at the age of 66. During her last 20 years of life, a complete follow-up study was carried out by repeated excision biopsies of the recurrent tumor. This was a unique soft tissue tumor, histologically resembling chordoma, and the case appears to fall within the category of "chordiod tumor", a term originally designated by F.W. Stewart (1948). The close histological resemblance between chordoma and chondrosarcoma has been discussed, and Heffelfinger and Dahlin et al. (1973) have coined the term "chondroid chordoma" for this lesion.

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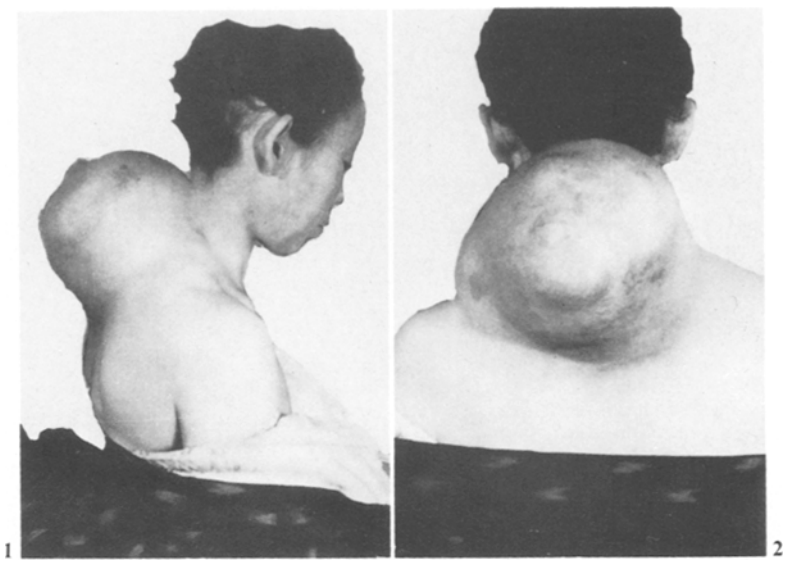

Fig. 1. A huge mass at the nape of the neck. At the first admission 20 years prior to death. Lateral view

Fig. 2. Posterior view

Case Report

A Japanese farm wife noticed a tumor measuring 1.5 cm in diameter in the subcutaneous tissue in the mid-line of the nape of her neck at the age of 37 (in 1944). It gradually grew larger during the following 10 years, reaching the size of child's head by 1954. There were no subjective symptoms. When the patient was admitted to a hospital in 1954, a huge firm hemispherical mass was observed (Figs. 1 and 2), measuring 50 cm in circumference at the tumor base. The overlying skin was freely movable, and the broad base of the tumor did not appear to be fixed to deeper structures. X-ray examination showed no pathological findings in the vertebral or other bony structures in the neck and skull. No calcified foci were detected in this mass.

The First Operation — April 1954. The mass was completely excised en bloc with abundant subjacent soft tissue, trapezius muscle and a sufficient elliptical skin flap. At operation it was confirmed that the mass was unattached to deeper structures.

The patient's progress was uneventful until 1970, when she first recognized a recurrent nodule in the left cervical region. The patient was hospitalized in September 1972, when elastic firm masses of various sizes, measuring up to 4 cm in greater diameter, were palpable on both sides of the neck and in the left axilla. To these tumor masses telecobalt radiation therapy was given, total dose of 5,400 rad each, between October and December 1972. The size of the tumors did not appear to alter greatly.

The Second Operation – November 1972. Excision biopsy of the left axillary mass was carried out. This measured 3.5 cm at its greatest diameter.

By January 1973, further nodules had appeared on both sides of the neck, where they formed a conglomerate mass. By September 1973, the site of the first operation, the right suprascapular region, had begun to enlarge due to tumor recurrence and a mass of 7.5×5.5 cm was present.

The Third Operation – October 1973. The tumor in the right suprascapular region was excised. A large mass measuring $5 \times 4 \times 3$ cm was removed.

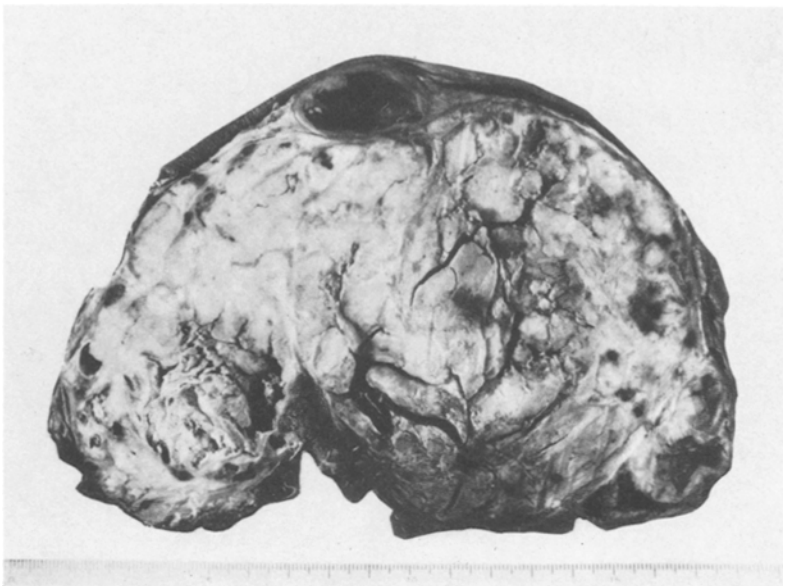

Fig. 3. The mass measuring 20 × 19 × 15 cm. and weighing 1,825 g. Greater part of tumor consisted of mucoid gelatinous neoplastic tissue, with intermingled medullary and firm solid areas. Small cystic and slit-like spaces were present

The Fourth Operation – February 1974. Recurrent tumor masses were removed from the same area as the third operation. They comprise 3 masses each measuring 3.5 cm in diameter.

The patient died from the effects of pulmonary metastasis in March 1975 at the age of 66, approximately 30 years after the onset of the disease and 20 years after the initial radical operation. Permission for autopsy could not be obtained.

Pathological Findings

The Surgical Specimen from the First Operation. A huge hemispherical mass measured 20 × 19 × 15 cm and weighed 1,825 g. It was well defined by dense fibrous tissue and surrounded by muscle. The overlying skin flap was not involved by tumor tissue. Cut section revealed a well encapsulated neoplasm having a close connection with the fascia. The tumor parenchyma was irregularly divided into areas with a different appearance by fibrous trabeculae – the greater part consisted of soft grayish and/or whitish gray gelatinous areas, with some firm elastic and some medullary areas. There were irregular-shaped slit-like and small cystic spaces with friable intraluminar and mucoid tissue. Focal areas of old and recent hemorrhages were present. No bony structure was detected (Fig. 3).

Histological findings: Over 30 blocks were sectioned from various portions. Besides routine methods, PAS, mucicarime, Best' carmine, toluidin blue metachromasia, fat stains, silver impregnation, etc., were employed. Histological appearance could be roughly divided into 3 variants.

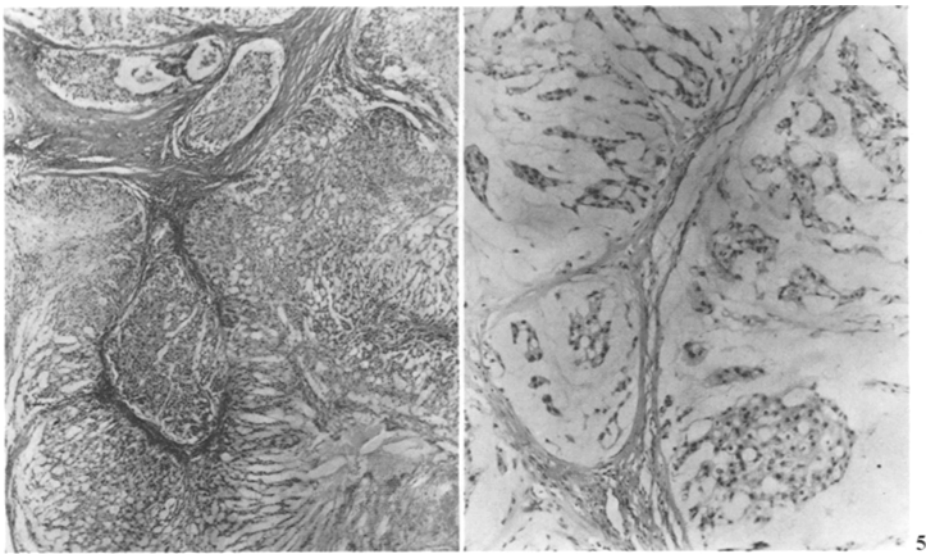

Fig. 4. Pseudoepithelial trabecular arrangement of cells in abundant mucoid matrix. Radially arranged neoplastic cells form septae. (H & E, ×40)

Fig. 5. Chordoma-like feature of neoplastic tissue and foamy vacuolar cells resembling physaliphorous cells. (H & E, ×100)

Area A) The largest area, which corresponded to the area with a myxoid appearance on gross inspection, consisted of an epithelioid trabecular arrangement of round and/or polyhedral cells of various size, often forming narrow pseudoalveolar structures in an abundant mucoid and/or chondro-myxoid matrix. They were radially arranged from the fibrous capsule or septum and showed a tendency to transition into a myxomatous structure composed of spindle- or stellate-shaped cells in the central portion (Fig. 4). These cells had acidophilic cytoplasm with abundant PAS positive granules (glycogen) and/or foamy vacuolar cytoplasm, resembling physaliphorous cells (Fig. 5). In this type of area, neoplastic cells showed occasional angiomatoid arrangements in the myxomatous matrix.

Area B) Occasional solid nests, composed of relatively large polyhedral cells of various size were packed between fibrous septae. Some fine reticulin fibrils formed a network in the intercellular spaces. These epithelial-like cells were sometimes very large, reaching 100 μ in diameter and contained a single pyknotic nucleus containing glycogen granules located in the center of the foamy clear cytoplasm (Fig. 6). They had the appearance of the physaliphorous element of a chordoma. These areas blended into those with myxomatous cobweb patterns with abundant chondromyxoid or mucoid matrix, compatible with Area A. Multinucleated giant cells with acidophilic cytoplasm, containing abundant glycogen were occasionally observed.

Area C) In some portions, the neoplastic tissue was acellular and hyalinated. Signet ring type foamy cells were embedded in fibrohyalinous tissue with positive

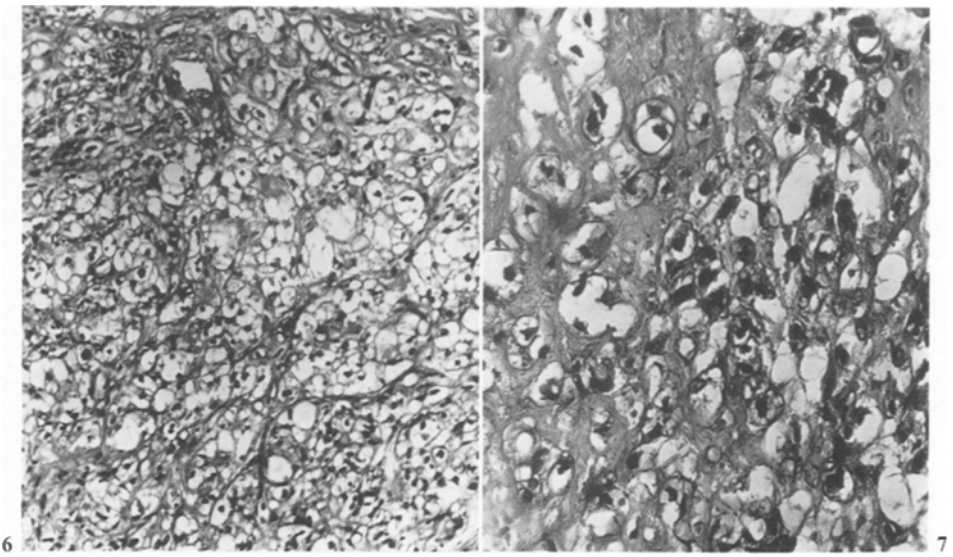

Fig. 6. In the medullary areas, cells are densely arranged and have large and foamy cytoplasm, resembling physaliphorous cells (H & E, ×200)

Fig. 7. Pleomorphic cells have foamy and/or acidophilic cytoplasm. Matrix appears chondomyxoid. (H & E, ×400)

mucicarmine staining and toluidin blue metachromasia below pH 2.6. No definite cartilagenous nor bony structure was detectable.

Surgical Material from the Second Operation. This had multiple cystic structures on the cut surface, into which there were gelatinous and friable neoplastic tissues. Histological examination revealed finding similar to that from the first excision, resembling that seen in Area A characterized by pseudoepithelial structures of spindle- and/or polyhedral-shaped cells in an abundant mucoid matrix. The pseudoglandular arrangement of the cells was reminiscent of an angiomatous or synovial appearance.

Surgical Material from the Third Operation. For all but a few small areas, the tumors were completely encapsulated and showed similar findings to the previous specimens. Histological examination revealed tissues, which were almost identical to that of the chondromyxoid Area A in the first surgical specimen (Fig. 7), resembling that seen in the chondrosarcomatous variant of chordoma.

The Surgical Specimen from the Fourth Operation. Cut section revealed lobulated structures with a focal gelatinous and/or myxomatous appearance. Histological examination revealed pseudoepithelial arrangement of cells forming narrow strands in an abundant mucoid and/or chondromyxoid matrix, resembling the so-called mixed tumor of salivary gland type or chordoma.

In all the surgical specimens, the neoplastic cells showed the characteristics

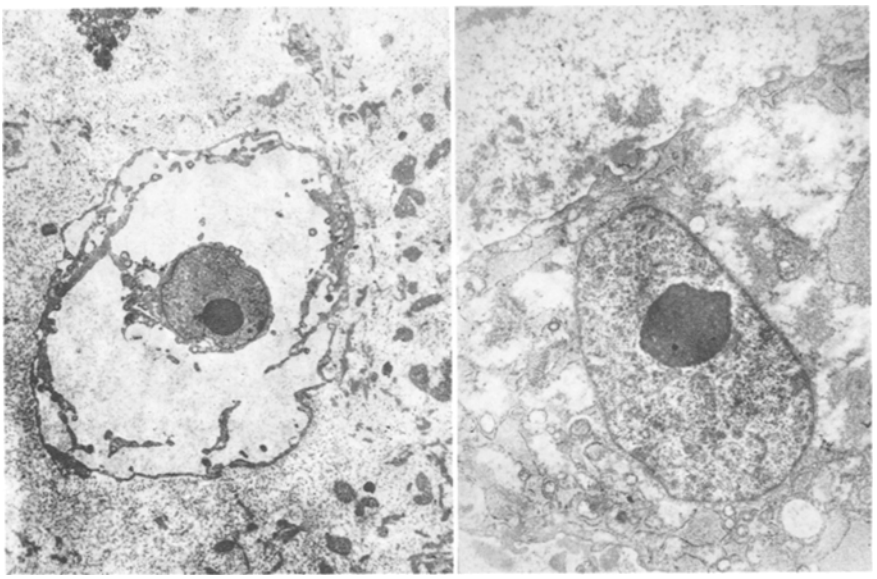

Fig. 8. Electron microphotograph was taken from the formalin-fixed specimen of the fourth operation. Ground substance is finely granular or amorphous. The cells have large and cystic structures, probably of dilated endoplasmic reticulum. Their features resemble chondroblasts. (× 35,000)

of physaliphorous cells with considerable numbers of glycogen granules. The abundant myxoid and/or chondromyxoid ground substance showed alcian blue positivity and was occasionally positive for toluidin blue metachromasia. There were scanty glycogen granules.

Electron microscopic examination was carried out on the fourth surgical specimen. Blocks were taken from the myxomatous area after buffered-formol fixation. On electron microscopy the matrix is seen to be finely granular or amorphous. Few fibrils were detected. Within the cells there are cystic spaces, probably representing dilated endoplasmic reticulum. A single distinct large nucleolus is observed, and in general the appearances resembled those seen in chondroblasts.

Discussion

The variability of histological features of mesenchymal neoplasms is sometimes extensive. Certain skin appendage tumors, e.g. chondroid syringoma, mixed tumor of salivary gland type, etc. (Stout and Gorman, 1959) may have a mixed appearance with chondroid and epithelioid areas. However, the mass in the present case was deep to the skin and no real epithelial component was observed. Soft-part chondrosarcoma, tenosynovial chondromatosis, mesenchymal chondrosarcoma of the soft parts, or chondromyxoid fibroma (Lichtenstein and Bernstein 1949; Stout and Verner 1953; Goldman 1967; Enzinger 1972) often show features similar to the present case. We are also aware of the variety

of tumors arising in connection with such structures as fascia, tendon or tendon-sheath, and synovia. Our first impression of the present case was that it represented a chordoma, because of the distinctive histological appearances and because the histological versatility of notochordal tumor may be extensive (Dahlin and MacCarty, 1952).

Heffelfinger and Dahlin et al. (1973) have coined the term "chondroid chordoma" for this type of lesion due to the striking histological similarity of chordoma and cartilagenous tumors. The topographical findings in this case are too uncertain to enable us to identify it readily was a variant of chordoma, in spite of its striking histological similarity. There have been reports of chordoma originating in the cervical region (Mabrey, 1935; Toole et al., 1961; Firkins, 1964; Higinbotham, 1967; Bach, 1970; Filbert, 1974) without any close connection to the vertebral column. Mabrey (1935) collected 150 cases of chordomas from literature of which 7 had occurred in the neck; Owen et al. (1932, cited from Mabrey, 1935) reported 8 cases of chordoma arising from the same region. Higinbotham et al. (1967) accumulated 46 cases of chordoma during 35 years at the New York Memorial Hospital, 5 cases being from the neck and one from the scapula. Thus, histopathological diagnosis may be particularly difficult when this kind of tumor arises in bone or soft tissue remote from the notochord (Ackerman et al., 1974; Mindan et al., 1975) as in the present case and in previously reported cases (Martin et al., 1973) originating from the mandible and maxilla.

In 1948, Fred W. Stewart (1948) coined the term "chordoid tumor" or "chordoid sarcoma" for these soft tissue neoplastic lesions. This term appears to be fitting descriptive designation for the present case, which morphologically resembles chordoma but which has no connection to the notochord. Weiss (1976) has examined his case of chordoid tumor of the flank ultrastructurally and concluded that chordoid tumor might be a variant of extraskeletal myxoid chondrosarcoma. Martin et al. (1973) pointed out that the histochemical data do not support a definite cartilagenous origin for the chordoid tumor, nevertheless, on the basis of electron microscopic findings we suppose its origin to be an immature chondroblastic mesenchymal cell.

Reference

Ackerman, A.B., Koch, K., Pardo, V.: Chordoma-like subcutaneous nodules in an infant. Arch. Dermatol. **109**, 247–250 (1974)

Bach, S.T.: Cervical chordoma. Report of a case and a brief review of the literature. Acta Otolaryngology **69**, 450–456 (1970)

Dahlin, D.C., MacCarty, C.S.: Chordoma – A study of 59 cases. Cancer **5**, 1170–1178 (1952)

Enzinger, F.M.: Clear cell sarcoma of tendon and aponeuroses. An analysis of 21 cases. Cancer **18**, 1163–1174 (1965)

Enzinger, F.M.: Extraskeletal myxoid chondrosarcoma: An analysis of 34 cases. Hum. Pathol. **3**, 421–435 (1972)

Filbert, C.G.: Chordoma of the cervical spine: A case report with a brief review of the literature. J. AOA **73**, 836–842 (1974)

Firkins, R.T.: Cervical chordoma. Arch. Otolaryngology **79**, 609–612 (1964)

Goldman, R.L.: Mesenchymal chondrosarcoma, a rare malignant chondroid tumor usually primary in bone. Report of a case arising in extraskeletal soft tissue. Cancer **20**, 1494–1498 (1967)

Heffelfinger, M.J., Dahlin, D.C., MacCarty, C.S., Beabout, J.W.: Chondromas and cartilaginous tumors at the skull base. Cancer **32**, 410–420 (1973)

Higinbotham, N.L., Phillips, R.F., Farr, H.W., Hustu, H.O.: Chordoma; Thirty-five year study at Memorial Hospital. Cancer **20**, 1841–1850 (1967)

Lichtenstein, L., Bernstein, D.: Unusual benign and malignant chondroid tumors of bone. A survey of some mesenchymal cartilage tumors and malignant chondroblastic tumor, including a few multicentric ones as well as many atypical benign chondroblastomas and chondromyxoid fibromas. Cancer **12**, 1142–1157 (1949)

Mabrey, R.E.: Chordoma – A study of 150 cases. Am. J. Cancer **25**, 501– 517 (1935)

Martin, R.F., Melnick, P.J., Warner, N.E., Terry, R., Bullock, W.K., Schwinn, C.P.: Chordoid sarcoma. Am. J. Clin. Pathol. **59**, 623–635 (1973)

Mindan, F.J.P., Canadell, J.M., Herranz, P., Imizcoz, J.L., Vazquez, yJ.J.: Sarcoma cordoide de fémur, Estudio a microscopio óptico y electronico de un case. Rev. Med. Univ. Navarra **XIX**, 133–141 (1975)

Stewart, F.W.: Case 211 Division of Laboratories and Research. Albany New York, 1948 (cited by Martin et al.)

Stewart, F.W.: personal communication (1955)

Stout, A.P., Verner, F.W.: Chondrosarcoma of the extraskeletal soft tissue. Cancer **6**, 581–590 (1953)

Stout, A.P., Gorman, I.G.: Mixed tumors of skin of the salivary gland type. Cancer **12**, 537–543 (1959)

Tanaka, N., Chen, W.C., Oota, K., Kuroda, H.: A huge unusual chordoid tumor of the nape arising in the soft tissue. Jap. J. Cancer Clin. **1**, 480–486 (1955)

Tanaka, N., Chen Wi-cha.: A giant chordoma-like tumor the subcutaneous soft part of the nape. Gunn **46**, 233–236 (1955)

Toole, H., Ioannovich, D., Papadimitriou, D.: Chordoma of the cervical spine. Arch. Otolaryngology **74**, 707–710 (1961)

Weiss, S.W.: Ultrastructure of the so-called chordoid sarcoma. Evidence supporting cartilagenous differentiation. Cancer **37**, 300–306 (1976)

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