Mesenchymal chondrosarcomas of the head and neck

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MESENCHYMAL chondrosarcoma is a distinctive type of chondrosarcoma that exists in both skeletal and extra-skeletal forms (Salvador et ah, 1971; Guccion et ah, 1973). The neoplasm rarely affects tubular bones and has a predilection for the facial bones and ribs. Over one-third of the reported examples have been in soft tissues and here too the region of the head and neck (principally the orbit) is typically a site of involvement.

In this report we present two additional examples of mesenchymal chondrosarcoma of the head and neck. Both neoplasms involved the paranasal sinuses, one apparently of osseous origin, the other from sinus soft tissues.

Report of cases

Case 1.

An 18-year-old male presented to his dentist with a two-month history of loosening of two upper molar teeth accompanied by a progressive and painless swelling of his right cheek. The affected teeth were removed and the alveolar ridge was biopsied. Tissue from the latter was interpreted as a hemangiopericytoma. The patient was then referred to the University of Michigan Hospital. Examination there revealed a swelling of the right cheek with a red, hemorrhagic mass in the right lateral nasal cavity, superior to and medi ally displacing the right posterior inferior turbinate. The right upper alveolar mucosa at the site of dental extraction was swollen, with visible tumour extension into the right gingival-buccal sulcus and right hard palate. Tumor was palpable in the inferior right orbit and the right eye was displaced upward. The remainder of the physical examination was unremarkable.

Paranasal sinus roentgenograms demonstrated a large mass filling the right maxillary antrum with extension into the right nasal cavity and ethmoid air cells (Fig. 1). Destruction of the bones of the anterior, medial and inferior walls of the antrum and right inferior orbital rim was noted. X-ray examination of the chest was negative.

Transoral biopsy of the right antral mass was performed and again a diagnosis of hemangiopericytoma was made. Following this, he received 6043 rads (Cobalt 60) over a 43-day period to the right anterior and oblique facial areas. During the time of irradiation a visible decrease in the size of the tumour was achieved, but at a one-month post-therapy visit, tumour remained in the right maxillary antrum and extended into the nasopharynx. Paranasal sinus x-ray

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FIG. 1
X-ray appearance of the mesenchymal chondrosarcoma involving the right maxillary sinus and nasal cavity of the patient in Case 1.

examination demonstrated a progressive destruction of the bones of the lateral and medial walls of the antrum and the floor of the orbit.

A right radical maxillectomy and orbital exenteration were performed. The margins of excision were free of a neoplasm that measured $6 \times 5 \times 4$ cm and apparently originated in the superior maxilla.

Prosthetic devices were inserted and the patient remains well without recurrent disease or metastases seven years after the surgical removal of the primary neoplasm.

Case 2.
A 24-year-old woman was first seen by a physician in 1963 for epistaxis from the left side of her nose. A small bleeding point was cauterized at that time but
epistaxis persisted. In a two-month interval a firm cherry-red mass (from the middle turbinate) had partially occluded her left nasal passage. No erosion of bone was demonstrated on sinus x-rays. Biopsy was carried out and a tentative and presumptive diagnosis of hemangiopericytoma was made.

In July, 1964, a left lateral rhinotomy and subtotal maxillectomy was performed with removal of a ‘cherry-sized’ mass from the posterior portion of the ethmoid sinus. Following this, she received what was called a ‘full course’ of cobalt irradiation.

The patient did well, even having a cosmetic rhinoplasty in 1975, until February, 1978 when a mass appeared in her left maxilla. This was excised and demonstrated to be matted lymph nodes measuring $8 \times 6 \times 4.5$ cm and containing metastatic neoplasm. The nasal cavity and sinuses contained no recurrent neoplasm.

Pathologic examination

Since the neoplasms from both patients presented with nearly identical histologic appearances, they are presented together. In each instance, the original diagnosis of hemangiopericytoma was corrected by the discovery of islands of cartilage in the neoplasm. For the tumour in Case 1, this required additional sectioning of the primary tumour. In Case 2, the cartilage was identified in sections from the long-delayed metastasis.

The light microscopic appearance of both neoplasms conformed to the descriptions of mesenchymal chondrosarcomas given in the literature (Salvador et al., 1971; Guccion et al., 1973). They were composed of cellular areas consisting of undifferentiated mesenchymal cells (Fig. 2) and islands of cartilage, varying in their stage of development, yet benign in appearance (Fig. 3). The undifferentiated cells contain a dominant, hyperchromatic nucleus and scant cytoplasm. Retrogressive changes such as fibrosis and hyalinization were found in the mesenchymal chondrosarcoma of Case 1. Each neoplasm presented numerous, slit-like vascular spaces about which the mesenchymal cells tended to aggregate, hence the hemangiopericytoma appearance. Mitoses were spare and not atypical. The islands of cartilage usually merged abruptly with the mesenchymal cells (Fig. 4). Only small areas of calcification were found in the cartilage of the tumour in Case 2.

Electron microscopic examination was able to be carried out in the mesenchymal chondrosarcoma of Case 2. In this instance, also, the ultrastructural features conformed to those described earlier by Steiner et al. (1973). Figure 5 presents the ultrastructural appearance of the tumor cells from the mesenchymal chondrosarcoma of Case 2. The cells are round-to-ovoid and poorly differentiated, with a paucity of organelles. Bundles of collagen fibrils are noted in the matrix.

Discussion

All major reports concerning mesenchymal chondrosarcomas indicate the apparent preference of this histologically unique malignancy for a primary origin (osseous or non-osseous) from structures in the head and neck (Salvador et al., 1971; Guccion et al., 1973; Pittman and Keller, 1974). Figure 6 depicts the sites of origin of 30 mesenchymal chondrosarcomas of the head and neck (excluding
Fig. 2

Case 2. Section taken from the metastasis in the axillary lymph nodes. In this cellular area, the typical, poorly differentiated cells have a tendency to be arranged about vascular spaces. Hematoxylin and eosin ×60.

Fig. 3

Case 2. Section from the metastasis to the axilla. Young cartilage that has arisen from the cellular undifferentiated component of the mesenchymal chondrosarcoma. Hematoxylin and eosin ×160.
Case 1. Mesenchymal chondrosarcoma. In this field both the young cartilage and undifferentiated elements are present. The hemangiopericytoma pattern is less prominent. Hematoxylin and eosin × 90.

The two present cases). Fourteen have had a definable origin from bone; seven from the maxilla, four from the mandible, and three from skull bones. The orbit and meninges have been most often cited as giving rise to the extraskeletal forms. These are followed by single cases reported from the nasal cavity, nasopharynx, lateral neck, temporalis muscle and paramandibular region.

Also clear from the literature and reaffirmed by our cases is the observation that more than one-half of all patients are in their second or third decades of life when the diagnosis is made. This holds true regardless of a soft or hard tissue origin. The sexes are equally effected.

While there is variability in clinical course, the tumour, at any site, cannot be judged as anything but malignant and requires radical surgical excision. Excision, radiation, or curettment leads to nearly predictable recurrence and, worse, metastases and death. Like the melanoma, however, no patient with a mesenchymal chondrosarcoma of the head and neck can ever be considered 'cured' since long latent periods have occurred, followed by metastases. Hematogenous spread, primarily to the lungs, is favoured over lymphatic metastases. Five-year survival is consequently of limited significance.

Light microscopic descriptions have been well detailed in the literature (Dahlin and Henderson, 1962; Salvador et al., 1971; Guccion et al., 1973) and conform to those of our cases. In brief, the histologic diagnosis is established by
the finding of a richly cellular neoplasm composed of undifferentiated mesenchymal cells in which islands of relatively well-differentiated benign-appearing cartilage is found. The undifferentiated mesenchymal cells vary from uniform round and small cells to spindle-shaped cells. In both cell types, the densely chromatic nucleus dominates, with the cytoplasm barely visible. A tendency for the undifferentiated cells to arrange themselves about vascular spaces is often commented on by observers. While this feature is variable, not only between tumours, but also within a single neoplasm, it is this feature that so often misleads the unwary to the diagnosis of hemangiopericytoma (Fig. 7). The cartilage is essential for accurate diagnosis. Its dispersion in the tumours is usually unequal and may be missed if the lesion is not adequately sampled. Typically there are rather abrupt transitions from the undifferentiated mesenchymal cells to the islands of cartilage. Less often, the transition is more gradual.

The first electron microscopic evaluation of a mesenchymal chondrosarcoma was reported in 1973 (Steiner et al.). To our knowledge, the study of our Case 1 is the third such report (Fu and Kay, 1974). From these investigations, it is considered that mesenchymal chondrosarcomas originate from primitive mesen-

**Fig. 5**

Case 2. Electron micrograph of the cellular component of a mesenchymal chondrosarcoma. The cells are undifferentiated and lack organelles. In the upper right, fibrils of the matrix lie apposed to the cells. Magnification ×18,000.
FIG. 6
Schematic representation of the location of 30 previously reported mesenchymal chondrosarcomas of the head and neck region. Open circles signify osseous origin, closed circles signify deep, soft tissue origin. The orbit and meninges have been the most frequently reported sites.

FIG. 7
Hemangiopericytoma of nasal cavity. Note that, while the peritheliomatous arrangement of the cells is similar to the mesenchymal chondrosarcoma, the cells are better differentiated and cartilage is not found. Hematoxylin and eosin ×130.
chyme with the potential to differentiate into cartilage. The vascular pattern, resembling hemangiopericytoma, is due to a proliferation of the undifferentiated tumour cells around vascular spaces rather than pericytes. While there are many ultrastructural features in common between the mesenchymal chondrosarcoma and myxoid chondrosarcoma, the two tumours are distinctive clinico-pathologic entities and should not be confused (Enzinger and Shiraki, 1972; Fu and Kay, 1974).

Summary

Mesenchymal chondrosarcoma, in both osseous and extra-skeletal forms, has a decided predilection for the head and neck region. The two cases presented in this report affirm this tendency and also illustrate the capricious biologic behavior of the neoplasms as manifested by the 30 additional cases recorded in the literature. Histopathologic confusion with hemangiopericytoma is avoided by identification of the cartilage component in the mesenchymal chondrosarcoma and by the undifferentiated nature of the cellular (non-cartilaginous) component.

REFERENCES


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