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Case Report

Mesenchymal chondrosarcoma of the maxilla: Case report and clinicopathologic review

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ABSTRACT

Mesenchymal chondrosarcoma is extremely rare, accounting for around 1% of all chondrosarcomas. It occurs in the axial skeleton, the head and neck region and a high proportion of extraskeletal sites. Fewer than 40 cases of mesenchymal chondrosarcoma in the maxilla have been reported in English literature. The neoplasm is aggressive, with a high tendency for late recurrence and delayed distant metastasis. Here we present a 50-year-old man with palatal mucosal swelling for 6 months. The initial clinical impression was a benign minor salivary gland neoplasm. The biopsy revealed hyaline cartilage, alternating with undifferentiated small round cells, pathognomonic for mesenchymal chondrosarcoma. Diagnosis of biopsy specimens remains a challenge because of variable tumor components, and specimens may contain only one of the two neoplastic elements. The patient received radical surgery and has been disease free for 40 months. Clinicians and pathologists should be aware of this entity in the differential diagnosis of maxillofacial neoplasms, to prevent misdiagnosis and delayed intervention.

1. Introduction

Mesenchymal chondrosarcoma is a rare subtype of chondrosarcoma among World Health Organization classifications and it accounts for around 1% of all chondrosarcomas [1,2]. The tumor affects a relatively young patient population compared with conventional chondrosarcoma. The majority of mesenchymal chondrosarcomas occur in the axial skeleton. The head and neck region is also a common site for the neoplasm [2,3]. The sinonasal tract has rarely been reported and the maxillary sinus is the most commonly affected. Commonly reported symptoms include swelling, mass formation, and nasal obstruction. Here we present a rare case of mesenchymal chondrosarcoma originating in the maxilla and discuss the clinicopathological features.

2. Case report

A 50-year-old man presented with palatal mucosal swelling and mild nasal obstruction for 6 months. He had initially visited a local clinic with follow-up. On intraoral examination, one small elastic, immobile nodule was noted in the palatal area. No epistaxis or nasal discharge was noted and no enlarged neck lymphadenopathy was observed. The laboratory data were within reference levels. The initial clinical impression was a benign, minor salivary gland neoplasm. An incisional biopsy was performed for histopathological evaluation.

Microscopically, the biopsy revealed bland-looking hyaline cartilage, admixed with undifferentiated, small round cells in the stroma. The cartilage component was relatively well-differentiated and the undifferentiated areas exhibited blue round cells. No salivary component was observed. The picture was pathognomonic for mesenchymal chondrosarcoma. Subsequent image studies were done to stage the disease.

Magnetic resonance imaging revealed a T1-isointense, T2-hypointense 5 cm mass lesion in the left maxillary-alveolar area, involving the nasomaxillary lamina and hard palate (Fig. 1A). The lesion exhibited prominent contrast enhancement (Fig. 1B). Further specimens, taken during radical surgery, disclosed a tan tumor, 4.8 cm in diameter, mainly in the left maxilla (Fig. 2A).

The histopathological features revealed a picture of mesenchymal chondrosarcoma, composed of biphasic patterns with hyaline cartilage and undifferentiated small blue round cells (Fig. 2B). The proportions of the two components were relatively

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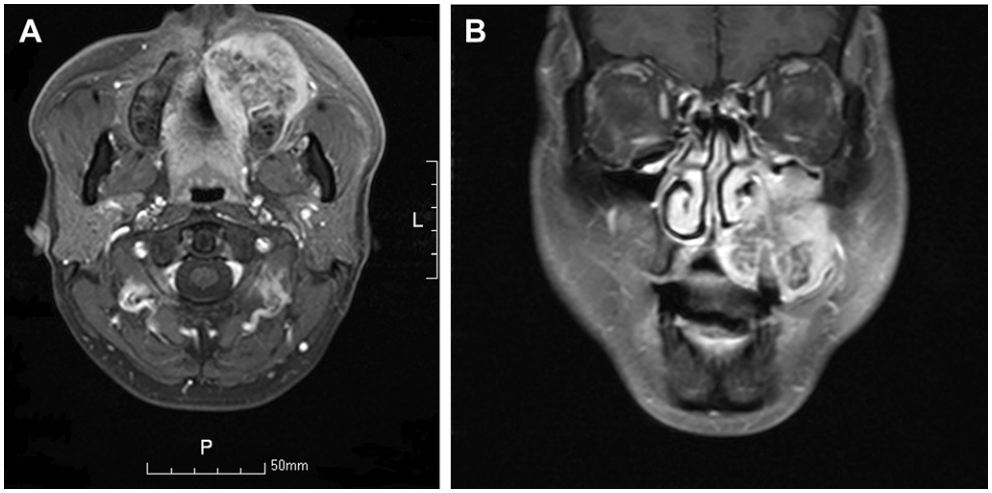

Fig. 1. (A) Magnetic resonance imaging displays a 5 cm mass lesion in the left maxillary-alveolar area; (B) the lesion exhibits contrast enhancement.

equal. The transition of the two patterns was abrupt in some regions and gradual in other areas. The hyaline cartilage component was well-differentiated and benign-appearing, with minimal atypia. These small cells exhibited round to oval hyperchromatic nuclei (Fig. 2C), with a focal hemangiopericytomatous-like vascular pattern (Fig. 2D). Immunohistochemically, the chondroid areas were positive for S-100 protein. The blue round cells were reactive for CD99. The lesion was mainly located in the left maxillary alveolar area, invading the hard palate and soft tissue. The section margins were free of disease. The patient has been followed up for 40 months, with no evidence of recurrence.

3. Discussion

Mesenchymal chondrosarcoma is a rare subtype of chondrosarcoma [1,2]. It shows no sex predilection. Mesenchymal chondrosarcoma is regarded as a high-grade sarcoma in the grading systems of the French Federation of Cancer Centers Sarcoma Group, the National Cancer Institute and the WHO classification [4]. The majority of mesenchymal chondrosarcomas occur in the axial skeleton. It may occur in the head and neck region, with a predilection for the maxillofacial skeleton [2,3]. The sinonasal tract has rarely been reported and the maxillary sinus is most

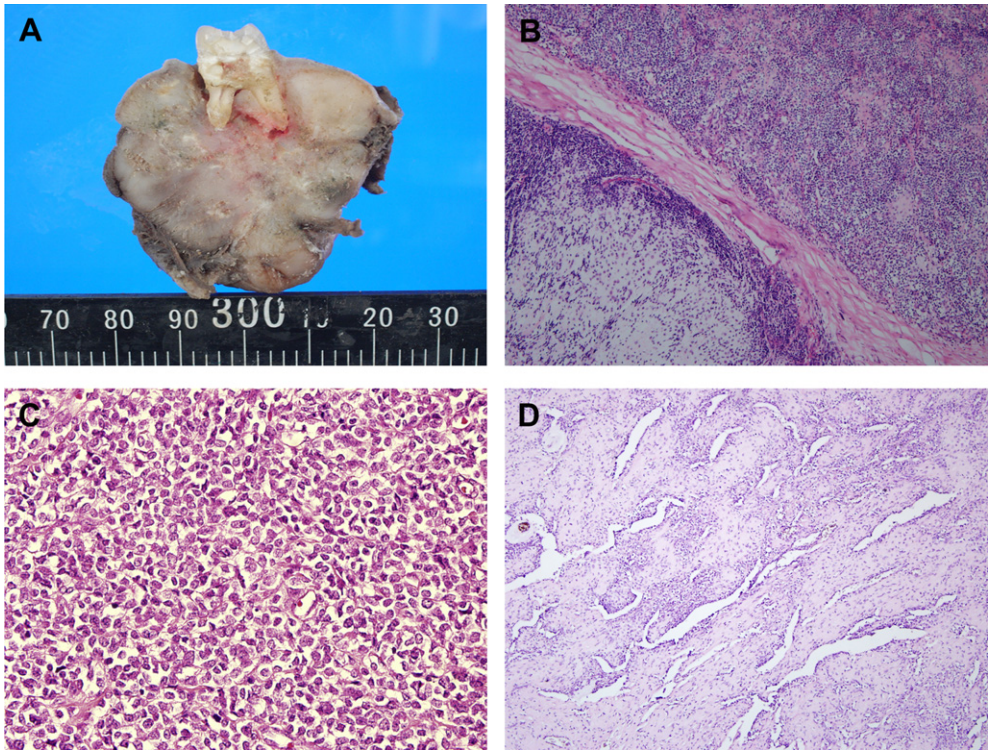

Fig. 2. (A) The tumor is tan and firm, mainly in the left maxilla, and bulges into the left palatal mucosa; (B) biphasic patterns of hyaline cartilage blending with undifferentiated small round cells (hematoxylin-eosin stain, ×100); (C) a small blue round cell area (hematoxylin-eosin stain, ×400); (D) there is a hemangiopericytomatous-like vascular pattern (hematoxylin-eosin stain, ×100).

commonly affected. Fewer than 40 cases of mesenchymal chondrosarcoma originating in the maxilla have been reported in English literature [5].

Computed tomography demonstrates a well-defined mass, with stippled calcified areas. Magnetic resonance imaging findings can be quite variable. The noncalcified areas demonstrate isointense to hypointense signal intensity on T1-weighted images and are hyperintense on T2-weighted images. Images after gadolinium contrast reveal inhomogeneous enhancement. The findings also include variable calcification, hypervascularity, lobulation, and bony destruction [2,6].

The tumor is typically large in the axial skeleton and relatively smaller in the maxillofacial regions and sinonasal tract [2]. The cut surface shows blue-gray fragments, with focal hemorrhagic areas. Microscopically, a biphasic pattern of well-differentiated chondroid elements, with an abrupt boundary from undifferentiated small round cells, is pathognomonic of the neoplasm. Focal myxoid degeneration and hemorrhagic necrosis may be observed. Tumor cells around vessels with a hemangiopericytoma-like growth pattern are common. Immunohistochemically, the small round cells are positive for CD99. The chondroid areas are positive for S-100 protein.

Diagnosis of biopsy specimens remains a challenge because of the variable tumor components and specimens may contain only one of the two elements. In one clinical series, only 38% of cases were accurately diagnosed on initial analysis [7]. The differential diagnosis includes a small blue round cell neoplasm as Ewing's sarcoma/primitive neuroectodermal tumors, small cell osteosarcoma, embryonal rhabdomyosarcoma, and lymphoma [2,5,7]. The identification of chondroid matrix and undifferentiated mesenchymal cells is crucial for an accurate diagnosis.

The primary treatment modality is radical surgery with wide safe margins. The overall role of multimodality treatment with radiation and/or systemic chemotherapy remains uncertain [2,8–10]. Mesenchymal chondrosarcoma has a tendency towards both local and distant recurrence [2,8]. The tumor affects a relatively younger patient population compared with conventional chondrosarcoma. There is also a high proportion of extraskeletal primary tumors, which are not observed with other chondrosarcoma subtypes [10]. The prognosis is markedly worse than for conventional primary chondrosarcomas, with 10-year survival rates of 20–30% [2,8]. A few series revealed better prognosis than other historical results with the tumors located in the sinonasal tract and jaws [3,7,9]. This could be attributed to more favorable tumor characteristics (e.g., smaller tumors). It is crucial to gain more information on the clinical behavior, pathogenesis, cytogenetic study and treatment from more series and prospective data of this rare neoplasm. Clinicians and pathologists should be aware of this rare entity in the differential diagnosis of maxillofacial neoplasm, to prevent misdiagnosis and delayed intervention.

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