Case Report

Mesenchymal chondrosarcoma of mandible

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Abstract Mesenchymal chondrosarcomas (MC) are rare and aggressive forms of chondrosarcoma. They are distinct tumors arising in unicentric or multicentric locations from both skeletal and extraskeletal tissues. The most affected region is the facial skeleton, especially the jaws. In this report, we present a case of MC primarily involving the mandible in a 60-year-old female patient.

Key Words: Chondroid, hemangiopericytoma mandible, mesenchymal chondrosarcoma

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INTRODUCTION

Mesenchymal chondrosarcoma (MC) is one of the most unusual cartilaginous malignancies of bone and soft tissues with distinct histopathological appearance and biological aggressive behavior. It was first described in 1959 by Bernstein and Lichtenstein as a separate entity of chondrosarcoma (CS).[1] It has a high metastatic and recurrence potential with poor survival.[2] This article describes a patient with MC affecting mandible. The clinical and pathological aspects of this lesion are presented and the relevant literature is reviewed.

CASE REPORT

A 60-year-old female patient visited with a chief complaint of slow growing painless swelling in the left lower back tooth region since 4 months. Extraoral examination revealed buccolingual expansion on the left side of the mandible. On palpation, the swelling was hard and nontender. Intraoral examination revealed an oval, solitary bony hard swelling with smooth margins measuring about 7 cm × 8 cm in size which obliterated left buccal vestibule extending from 43 to 38. The left submandibular and submental lymph nodes were about 2 cm, nontender and fixed. Past medical history was noncontributory.

Orthopantomogram and occlusal radiograph revealed mixed radiopacity and radiolucency extending from the roots of 43 to 38. No displacement of teeth and resorption of root were seen. Incisional biopsy was performed, and histopathological examination revealed proliferation of tumor cells which were spindle and epithelioid. Intracytoplasmic vacuolation admixed in a myxoid stroma was evident. Hyalinized capillaries lined by spindle to basaloid endothelial cells were also seen. Ossification was noticed in one area. No atypical features of tumor cells were observed. Overall histopathology was suggestive of intermediate malignancy of vascular origin possibly hemangioendothelioma [Figure 1]. Surgical excision of the lesion was done and referred for histopathological diagnosis [Figure 2].

Microscopically, tumor revealed areas of well-differentiated cartilage along with neoplastic chondrocytes and undifferentiated

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small round, ovoid and spindle cells with scanty cytoplasm arranged in clusters and sheets [Figure 3a]. Ossification of the chondroid areas was observed in few areas [Figure 3b]. Round to ovoid cells with indistinct cytoplasm were seen around the blood vessels imparting a hemangiopericytoma pattern [Figure 3c]. Focal areas showed clear cell changes. Immunohistochemical analysis showed diffuse positivity for S100 [Figure 4a] and strong positivity for vimentin [Figure 4b]. The cells were negative for cytokeratin (CK), CD31 and CD34 [Figure 4c]. Hence, the diagnosis of hemangioendothelioma on incisional biopsy was ruled out. We concluded a final diagnosis of MC. Patient was further referred to the Oncology Department where metastasis was ruled out and patient is now under close follow-up.

DISCUSSION

MC is one of the most unusual neoplasms of CS.[3] Four types of CS have been cited in the literature: Grade I, Grade II, mesenchymal and myxoid. The mesenchymal type is the most aggressive because of its tendency to grow in deeper tissues.[4] MC of the head and neck occurs most often in the third to sixth decades of life.[5] Both skeletal and extraskeletal lesions have been reported in head and neck region. Extraskeletal tumors commonly arise in orbit, meninges, nasal and paranasal regions.[6] This neoplasm affects females more frequently than it does in males (female/male = 1.4/1).[3] The clinical presentation of MC depends on the location and size of the tumor. Generally, they present as painless swellings causing facial deformity and malocclusion as reported in our case. Few patients may develop neurological disturbances such as facial and lip paresthesia.[7]

Among the facial skeletal region, jaws are most commonly involved with more preference to maxilla unlike the present case involving the mandible.[7] In mandible, the most common location is the premolar-molar area, but the symphysis or coronoid condylar processes may be involved.[3] They are

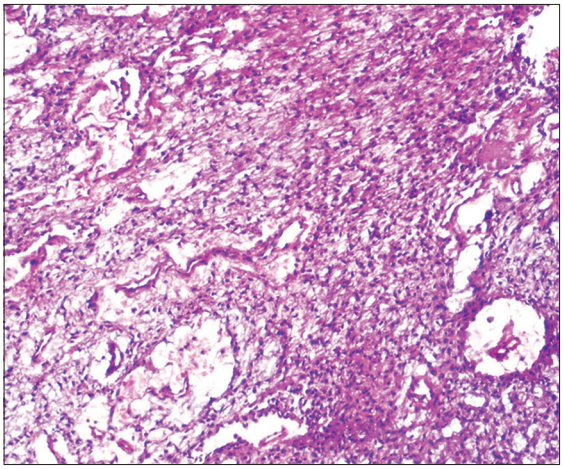

Figure 1: Photomicrograph showing proliferation of spindle and epithelioid tumor cells admixed with numerous blood vessels (H&E stain, ×100)

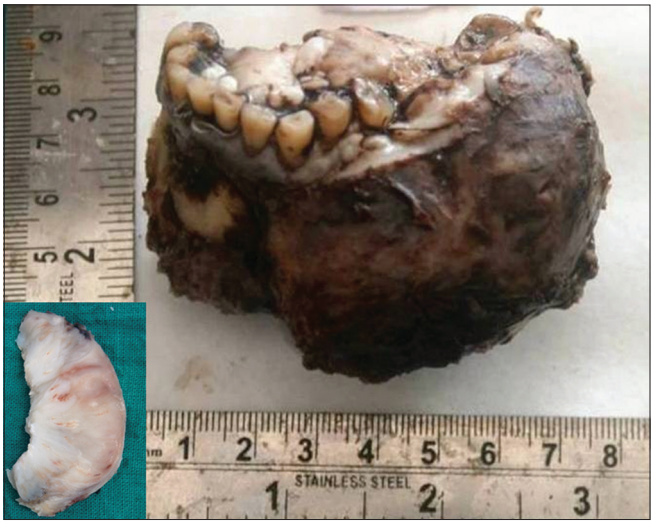

Figure 2: Macroscopic appearance of the lesion

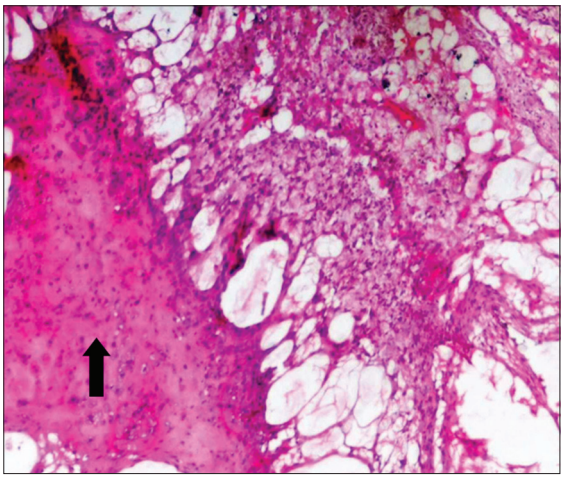

Figure 3a: Photomicrograph showing sheets or clusters of lesional cells alternating with well-differentiated cartilaginous areas (H&E stain, ×100)

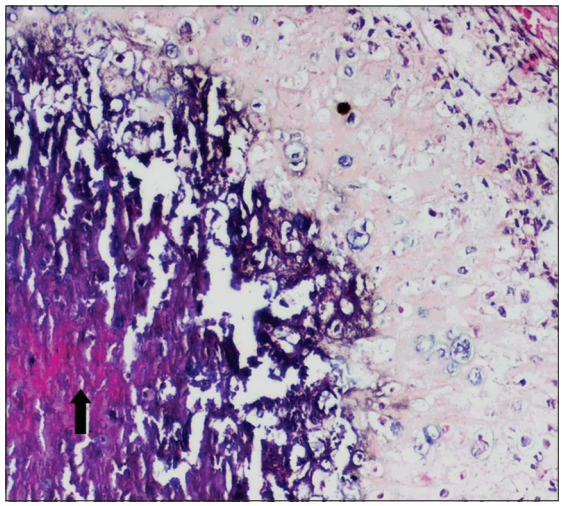

Figure 3b: Photomicrograph showing chondroid area along with ossification (H&E stain, ×100)

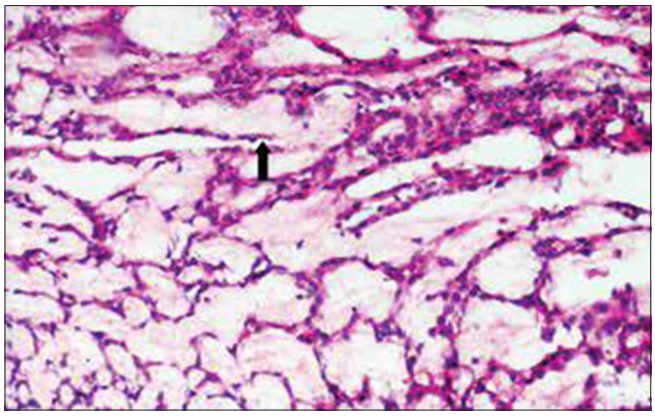

Figure 3c: Photomicrograph showing round to ovoid cells with indistinct cytoplasm around the blood vessels imparting a hemangiopericytoma pattern (H&E stain, ×200)

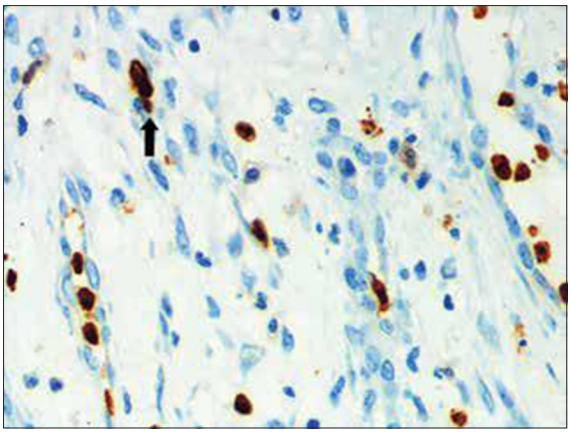

Figure 4a: Tumor cells showing varied positivity for S-100 (IHC stain, ×400)

Figure 4b: Tumor cells showing strong positivity for vimentin (IHC stain, ×40)

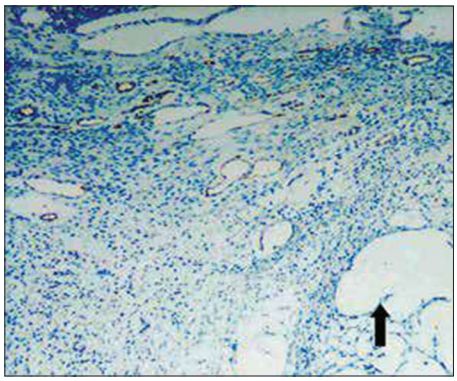

Figure 4c: Tumor cells were negative for CD34 (IHC stain, ×40)

relatively firm with few calcifications and fair vascularity. They show direct bone expansion and erosion of the mandible.[8]

The radiographic appearance of MC in the jaws varies from being radiolucent to a mixed radiolucency/radiopacity. In the mandible, these lesions are generally radiolucent, with zones of radiopacity in the lytic areas.[9] Present case showed mixed radiolucency and radiopacity.

Obtaining adequate tissue to diagnose MC may be problematic because of its propensity to occur in inaccessible deeper maxillofacial areas.[7] Incisional biopsy may sometimes fail to identify adequate features required to secure the final diagnosis and are usually misdiagnosed as odontogenic fibroma, chondromyxoid fibroma, fibrosarcoma and angiosarcoma.[1,6] The essential light microscopic features required for the diagnosis include bimorphic appearance showing both highly cellular undifferentiated tumor cells with apparent small cell population transitioning into ovoid, spindle cells and multifocal areas of chondroid differentiation.[10] Clustering of intratumoral blood vessels akin to hemangiopericytoma-like areas was also evident.[11] The larger cartilage islands may undergo calcification or ossification as in our case.

As diagnosis is challenging when limited tissue is available for analysis, it may be assisted by ancillary pathologic techniques such as immunohistochemistry (IHC) [Table 1]. S-100 positivity is varied with loss of expression in the round cell component unlike the conventional CS which is strongly positive. The round cell component is strongly positive for CD99. Type II collagen is a selective marker for chondrogenic neoplasms which is specific for MC as it is not expressed in other small cell sarcomas.[8]

Histopathological differentials include, Ewing's sarcoma showing similar histological findings and is positive for vimentin and CD99 but lacks chondroid areas and hemangiopericytoma-like pattern. MC shows clustering of intratumoural blood vessels which may have to be distinguished from angiosarcomas. However, these lesions do not demonstrate chondroid components such as those of MC and are positive for vascular markers, CD31 and CD34. Synovial sarcoma and some spindle cell variant share the similar histology, but S-100 positivity and CK negativity will enable the demarcation of these tumors.[12,13] Differentiating chondroblastic osteosarcomas may not be difficult as it shows malignant spindle cells toward

Table 1: Differential diagnosis for mesenchymal chondrosarcoma

Histopathological features	MC	Conventional CS	Chondroblastic osteosarcoma	Ewing's sarcoma	Angiosarcoma
Histopathology	Biomorphic with highly cellular undifferentiated tumor cells and small round cell population Multifocal areas of chondroid Hemangiopericytoma pattern	Lobular growth pattern showing chondrocytes of varying degrees of maturation and cellularity Peripheral areas show immature cartilage and mesenchymal tissue	Malignant spindle cells toward the periphery of the lobules with the presence of lace-like tumor osteoid Osteoclastic multinucleated giant cells are usually observed	Similar histology of MC but lacks chondroid areas and hemangiopericytoma-like pattern	Anastomosing vascular channels lined by enlarged endothelial cells with hyperchromatic or vesicular nuclei Hemangiopericytoma pattern with no areas of chondroid
Immunohistochemistry	S-100 - focal positive with loss of expression in round cell component CD99-round cell component is strongly positive Type-II collagen-specific marker for MC Keratin negative	Strong positive for S-100 Keratin negative Collagen II positive	Positive for osteonectin and osteocalcin	Positive for CD99 and O-13 product of MIC-2 gene	Positive for CD31, CD34 and Factor VIII (focal areas)

MC: Mesenchymal chondrosarcoma; CS: Chondrosarcoma

the periphery of the lobules with the presence of lace-like tumor osteoid, whereas MC lacks the tumor osteoid.[14]

There is diversity of opinion in the diagnosis and treatment of MC.[15] The most effective therapeutic modality is wide surgical excision along with chemotherapy and radiotherapy. In case of mandible, a wide local excision with a tumor-free margin of 2 to 3 cm is recommended.[3] According to Nakashima *et al*., extensive resection has less recurrence and a better survival rate than limited surgical resection.[10] Prognosis of MC is poor because the lesion has a high tendency for late recurrence either locally or metastasizes by hematogenous route. Lungs are the most common site for metastasis. Five-year survival rates for craniofacial MC are 40–60%.[3]

Present case emphasized that the diagnosis of MC should be evaluated cautiously. Adequate biopsy with meticulous histopathological examination of the multiple sections along with adjunctive IHC is the key for the definitive diagnosis. As recurrence and metastasis are very frequent with MC, follow-up for a long period should be advised.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Naik LR, Shetty P, Teerthanath S, Makannavar JH, Shroff SE. Mesenchymal chondrosarcoma affecting the mandible. J Oral Maxillofac Pathol 2008;12:29-33.
2. Singh P, Singh A, Saxena S, Singh S. Mesenchymal chondrosarcoma of mandible: A rare case report and review. J Oral Maxillofac Pathol 2014;18 Suppl 1:S167-70.
3. Akpolat N, Gok O. Mesenchymal chondrosarcoma of the mandible. Turk J Med Sci 2004;34:209-13.
4. El Mostarchid AB, Belhachmi A, Khaddar A, Gazzaz M, Boucetta M. Spheno-orbital mesenchymal chondrosarcoma: A rare cause of exophthalmos. Pan Arab J Neurosurg 2010;14:86-9.
5. Anil S, Beena VT, Lal PM, Varghese BJ. Chondrosarcoma of the maxilla. Case report. Aust Dent J 1998;43:172-4.
6. Williams HK, Edwards MB, Adekeye EO. Mesenchymal chondrosarcoma. Int J Oral Maxillofac Surg 1987;16:119-24.
7. Zakkak TB, Flynn TR, Boguslaw B, Adamo AK. Mesenchymal chondrosarcoma of the mandible: Case report and review of the literature. J Oral Maxillofac Surg 1998;56:84-91.
8. Pellitteri PK, Ferlito A, Fagan JJ, Suárez C, Devaney KO, Rinaldo A. Mesenchymal chondrosarcoma of the head and neck. Oral Oncol 2007;43:970-5.
9. Aziz SR, Miremadi AR, McCabe JC. Mesenchymal chondrosarcoma of the maxilla with diffuse metastasis: Case report and literature review. J Oral Maxillofac Surg 2002;60:931-5.
10. Nakashima Y, Unni KK, Shives TC, Swee RG, Dahlin DC. Mesenchymal chondrosarcoma of bone and soft tissue. A review of 111 cases. Cancer 1986;57:2444-53.
11. Bertoni F, Picci P, Bacchini P, Capanna R, Innao V, Bacci G, *et al*. Mesenchymal chondrosarcoma of bone and soft tissues. Cancer 1983;52:533-41.
12. Nguyen DV, Muda AS, Yaacob Y. Mesenchymal chondrosarcoma: A case report. Malays J Med Sci 2013;20:71-7.
13. Thomas RS, Sherlin HJ, Natesan A, Premkumar P, Ramani P, Chandrasekar T, *et al*. Mesenchymal chondrosarcoma: A report of two cases with immunohistochemical study. Iran J Pathol 2010;55:100-4.
14. Gnepp DR. Diagnostic Surgical Pathology of the Head and Neck. 2nd ed. Philadelphia: Saunders Elsevier; 2009.
15. Krishnamurthy A, Vaidhyanathan A, Srinivas S, Majhi U. A fatal case of mesenchymal chondrosarcoma of the mandible. J Cancer Res Ther 2011;7:192-4.