Mesenchymal chondrosarcoma: An unusual lump in posterior maxilla

S. Ravi Raja Kumar, Anuradha A., Namineni Kiran Kumar, Horatti Puneeth Kuberappa, Venkata Raju K., Mahanthi Vijayalakshmi

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Case Report: We report an unusual case of mesenchymal chondrosarcoma on posterior palate in a 45-year-old male, who was previously diagnosed as pleomorphic adenoma in private clinic.

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CASE REPORT

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Keywords: Cartilage tumor, Mesenchymal chondrosarcoma, Posterior maxilla, Recurrent lesion

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INTRODUCTION

Mesenchymal chondrosarcoma is an uncommon, slow growing malignant tumor which is a rare variant of chondrosarcoma, having a predilection for the maxillofacial skeleton; less often involves the soft tissue sites in head and neck. They usually occur in middle aged individuals, but are rare in young patients, with predilection for anterior portion of maxilla [1]. The tumor is unique due to high tendency for late recurrence and delayed metastasis to lung, bone, and lymph nodes [2]. Histopathologically, it is characterized by a biphasic pattern consisting of hyaline cartilage mixed with undifferentiated pleomorphic mesenchymal cells [1, 3]. To the best of our knowledge, only 17 cases have been reported in English literature affecting posterior maxillary region. Thus we report an unusual case of mesenchymal chondrosarcoma on posterior palate in a 45-year-old male.

CASE REPORT

A 45-year-old male presented with painless swelling in the left palatal region present since four months. Patient gave a history of an excision of lesion in posterior left palatal region nine months ago, which was diagnosed as pleomorphic adenoma in a private clinic. In duration of four months, patient again noticed a swelling in the site of previous excision. On clinical examination a well-defined solitary swelling of size approximately 3x1.5 cm extending from distal aspect of 21 to distal aspect of 26 on the left buccal alveolar ridge anteroposteriorly and mediolaterally extending from palatine gingiva to mid palatine raphe in relation to 22–26. The surface appears to be lobulated and mucosa over the growth appeared normal (Figure 1A–B).

Computed tomography scan and orthopantomogram showed no definite changes, whereas paranasal sinus (PNS) view showed haziness in maxillary sinus area. Magnetic resonance imaging (MRI) scan and positron emission tomography (PET) scan revealed a well-defined mass seen on the left side of the buccal vestibule extending into the maxillary sinus (Figure 2A–B). Based on clinical and radiographic features provisional diagnosis of recurrent pleomorphic adenoma was given.

The patient was advised to undergo surgery, and the tumor was resected by subtotal maxillectomy under general anesthesia. The excised specimen was sent for histopathological examination which revealed focal ulceration of the lining epithelium. The stroma revealed biphasic pattern (Figure 3A) with areas of relatively mature cartilage formation with focal areas showing calcification of mature cartilage and areas of primitive spindle to round shaped mesenchymal cells. Primitive appearing mesenchymal cells had scanty cytoplasm, eccentric placed irregular vesicular nuclei and exhibited moderate pleomorphism with variable mitotic activity (Figure 3B). Focally hemangiopericytoma like vascular pattern was evident (Figure 3C). Transitional zone between the chondroid foci and the mesenchymal component was more plodding but not sharp. Immunohistochemical study was done and the proliferating chondrocytes showed nuclear positivity with S100 protein (Figure 4A) and were negative for pancytokeratin (Figure 4B).

Correlating with the clinical, radiographical, histopathological and immunohistochemical findings the

Figure 2: (A, B) Magnetic resonance imaging scan and positron emission tomography scan showing a well-defined radiodense mass on the left side of the buccal vestibule extending into the maxillary sinus.

Figure 3: (A) Biomorphic patterns of islands of cartilage surrounded by primitive mesenchymal cellular areas (H&E stain, x200), (B) Primitive spindle to oval shaped pleomorphic mesenchymal cells admixed with calcified chondroid areas with atypical chondrocytes (H&E stain, x200), and (C) Mesenchymal cells arranged around blood vessels (H&E stain, x200).

Figure 4: Photomicrograph of IHC stain showing (A) S-100 positive proliferative chondrocytes, and (B) Negative stained for pancytokeratin.

Figure 1: (A, B) Intraoral photomicrograph showing lobulated growth on left buccal and palatal region.

case was diagnosed as mesenchymal chondrosarcoma. The present case was followed for a period of one year with no postoperative recurrence.

DISCUSSION

Chondrosarcoma is the third most common primary malignancy of bone after myeloma and osteosarcoma [4].

Mesenchymal chondrosarcomas are rare biphasic tumor with areas comprising spindle cell mesenchyme interspersed with areas of chondroid differentiation accounting for only 1% of all chondrosarcomas [5].

Mesenchymal chondrosarcomas may develop from pluripotent mesenchymal stem cells and can differentiate into angioblastic, fibroblastic or cartilaginous structures [3], but precise pathogenesis and biological behavior is not fully understood in head and neck mesenchymal chondrosarcoma. But recent studies have shown the identification of new gene HEY1-NCOA2 fusion which appears to be diagnostic, but missing in other subtype of chondrosarcoma [6].

Mesenchymal chondrosarcomas arise from soft tissue or bone in the ratio of 1:2 to 1:6 [3]. In head and neck region commonly involved extra skeletal sites are orbit, meninges and sinonasal tract, whereas intraskeletally anterior maxilla is the most common site [3], where preexisting nasal cartilage is present [6, 7], others sites in descending order of frequency of involvement are body of the mandible, the ramus, the nasal septum and paranasal sinuses [8]. The present case was reported in left posterior maxillary region involving both buccal and palatal areas.

Most of chondrosarcoma occur in 3rd–6th decade of life, where as mesenchymal chondrosarcoma variant occurs in younger age group i.e., 2nd–3rd decade of life in 70% of cases, but few cases have been reported between 5–7th decade [5, 8], where present case also falls in 5th decade. Concerning sex prevalence there some disagreement, as few authors' state male predominance and few state female predominance [1, 8].

Mesenchymal chondrosarcomas show no specific clinical signs and symptoms. The predominant symptom is usually a painless mass or swelling (53%) [5], as in the present case. However, other reported symptoms are nasal obstruction (32%), epistaxis (32%) tooth mobility (24%) [6] and rarely, lymphadenopathy and neurological disturbances such as facial paresthesia and lip paresis can also occur [1, 3].

The radiographic appearance of mesenchymal chondrosarcoma of jaws are not classic, usually exhibit features of a malignancy, consisting of osteolytic process with poorly defined borders. The ill-defined radiolucent area contains scattered foci of neoplastic cartilaginous tissue [3, 8]. The lesion may also cause symmetrical widening of the periodontal ligament space of involved teeth [8, 9], but was not evident in the present case.

Histologically, depending on the cellularity, nuclear staining (hyperchromasia) of the tumor cells and size of the nuclei, chondrosarcomas have been graded into low, intermediate and high grade [5]. Previously, the same case was diagnosed as pleomorphic adenoma due to the presence of chondroid area in the background of myxomatous stroma and also due to inadequate biopsy taken initially might have missed the characteristic areas of mesenchymal chondrosarcoma.

Histopathologically, mesenchymal chondrosarcoma shows characteristic biphasic pattern. Highly cellular undifferentiated/primitive spindle cells is similar to small cell tumor, but presence of islands of chondroid differentiation help in making proper diagnosis, but difficultly arises with small biopsy sample [6, 8]. Most of the mesenchymal chondrosarcomas show rich vascular component, as seen in the present case, often confused with hemangiopericytoma but hemangiopericytoma lacks hyaline cartilage component, additionally, it is positive for CD34.

Histologically, the lesion should be differentiated from small cell osteosarcoma and PNET/ Ewing sarcoma, malignant peripheral neuroectodermal tumor, synovial sarcoma (poorly differentiated type), dedifferentiated chondrosarcoma [10, 11].

Ewing's sarcoma usually contains moderate amount of cytoplasmic glycogen and presence of t(11;22) but lacks cartilaginous component, reticulin meshwork and vascular pattern of mesenchymal chondrosarcoma. Malignant peripheral neuroectodermal tumor contains more pleomorphic small cells than in mesenchymal chondrosarcoma and presence of rosettes. Tumors cells of mesenchymal chondrosarcoma and malignant peripheral neuroectodermal tumors are positive for CD99 and S100 but stromal cells of mesenchymal chondrosarcomas are negative for S100. Small cell osteosarcoma can be differentiated from mesenchymal chondrosarcoma by the presence of osteoid in lace like pattern. Though mesenchymal chondrosarcoma contains bone, it is not produced directly by the stromal cells but formed by endochondral ossification of cartilage islands [3, 11]. Immunohistochemically, the small cell components are positive for vimentin, CD99, and Leu7 but not for S-100 protein; latter is found instead in chondroid areas [12].

Dedifferentiated chondrosarcoma shows large and highly pleomorphic spindle cells with atypical nuclei in contrast to uniform nuclei of small cells of mesenchymal chondrosarcoma. In dedifferentiated chondrosarcoma, there is sharp margin between both components but in mesenchymal chondrosarcoma both are admixed. Monophasic synovial sarcoma (poorly differentiated type) can be separated from mesenchymal chondrosarcoma by the absence of hyaline cartilage. Absences of keratins, especially CK7, and EMA and t(x;18) can differentiate mesenchymal chondrosarcoma from synovial sarcoma [11]. Hemangiopericytoma is distinguished from mesenchymal chondrosarcoma by its lack of cartilage and positive immunohistochemical staining for CD34 [5]. The characteristic expression of type II collagen in matrix of mesenchymal chondrosarcoma helps in differentiation from other small cell sarcomas [6].

In relation to the histological diagnosis, immunohistochemical analysis is of great helpful. Immunohistochemistry is advised for difficult cases. Chondroid areas are positive for S-100 protein and vimentin [3, 5].

Wide surgical excision is the mainstay treatment for chondrosarcoma in the jaw bones [2]. These tumors are radio resistant, so chemotherapy can be used as an adjuvant therapy after wide surgical excision [7]. Our case was treated with wide surgical excision and followed up for three years with no recurrence.

The prognosis of the chondrosarcoma of the jaws is poor as compared to that of long bones. The cause of death is usually by direct extension of tumor into the base of skull, and also through distant metastasis, primarily to lungs and bones. The prognosis is good for low and intermediate grade chondrosarcomas. Especially, the maxillary (0.7% of whole body tumor) and mandibular locations of the tumor have documented inferior prognosis [12].

Fu and Perzin in 1974 described three prognostic factors: location and extent of the lesion, adequacy of surgical therapy, and degree of differentiation of the tumor. Few studies have shown five-year survival rate up to 40–60% and few showed recurrence even after 10–20 years [1]. Hence adequate treatment and a lifelong follow-up after surgery is recommended for patients with mesenchymal chondrosarcomas of the maxillofacial region.

CONCLUSION

Mesenchymal chondrosarcoma shows varied clinical and radiographic features, with occurrence of this lesion in unusual locations like posterior region of jaw which shows ill-defined feature and ossification which may lead to error in clinical and histopathological diagnosis and delay in early treatment. It is an aggressive neoplasm with high penchant for recurrence and delayed metastasis. Hence, patient should be kept under long-term follow-up.

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Author Contributions

S. Ravi Raja Kumar – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Anuradha A. – Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Namineni Kiran Kumar – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Horatti Puneeth Kuberappa – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Venkata Raju K. – Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Mahanthi Vijayalakshmi – Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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