INTRODUCTION

Chondrosarcoma (CS) is a malignant tumor characterized by the formation of cartilage and usually arises in long and flat bone. It accounts for approximately 10-20% of malignant bone tumors. Head and neck CS is an uncommon entity accounting for approximately 0.1% of all head and neck malignancies. Several variants of CS have been proposed including clear cell, dedifferentiated, myxoid and mesenchymal. Mesenchymal chondrosarcoma (MC) is a rare variant of CS that accounts for up to 3-9% of all CS and has high predilection for the head and neck region. MC is one of the most unusual rare malignant cartilaginous malignancy of bone and soft tissues with distinct histopathological appearance and biological behavior and was described by Lichteinstein in 1959.

Approximately 33% of these tumors occur in an extraskeletal location or are associated with a large, extraskeletal, soft tissue mass, commonly in the meninges. Similar to conventional chondrosarcomas, lesions occur in the ribs, pelvis, and jaw bones more frequently than other bone neoplasms. They also are identified commonly in the extremities, particularly the femur. There is a predilection for the maxilla followed by mandible in the craniofacial complex. MC occurs between ages 30-60 years with a mean age of 47.5 years. Patients typically present with vague symptoms of pain and swelling that may have been present for a relatively long duration. Histologically, it is characterized by a biphasic pattern consisting of areas of hyaline cartilage mixed with small cell malignancy.

CASE REPORT

A 60-year-old male patient reported with the chief complaint of swelling in the lower right side of the face since 1 month. History revealed that the swelling was small 1 month back which rapidly grew to attain the present size. Extra-oral examination revealed a diffused swelling on the right side of the face measuring approximately 6 × 7 cms in size, extending from the temporal region to inferior border of the mandible superior-inferiorly and from the nose to the ear anteroposteriorly. The skin over the swelling was slightly stretched with no secondary changes. On palpation the swelling was non-tender, varied in consistency from soft in the center to hard at the periphery. The right sub-mandibular lymph nodes were palpable, non-tender, hard and fixed to underlying structure.

Intraoral examination showed an oval swelling present on the right side of the jaw in the retromolar area measuring approximately 4 × 3 cms in size. The swelling had diffuse margins with smooth surface.
Radiological examination revealed a diffused radiolucency involving the lower body and angle of the mandible extending from right second premolar to angle of the mandible [Figure 3].

Considering the size and aggressive nature of the lesion, surgical resection with wide margins of the lesion was carried out. The gross soft tissue specimen received for histopathological examination measured approximately 12 × 5 mms in size, was soft to firm in consistency, irregular in shape and whitish brown in color. The surgical material was fixed in buffered formalin and subjected to histopathological analysis.

On histopathological examination, the Hematoxylin and Eosin (H and E) stained section showed sheets of tumor cells interspersed by cartilage area showing calcification [Figures 4 and 5]. Higher magnification showed pleomorphic cells with large basophilic ovoid nuclei and small darkly stained nuclei with little cytoplasm [Figures 6 and 7]. High degree of vascularity was seen in some areas. Scattered, small, poorly circumscribed foci of cartilaginous tissue were also seen. Correlating the clinical, radiological and the histopathological features a diagnosis of MC was made. The patient was further advised for radiotherapy. The patient showed no signs and symptoms since 2 years and is being followed up 6 monthly periodically for possible recurrence.

**DISCUSSION**

MC was first described by Lichtenstein L and Bernstein D in 1959 as a biphasic tumor, comprising of spindle cell mesenchyme interspersed with areas of chondroid differentiation.[5] It is a specific variant of CS[7] and represents approximately 1% of all CSs.[6] MC of the head and neck occurs most often in the third to the sixth decades of life[8] that is very much consistent with the present case. It shows almost equal predilection in males and females.[9]

The etiology of these tumors is unknown. However, they are formed from cartilage in tissues not normally harboring cartilage or, secondly, from the cartilage cap of exostosis or enchondromas. Vestigial rests of multipotential differentiation
of mesenchymal cells may be the forerunner.\cite{5} It originates from bone, meninges or less commonly, soft tissues.\cite{10} According to Thomas RS et al., (2010), 3 to 25\% of all skeletal MC occurs in the maxillofacial region the premolar-molar region being the most common site. The predominant clinical symptom is painless mass or swelling. Other most commonly reported symptoms are nasal obstruction, epistaxis, tooth mobility, headache, bleeding, ulceration, facial asymmetry, paresthesia and trismus.\cite{8}

Radiographically, these lesions appear as osteolytic, radiolucent shadows with ill-defined, ragged borders.\cite{6} Diagnosis can only be established by combination of clinical, radiographic and histopathological examination.\cite{8}

The histologic appearance of MC is characteristically biphasic pattern. Undifferentiated areas appear as sheets of primitive mesenchymal spindle/round cells similar to small cell anaplastic sarcoma. However, islands of relatively well differentiated cartilaginous tumor help in making a specific diagnosis. Calcification or ossification may occur within the chondroid matrix. Neoplastic cartilage may be replaced by bone in a manner similar to normal endochondral ossification. Histologically, the lesion must be differentiated from similar other lesions like hemangiopericytoma, Ewing’s sarcoma, PNET, leukemia/lymphoma, rhabdomyosarcoma, and malignant melanoma\cite{1}

The most effective therapeutic modality is wide surgical excision. Wide local excision with a tumor free margin of 2-3 cm is recommended. Extensive resection has less recurrence and a better survival rate than limited surgical resection. Also, the postoperative radiotherapy and chemotherapy offer a good prognosis and eradicate chances of micrometastases. The prognosis for MC is poor because tumors have a tendency for late recurrence either locally or as metastasis. Metastasis of MC is hematogenous and the most common site is the lung. Five-year survival rates for craniofacial MC are 40-60\%.\cite{9}

CONCLUSION

MC is a rare mesenchymal tumor that occurs frequently in both hard and soft tissue locations. These tumors show local aggressive behaviour as well as a high metastatic and recurrence potential. Due to these features the prognosis of MC is poor. Considering the propensity of these tumors to metastasize and the poor prognosis of patients with MC, early identification may allow earlier, more aggressive interventions.

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