

# Mesenchymal Chondrosarcoma of the Mandible

Roobina Khan<sup>1</sup>, Syed Hasan Haris<sup>2</sup>, Veena Masheshwari<sup>3</sup>, Kiran Alam<sup>4</sup>, Arshad Hafeez Khan<sup>5</sup>

<sup>1</sup>Assistant Professor, Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, India

<sup>2</sup>Associate Professor, Department of General Surgery, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, India

<sup>3</sup>Professor, Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, India

<sup>4</sup>Associate Professor, Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, India

<sup>5</sup>Professor, Department of Plastic Surgery, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, India

## ABSTRACT

Chondrosarcoma is an uncommon malignant mesenchymal tumor of the head, neck, oral and maxillofacial regions characterized by the production of cartilaginous tissue and the absence of production of bone tissue. Mandibular localization of these tumors is rare. These tumors grow indolently to large sizes in young males. These neoplasms reveal aggressive local behavior and a high

metastatic potential. Wide surgical excision is one of the preferred treatments, although radiotherapy and chemotherapy are advised as an adjunct without many advantages. We report a case of mesenchymal chondrosarcoma of the mandible in a young female that exhibited massive extension of the tumor to the soft tissues.

**Keywords:** Chondrosarcoma; Malignant Mandibular Tumor; Mesenchymal Chondrosarcoma

## INTRODUCTION

Mesenchymal chondrosarcoma (MCS) was first described by Lichtenstein and Bernstein in 1959 as a special variant of chondrosarcoma [1]. It is a rare malignant tumor of bone and soft tissue. One of the most affected regions is the facial skeleton, especially the jaw. Males are more commonly affected [2]. Pain may be a late stage feature, and regional lymphadenopathy is very rare [3].

The lesion was originally grouped with osteosarcoma. These neoplasms are characterized by sheets or clusters of highly undifferentiated, small, ovoid cells that alternate with small zones of neoplastic cartilage. The prognosis for patients with MCS is unpredictable. This type of neoplasm shows aggressive local behavior as well as a high metastatic potential. Due to these features and the high risk of recurrence, the prognosis is poor.

## CASE REPORT

A 20-year-old female presented to the surgical out-patients department with a large swelling of the right jaw. It was painless and gradually increased in size over a six and a half months period. The swelling measured about 15x10 cm, being fixed to the mandible and the soft tissues especially the cheek with intra-oral extension

(Figure 1). The overlying skin was stretched and shiny. There were no engorged veins. The facial nerve was not involved with the tumor. The fixity to the masseter muscle could not be ascertained because of the large size of the tumor. There was no cervical lymphadenopathy. On abdominal examination, the liver was not palpable. There was no clinical evidence of distant metastasis. Fine needle aspiration cytology was

**Figure 1:** Swelling fixed to the mandible



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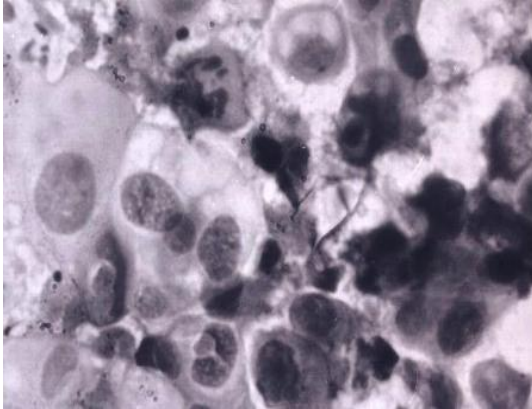
*Correspondence to:* Dr.  
Syed Hasan Haris

*Address:* Department of  
Pathology, Jawaharlal  
Nehru Medical College,  
Aligarh Muslim  
University, Aligarh, India

*E-mail:*  
[hasanharris@yahoo.com](mailto:hasanharris@yahoo.com)

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**Figure 2:** Isolated and loosely cohesive aggregates of oval or polygonal chondrosites with vacuolated granular cytoplasm and a background of chondroid matrix



characterized by the presence of isolated and loosely cohesive aggregates of oval or polygonal chondrosites with vacuolated granular cytoplasm and a background of chondroid matrix (Figure 2). A preoperative diagnosis of chondrosarcoma of the mandible was made. The patient was given doxorubicin-based combination neo-adjuvant chemotherapy. The patient was then operated on. A left hemimandibulectomy was performed with wide surgical margins under general anesthesia. The postoperative course was uneventful.

The contour of the face was maintained with the help of autologous rib graft (Figure 3). Postoperative radiation therapy was also given to the patient.

The resected specimen showed the tumor along with the mandible (Figure 4). The histopathological examination was characterized by sheets of highly undifferentiated, small, ovoid cells having nuclear atypia, hyperchromasia with myxoid stroma. The diagnosis of mesenchymal chondrosarcoma was established.

Three months after surgery, the patient was without local recurrence or metastasis.

## DISCUSSION

Patients with malignant bone tumors often report at a very late stage of the disease [4]. Chondrosarcoma has a very aggressive course locally and tends to recur or metastasize in the late stages and therefore the long-term prognosis becomes extremely grave [4].

It may occur at any age; however, most frequently it is found between the third and sixth decades of life. Males are more commonly affected than females [2]. A study of all cases of

maxillofacial chondrosarcomas revealed the maxilla to be more commonly involved than the mandible [5].

Our case, although being present in a young patient, was seen in the mandible and the diseased was a female highlighting the rarity of the lesion.

Painless swelling of the jaw is the presenting symptom in most of these patients [6, 7]. In developing countries, patients with these tumors mostly report late for treatment and this period ranges between two weeks to four years with an average of 18 months [4].

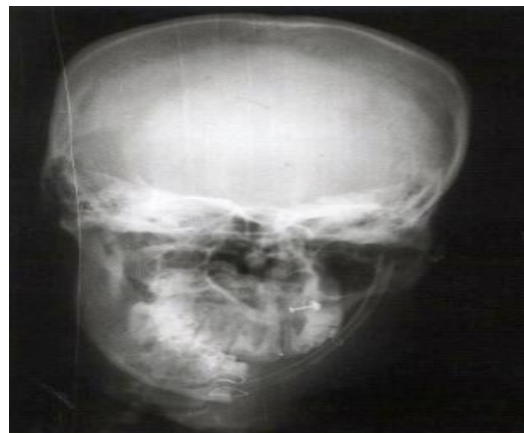
Fine needle aspiration cytology (FNAC) of these lesions due to their rarity and limited experience with the spectrum of cytological and histopathological features poses a challenge to many cytopathologists. FNAC diagnosed 87.2% of jaw chondrosarcomas in a series by Nanda et al [8].

Resection with wide margins of clearance of the tumor is the agreed treatment of choice and gives the best outcome for the patients [9, 10].

Although preoperative chemotherapy and postoperative radiation therapy as adjunctive modalities were given to our patient, both these modalities have undefined roles and offer no survival benefits [7, 9].

These tumors tend to recur locally after resection and metastasize even years after surgery. The most common site of distant metastasis is the lung. Crawford et al [9] noted that these tumors tend to recur or metastasize and found the five-year survival rate to be misleading. Overall, there is a ten-year survival rate of 28% [9]. Therefore, even after complete surgical resections with negative tumor margins, chondrosarcomas need a long-term follow-up.

**Figure 3:** Contour of face maintained by autologous rib graft



**Figure 4:** Tumor along with mandible



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