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Case Report *Volume 1; Issue 1*

Rare lesion of the Infratemporal Fossa

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Abstract

Mesenchymal chondrosarcoma (MC) is an aggressive, uncommon histological entity rising in bone and soft tissue. It is subtype of chondrosarcoma and second most common tumor of bone accounting of about 2 %. We are presenting a case of 35years old male patient treated at Liquate National Hospital with Infratemporal fossa mass. A Computerized tomography (CT) of face and sinuses demonstrated a lesion in Infratemporal fossa. Excision of Infratemporal fossa mass with sub ciliary extension was performed. It was diagnosed as Grade II Mesenchymal chondrosarcoma. Patient was advised post operative chemotherapy. No recurrence was noted on follow-up.

Keywords: Mesenchymal chondrosarcoma; Infratemporal fossa

Abbrevations: CT: Computerized Tomography; MC: Mesenchymal Chondrosarcoma; MCS: Mesenchymal Chondrosarcomas

Introduction

Mesenchymal chondrosarcoma (MC) of the head and neck is an uncommon tumour with a potential for exhibiting highly aggressive behaviour. In the head and neck region, they may involve skeletal or extra skeletal tissues. In skeletal tissue it involves maxilla and mandible. It was first described in 1954 by Lichtenstein and Bernstein in bone. Dowling Subsequently reported this entity in the soft tissue in 1964 [1]. It is a subtype of chondrosarcoma and is thought to arise from remnants of the embryonic cartilage or metaplasia of meningeal fibroblasts [2]. MC mostly affects children and young adults between the ages of 15 and 35 years and accounts for less than 1% of all sarcomas [3]. One-third of the cases occurs outside the bone and are seen more in young patients. Occurrence is mostly seen in the central nervous system, meninges, maxillary sinuses, eyelid, eye socket, and thyroid [4]. Four types of chondrosarcoma have been cited in the literature: grade I, grade II, mesenchymal, and myxoid. It has a variable clinical course with frequent recurrence and occasional distant osseous and visceral metastatic spread. The prognosis is poor, independent of the type of treatment [5] and shows no sex dominance [6]. We are presenting a case report of MCS of Infratemporal fossa in a young male which was excised and no recurrence was noted in follow-up.

Case Presentation

We are presenting a case of 35 years old male patient treated at Liquate National Hospital who presented with complaint of Swelling & numbness over left side of face for 5 months. Detailed history revealed that it was progressive and painless. Examination showed there was a diffuse swelling over left maxillary region causing facial symmetry. It was extending posteriorly up to left temporal region and superiorly up to left orbital region

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,overlying skin appears normal (Figure 1). It was firm in consistency measuring about 5x 5 x 4cm and no change in temperature of skin noted. However there was left eye proptosis was noted.

Figure 1: Blue arrow showed swelling of left side of cheek.

Endoscopic biopsy of Infratemporal fossa initially showed Ewing sarcoma, latter on a computerized tomography (CT) of Face and sinuses demonstrated a heterogeneously enhancing mass in left infra temporal fossa measures 5.7 x 4.4x5.5cm causing erosion of greater wing of sphenoid, floor of left orbit and lateral wall of left maxillary sinus. Superiorly it was extending into floor of the left globe, posteriorly extending up to left lateralpterygoid plate, medially it was causing mass effect over the maxillary sinus and laterally it was causing soft tissue thickening over the lateral aspect (Figure 2). His metastatic workup which included CT scan Chest with contrast and Bone scan were negative so, excision of Infratemporal fossa tumor through open approach was performed (Figure 3).

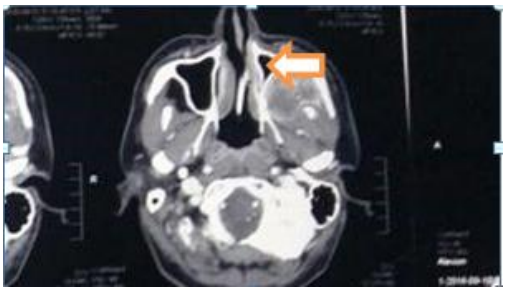

Figure 2: CT SCAN BASE OF SKULL AXIAL view Heterogeneous enhancing tumor involving left infra temporal fossa tumor with bowing of posterior wall of maxillary sinus (showed by arrow).

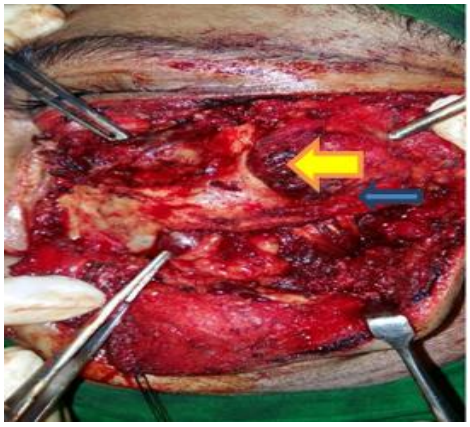

Figure 3: Yellow arrow shows tumor, Blue arrow shows zygomatic arch.

Final histopathology showed grayish brown piece of tissue measuring 7x 2.7 x 1.1cm.Microscopically it was composed of poorly differentiated small to round cells with hyper chromatic to vesicular nuclei, in conspicuous nucleoli & scanty cytoplasm. It was positive for CD99. Morphologically and immune histo chemical profile was in favor Mesenchymal Chondrosarcoma Grade 2 according to FNC LCC grading system. Later on this case was discussed in tumor board meeting & he was advised chemotherapy. Later on in his follow up no recurrence was noted. Preoperatively there was a well demarcated encapsulated tumor filling left infratempral fossa, superiorly eroding zygomatic arch and inferior wall of orbit, anteriorly bowing posterior wall of maxilla, posteriorly abutting lateral pterygoid plate (Figure 4).

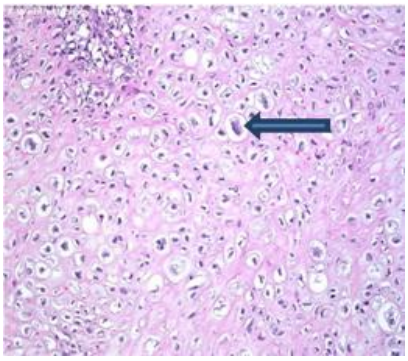

Figure 4: Histopathology of mesenchymal chondrosarcoma.
Blue arrow shows small to round cells with hyper chromatic to vesicular nuclei.

Discussion

Mesenchymal chondrosarcoma is thought to originate from cartilage precursor cells, or chondroblasts, that have failed to develop into mature chondrocytes. It is regarded as a high-grade sarcoma in the grading systems of the French Federation of Cancer Centers Sarcoma Group, the National Cancer Institute [7] and our patient had grade II according to this system. This tumor affects children and young adults, usually between the ages of 15-35 years, like in our case our patient was 35 years old. It presents with swelling and pain. In our case it presented with swelling of cheek and paresthesia due to involvement of Infratemporal fossa. Radio graphically, chondrosarcoma is seen as an irregularly shaped, ill-defined radiolucency with randomly. CT performed with contrast enhancement is the imaging modality of choice because it best delineates the bony involvement while defining local invasion into the adjacent soft tissues as seen in our case [8].

Mesenchymal chondrosarcoma can present with metastases or tumor that has spread to other parts of the body through the blood stream. Usually, patients feel symptoms from their primary tumor before they feel anything from their metastases. The most common site to which the tumor spreads is the lungs. Therefore it is important to do metastatic workup. In our patient metastatic workup showed no distant metastasis. Wide surgical excision is the mainstay of treatment for MC in the bones [9]. These tumors are radio resistant and chemotherapy can be used as an adjuvant therapy after wide surgical excision is made. The benefit of neo-adjuvant modalities for MC has not been proven. The overall survival rate at 5 and 10 years is 55% and 27% respectively [10]. Our case was treated with wide surgical excision followed by chemotherapy. Generally the prognosis of MC is considered to be poor. The grade of tumor, size and the adequacy of tumor resection margins are prognostic indictors. Local recurrence may occur hence adequate treatment and long-term follow-up is required which include periodic systemic evaluation.

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