Huge Mesenchymal Chondrosarcoma of Mandible

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Abstract Mesenchymal Chondrosarcomas [MCS] are rare subtype of chondrosarcoma. They are distinct tumors arising in unicentric or multicentric locations. It usually appears in second to third decade of life. The neoplasm affects female more frequently than in males. They reveals unusual clinical behavior, characteristic male histopathological features, and poor prognosis with late recurrences. We report a rare case of MC arising in a 19 year male patient.

Keywords Mesenchymal chondrosarcoma · Mandible · Maxilla · Swelling · Neoplasm

Introduction

Mesenchymal chondrosarcomas (MCs) are rare malignant neoplasms that can arise from both soft and hard tissues [1]. It was first described by Lichtenstein and Bernstein in 1959 as a distinct variant of chondrosarcoma from which it must be differentiated [2]. It usually appears in second and third decade of life [2]. Females are more frequently affected with MC as compared to males [3]. Facial bones and ribs are the commonest bones to be involved in this disease. MC is characterized by sheets or clusters of highly

undifferentiated small ovoid cells that are intermixed with small zones of neoplastic cartilage [1]. It commonly presents as a painless mass or swelling, although a painful mass may also be a presenting symptom. Neurological symptoms like paresthesia may also be present in few cases. This neoplasm shows aggressive local behaviour as well as a high metastatic potential [3]. The long term prognosis is extremely poor. Local recurrence and distant metastasis have also been reported [4].

Case Report

A 19 years male was referred to department of oral and maxillofacial surgery, CSM Medical University, Lucknow, with complaint of swelling in anterior region of the lower jaw. He was operated one year back in the ENT department, excision of lesion was done along with extraction of four incisors teeth. The tumor recurred once again after 6 months after the excision.

On examination the patient was well built, nourished and conscious. External examination showed a huge lump [7 × 8 × 8 cm] in size extending from left to right body of the mandible, with normal colour and texture of the overlying skin (Fig. 1). On palpation swelling was non tender, firm in consistency, non compressible and non pulsatile with smooth surface. Intra-oral examination showed swelling extending from right lower second premolar to left first molar involving labial and buccal vestibules. Mucosa over mass was reddish in colour. Both right and left submandibular lymphnodes were palpable, freely movable and measuring about 2 × 1.5 cm.

Laboratory investigations showed a normal haemogram. Serum alkaline phosphatase levels were within the normal range. OPG and CT scan of the mandible and neck

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Fig. 1 Pre-operative photograph

Fig. 2 Pre-operative radiograph

demonstrated a heterogeneous massive lesion with expansion of the bone and invasion of soft tissues (Fig. 2, 3, 4). X-ray chest and ultrasonography of the whole abdomen did not show any pathology except a 6 mm cyst in prostate.

A diagnostic incisional biopsy was done which revealed a tumor with biphasic pattern comprising of undifferentiated short spindled cells admixed with small area of hyaline cartilage, blended gradually with the undifferentiated areas. The short spindled cells showed predominantly a hemangiopericytomatous vascular pattern, with foci of myxoid changes, suggestive of mesenchymal chondrosarcoma (Fig. 5). After diagnosis was established, wide surgical resection, was done along with enlarged lymph nodes under general anaesthesia (Fig. 6), and a reconstruction plate was placed (Fig. 7). Histopathological examination of the excised specimen confirmed the previous diagnosis. No chemotherapy or radiotherapy was given preoperatively and postoperatively. No recurrence was seen after 2 years of follow-up. (Fig. 8)

Discussion

Mesenchymal Chondrosarcomas are, rare subtype of chondrosarcoma, constituting approximately 3–9% of all

Fig. 3 Axial section of the lesion

Fig. 4 Coronal section of the lesion

chondrosarcomas (5). Mesenchymal Chondrosarcomas usually presents as a painless mass or swelling 53%, although a painful mass is also a frequently reported presenting symptom 16%. Neurological symptoms like paresthesia and paresis may also occur (5).

Chondrosarcomas of the head neck occurs most frequently in the third to sixth decade of the life as compared to mesenchymal chondrosarcoma, which usually occurs in second to third decade of life (6). Common radiological findings of mesenchymal chondrosarcoma of the jaw are radiolucency but there may be radiopaque or mixed radiopaque and radiolucent image (7). Radiographically, it is difficult to distinguish between mesenchymal chondrosarcoma, other cartilaginous neoplasms or osteogenic

Fig. 5 Histopathology of the lesion

Fig. 8 Post-operative photograph of patient

Fig. 6 Excised specimen

Fig. 7 Post-operative OPG showing reconstruction with bone plate

sarcoma, thus there is no characteristic radiographic appearance (3).

The histological feature of mesenchymal chondrosarcoma is characteristically a biphasic picture. Undifferentiated areas appear as sheet of primitive mesenchymal cells, similar to small cell anaplastic sarcoma. However, island of

well-differentiated cartilaginous tumor enable the pathologist to make the specific diagnosis of the mesenchymal chondrosarcoma. Rich vascular component with short blue spindle cells in the mesenchymal component cause confusion with hemangiopericytoma [5]. In some cases initial diagnosis has been reported as Ewing's sarcoma, and Osteosarcoma [8]. So it is essential, that multiple sections should be examined to make a definite diagnosis [1].

The most effective therapeutic modality is wide surgical excision, with a tumor free margin of 2–3 cm [10]. Extensive resection has less recurrence and a better survival rate than limited surgical resection [1]. There are also reports of resolution of this tumor with chemotherapy and radiation alone [4]. The benefit from chemotherapy and radiation is still unclear [9].

The prognosis for mesenchymal chondrosarcoma is poor because tumours have tendency to recur either locally or as metastases [9]. Most common site of the metastases is the lung. Survival time is longer with maxillary than with mandibular lesions [5]. Survival time varies between 5–10 years. Overall the reported 10 year survival rate is 28% [4]. In our case recurrence occurred at 6 months after local excision. It explains that excision with wide margins should be done in cases diagnosed by incisional biopsy.

MCS is an uncommon tumor and rarely found on symphysis of the mandible. To best of our knowledge only 45 cases of MCS of jaws has been reported in literature till now, 23 in maxilla and 22 in mandible we are reporting 23rd case of MCS in mandible, which arised form symphysis of mandible and extended to both body of mandible. An adequate incisional biopsy is required for making a preoperative diagnosis. The lesion may be radiolucent, radio-opaque or mixed. The tumor requires surgical excision with wide margins. The role of pre and post operative radiotherapy and chemotherapy is unclear and as of current date complete excision with wide margins, is the treatment of choice.

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