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Case Report

Chondrosarcoma of the Mandible - A Rare Case Report

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ABSTRACT

Chondrosarcomas are the third most common malignant bone neoplasm. It has now been recognized that it is not a stereotypical disease but has variants. Although any portion of the skeleton may be affected, those involving the jawbones and the skull are decidedly unusual. A conventional chondrosarcoma of retro molar region in a 51-year-old female patient has been reported here.

Key-words: chondrosarcoma, conventional chondrosarcoma, mandible, jaws.

INTRODUCTION

Chondrosarcoma, as defined by WHO is a malignant tumor with pure hyaline cartilage differentiation. [1] They commonly arise in the central part of the skeleton, including pelvis, shoulder and ribs. In the facial skeleton, they account for 1-3% of all chondrosarcomas and arise primarily in the maxilla with a predilection for the anterior maxillary region. [2,3] Here, we report a case of chondrosarcoma of left mandible in the retro molar area in a 51-year-old female patient.

CASE REPORT

A 51-year-old female patient reported to the department with a diffuse swelling on left side of the face. The swelling extended from distal aspect of tooth 36 to retro molar pad area. Overlying skin appeared normal. On palpation, it was soft to firm in consistency and mildly tender. Intra orally, obliteration of buccal and lingual vestibules and expansion of buccal and lingual cortices were noted. Grade one mobility of teeth 35 and 36 were noted. Patient had a history of extraction of mobile tooth 37, three months back. The swelling was slow growing with no difficulty in tongue movements, trismus or numbness of lip or tongue. Bilateral submandibular lymph nodes were palpable but non tender.

CT showed a heterogeneously enhancing area involving left retromolar region of size 2.6 x 1.0 cm with hypodense lingual area showing rim enhancement, mild sclerotic changes in mandible and homogenously enhancing enlarged submandibular lymphnodes bilaterally.

An incision biopsy was performed and a pathological diagnosis of chondrosarcoma was made. The patient was posted for surgical excision of the lesion under general anaesthesia. Hemimandibulectomy was done. The lesional tissue obtained was an ivory white coloured soft to firm mass and was friable in nature (figure 1).

The H and E stained sections revealed a highly cellular connective tissue stroma with chondroid and non chondroid areas. The chondroid areas showed a hyalinised matrix with lacunae, most of which were occupied by chondrocytes. Nuclear hyperchromatism, pleomorphism and binucleation of the chondrocytes and foci of calcification were noted. The non chondroid areas showed proliferating spindle cells with nuclear pleomorphism and occasional mitotic figures (figure 2 & 3). The chondrocytic area showed diffuse strong positivity for S100 (figure 4). Vimentin and cytokeratin AE1/AE3 were negative. A diagnosis of grade 2 chondrosarcoma was made.

Post operative course of the patient was uneventful. A follow up period of 20 months didn't show any recurrence.

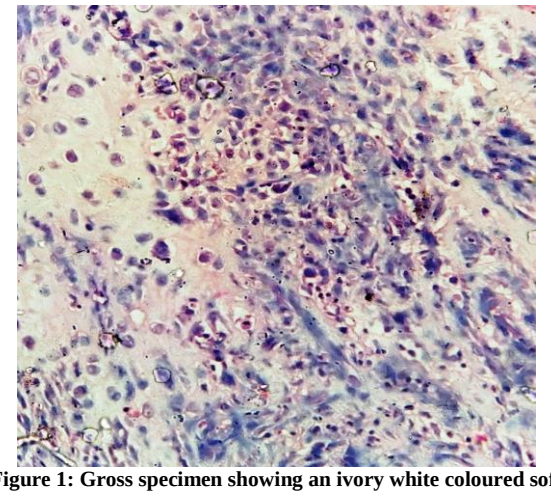

Figure 1: Gross specimen showing an ivory white coloured soft to firm mass, along with resected mandible.

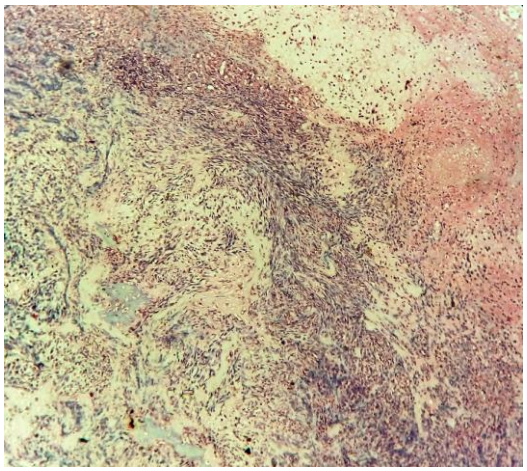

Figure 2: Photomicrograph showing chondroid and hypercellular non chondroid areas containing spindle cells (H/E 10x).

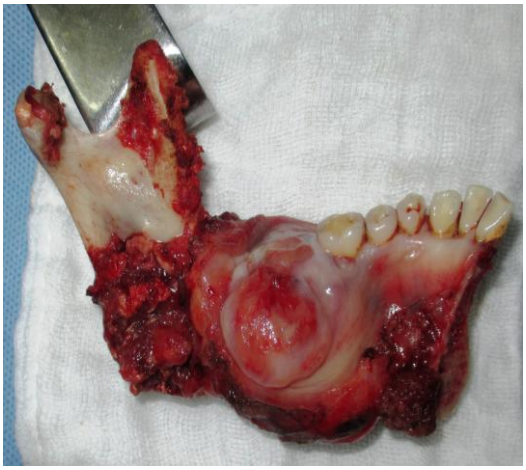

Figure 3: Photomicrograph showing atypical chondrocytes with nuclear pleomorphism and hyperchromatism (H/E 40x).

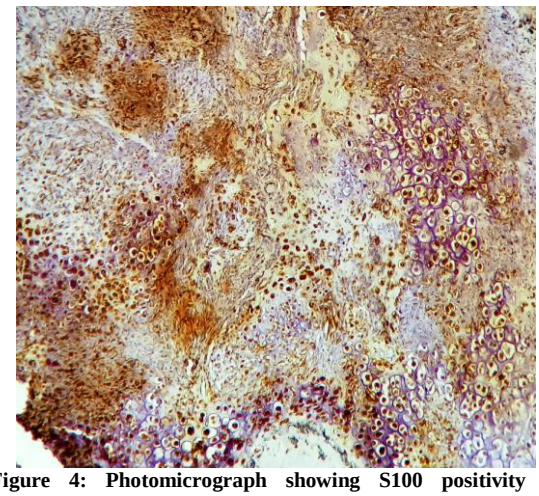

Figure 4: Photomicrograph showing S100 positivity of chondrocytes (40x).s

DISCUSSION

The tumor was recognized as a distinct entity in 1930 when Phemister stated that sarcomas, involving bone, which contained cartilage, were chondrosarcomas. [2] Conventional chondrosarcomas occur primarily in adults with a male to female ratio ranging from 2 to 3.7:1. [4] The most common sites are pelvis, proximal femur, proximal humerus, distal femur and ribs. Out of the minority of cases in facial skeleton, the mandible accounts only for 10%, [5] involving mostly the molar region and mandibular symphysis.

The tumor may originate from primitive mesenchymal cells, embryonal rests from cartilaginous matrix of cranium or a pre existing Paget's disease or fibrous dysplasia of bone. [4,6] Since the patient here failed to give any history of such pre existing lesions, it may have developed

from normal chondroid tissue or from embryonic cartilaginous rests.

The pathogenesis of chondrosarcomas remains poorly understood. Conventional cytogenetic analysis shows the tumor to be typically near diploid, but higher grades may show complex karyotypes. Aberrant signaling of the Indian hedgehog- parathyroid hormone-like hormone pathway, activated Src and Akt kinase signaling, functional NRAS and isocitrate dehydrogenase (IDH) mutations, alterations in pRB and derangements in the hypoxic and glycolytic pathways has been implicated in chondrosarcoma. Mutations affecting the coding sequence of COL2A1 were found in central chondrosarcomas, suggesting that matrix associated type II collagen may play a role in its formation. [7]

Categorized generally by their location within the parent bone, chondrosarcomas are of two types – central and peripheral. Central/primary or conventional chondrosarcomas arise de novo within the intramedullary cavity of a normal bone, whereas peripheral/periosteal chondrosarcomas arise on the surface of cortical bone, most commonly within a pre-existing osteochondroma or enchondroma (secondary chondrosarcoma). The International Classification of Diseases for Oncology classified chondrosarcomas into six types: clear cell, myxoid, dedifferentiated, juxtacortical, mesenchymal and chondrosarcoma not otherwise specified.

Chondrosarcomas of the mandible present most commonly as a painless swelling of long duration, with symptoms of pain, paresthesia, trismus and loosening of the teeth developing as the disease progresses. [8] Radiography is usually non pathognomonic. Single / multiple radiolucencies with ill defined borders and variably distributed punctate opacities, cortical destruction, fusiform expansion and thickening of the cortex, uniform periodontal ligament space widening and root resorption of the adjacent teeth have been reported. MRI helps in delineating the extent of the tumor and establishing soft tissue extension. CT scans aid in demonstrating matrix calcification.

Macroscopically, the cut surfaces show a translucent, blue-grey or white color corresponding to the presence of hyaline cartilage. Microscopically, chondrosarcomas shows abundant blue-grey cartilage matrix production. Three histological grades, primarily depending on cellularity, size and hyperchromasia of the tumor nucleus are noted.

- Grade 1(low grade) tumors are characterized by moderately cellular stroma containing hyperchromatic plump nuclei of uniform size. Occasionally binucleated cells are present. Mitotic figures are absent.
- Grade 2(intermediate grade) tumors are more cellular and contain a greater degree of nuclear atypia, hyperchromasia and nuclear size. Mitotic figures are occasionally seen.
- Grade 3(high grade) lesions are more cellular and pleomorphic and atypical than grade 2. Mitoses are easily detected with a count of 2 per 10 high power fields.
- The malignant tumor that has areas of spindling should be considered grade 4 and called either chondroblastic osteosarcoma or dedifferentiated.

The main differential diagnoses include chondroma, mesenchymal chondrosarcoma and chondroblastic osteosarcoma. Chondromas are usually hypocellular with minimal cellular atypia. Also, tumoral pain accompanied with radiologic findings of deep endosteal scalloping, cortical destruction and soft-tissue mass on CT or MRI, periosteal reaction facilitate the diagnosis of chondrosarcoma over chondroma. [9] Negative staining for brachyury and cytokeratin AE1/AE3 is a powerful identifier of chondrosarcoma over chondroma. [8] Mesenchymal chondrosarcoma usually peaks at second and third decades and typically presents a bimorphic pattern, composed of highly

undifferentiated small round cells and islands of well differentiated hyaline cartilage. Chondroblastic osteosarcoma can be excluded by the expression of alkaline phosphatase by the tumor cells and absence of osteoid matrix, bony trabeculae and IDH mutation.

Chondrosarcomas are radio-resistant lesions. Radical surgical excision is the treatment of choice. As distant metastases to lymph nodes are rare, neck dissection is not obligatory. The prognosis appears to be related to the location of the tumor, the adequacy of the primary surgical resection and the histological grade of the neoplasm. The five-year survival is 89% for patients with grade 1; the combined group of patients with grade 2 and 3 has a five-year survival of 53%. [10]

CONCLUSION

Chondrosarcomas of jaw bones is rare and present a poorer prognosis compared to other parts. Since the tumor shows a wide variation in time of recurrence and metastasis, it is warranted that the patient should be kept in long term and periodic surveillance.

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