Marginal resection in a case of juxtacortical chondrosarcoma of the mandible

ABSTRACT
Chondrosarcomas usually invite extensive surgical resection. These are malignant tumors of cartilaginous origin and their involvement in the maxillofacial region is rare. Juxtacortical chondrosarcoma, a distinct malignant cartilage-forming tumor arising from the external surface of a bone, is extremely rare in the mandible. Here, one such case with a relatively good prognosis is presented where marginal resection as a surgical option yielded good result.

Keywords: Chondrosarcoma, juxtacortical, mandible, marginal resection

INTRODUCTION
Chondrosarcomas commonly occur in the long bones such as femur, humerus, pelvis, and sacrum. Chondrosarcoma of maxillofacial region is rare, mandible being even rarer. These malignant tumors arise from primitive mesenchymal cells.

According to the location, chondrosarcomas are classified as central, peripheral, and juxtacortical chondrosarcomas.

Juxtacortical chondrosarcoma is a distinct entity typically manifesting as a slow-growing, painless mass from the external bony surface. It accounts for <2% of all chondrosarcomas. This tumor usually involves the shaft of long bones, especially the femur. In the maxillofacial region, only a few cases of juxtacortical chondrosarcoma affecting the mandible, other than the symphysis region, have been reported so far.

CASE REPORT
A 43-year-old male reported with a swelling in the lower jaw of 8 months’ duration, in the mandibular symphysis region with no pain, trismus, or paresthesia associated with the swelling. Examination revealed an ulcerated erythematous mass arising from the lower anterior alveolar region from canine to canine and obliterating the labial vestibule. The mass was hard in consistency, nontender, and seemed fixed to the mandible. Tongue movements were normal and the floor of the mouth was uninvolved. No regional lymphadenopathy was present. Metastasis was evaluated by ultrasonogram and computed tomography (CT) with negative results.

The mandibular basal bone appeared intact on orthopantomogram.

CT images revealed a well-circumscribed hypointense area of 3.3 cm × 3.3 cm × 2.9 cm in the symphysis region extending from the labial cortex into the soft tissue. Within this, patchy...
hyperintense areas were noted. There was thinning of the labial cortex with evidence of minimal cortical involvement in the symphysis region [Figure 2].

Taking into consideration the radiological appearance of intact basal bone and the low-grade nature confirmed histologically (incisional biopsy), plan was made for a marginal resection. Intraoperatively, the mass was well demarcated and firm in consistency. The labial alveolar bone was minimally invaded by the tumor. The cortical bone that bordered the lesion was clinically intact. Adequate clearance of 2.5 cm of bony and soft-tissue margin was given [Figure 3]. Postoperative histopathological evaluation revealed that the bony and soft-tissue margins were free of tumor cells. No complication has been reported in the 24-month follow-up. No additional radiotherapy or chemotherapy was given.

**Histopathological findings**

Biopsy specimen consisted of lobules of chondrocytes separated by thin fibromyxoid stroma. The chondrocytes show varying degrees of differentiation with areas of malignant cartilage production. Cords of tumor cells, mostly spindle shaped, were seen infiltrating the stroma. The cells in some areas showed atypia with hyperchromatic nuclei. Mitotic figures were identified easily [Figure 4]. The final diagnosis was given as low-grade chondrosarcoma. The surgical margins were free of tumor cells.

**DISCUSSION**

The term juxtacortical chondrosarcoma was introduced by Jaffe in 1958.[6] Juxtacortical chondrosarcoma mainly affects the shaft of the long bones, especially the femur.[3] Juxtacortical chondrosarcomas are extremely rare in the jaws. Only a few cases of juxtacortical chondrosarcoma of the mandible have been reported till date.

Juxtacortical chondrosarcoma typically presents as a slow-growing, smooth painless lump arising from the surface of the bone without the medullary component. The clinicopathologic characteristics include peak occurrence at approximately 20 years of age with a range of 15–65 years, males being affected more. Radiographically, the cortex is intact and sometimes thickened and there may be minimal tumor invasion into the cortex without medullary involvement. Histopathologically, it is composed of poorly differentiated malignant cartilaginous tissue. Local recurrence and distant metastasis is rare.

CT image of juxtacortical chondrosarcoma appears as a well-defined hypodense mass within which there are multiple areas of fine and coarse calcifications. Soft-tissue lesions appear as hypodense mass, with stippled radio-opacity representing small foci of cartilage formation which appears as a ring- or crescent-shaped calcification. The lesion is
present on the surface of the bone. The medullary cavity is characteristically not involved. [4]

In this case, CT scan clearly demonstrated the absence of medullary involvement and extensions into the soft tissue with areas of spotty calcifications (hyperintense patches). These findings were consistent with the classic imaging aspects described for juxtacortical chondrosarcoma of long bones.

Reinforcing the diagnosis of juxtacortical chondrosarcoma in our case were the factors such as the lesion running a slower, less aggressive course and absence of radio-opaque striations (spicules) running perpendicular to the underlying cortex which is a common feature of osteosarcoma. [7]

Histologically, juxtacortical chondrosarcoma exhibits features similar to that of conventional chondrosarcoma. Resected specimen shows chondrocytes in varying degrees of differentiation and cords of spindle-shaped tumor cells infiltrating the fibrous stroma. [2]

The treatment option of chondrosarcoma is multimodal: wide en bloc resection, local curettage, cryotherapy, chemotherapy, radiotherapy, and immunotherapy. Treatment mainly depends on the following: (1) size of the tumor, (2) extent and location of the tumor, and (3) histological grading. Grade 1 and Grade 2 chondrosarcomas of the jaws and facial skeleton are best treated with local resection, giving a clearance of 1.5-cm margins of bony and soft tissues. [8] Chemotherapy and radiotherapy are not indicated as the primary treatment. Grade 3 chondrosarcomas are treated with aggressive resection followed by chemotherapy and radiotherapy. [2]

In this case, the surgical treatment planned was marginal resection of the anterior mandible based on the grade (histopathology), lack of intramedullary involvement (radiographic), and clinically intact basal bone (surgical exposure), enabling 2.5 cm of hard- and soft-tissue clearance.

Prognosis of juxtacortical chondrosarcoma of long bone seems to be better than that of central variety of similar grade. Prognosis in maxillofacial region depends on factors such as the direct extension of the tumor to the skull base and distant metastasis. However, metastasis is rare, which occurs only in high-grade or recurrent cases. [4] A 10-year follow-up of the case reported by Van Damme et al., 2005, showed a tumor-free course, thus showing the indolent nature of this tumor. [1]

Our follow-up was for a period of 35 months, and no clinical and radiological evidence of recurrence or metastasis was noted during this period.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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