Juxtacortical osteosarcoma of the mandible: Challenges in diagnosis and management

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ABSTRACT

Parosteal osteosarcoma (OS) of the jaw is a rare type of OS with peculiar clinical radiographic and microscopic features. The aim of this article was to report and discuss a case of high-grade parosteal OS in the mandible of a 35-year-old woman. The patient reported sensing mild pain and swelling in the retro molar area on the left side of the mouth for a period of 4 years, despite continuous dental treatment. The radiographic evaluation showed a mixed radiopaque/radiolucent lesion in the body of the left side of the mandible. Destruction of the mandibular cortex in that area was also observed. After the initial histological study, the patient underwent partial hemi-mandibulectomy. Microscopic findings showed a tumor exhibiting spindle cells with nuclear hyperchromasia, moderate pleomorphism, and irregular osteoid formation, with chondroid differentiation noted with tumor-free margins. The immunohistochemical analysis showed the expression of negativity to p53, human epidermal growth factor receptor 2/neu, and positivity to S-100. The diagnosis was high-grade parosteal OS of the jaw. The 4 years clinical and imaging postoperative follow-up showed no evidence of recurrence. The literature on this unusual pathologic entity reviewed and diagnostic challenges described.

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Key words: Mandible, oral osteosarcoma, parosteal or juxtacortical variant

Introduction

Osteosarcoma (OS), a rare malignant bone tumor arising from primitive bone forming mesenchyma, most often arises in the metaphyses of long extremity bones.[1] Craniofacial OS (CFOS), most often located in the mandible or maxilla,[2,3] accounts for only 6–13% of all OSs.[4-7]

The term OS refers to a heterogeneous group of primary malignant mesenchymal neoplasms showing evidence of osteogenic differentiation.[8] In general, OSs of the jaws are high-grade lesions. Low-grade lesions are rare and include the well-differentiated intramedullary OS (low-grade central OS) and parosteal OSs.[9]

Whereas classical OS of the long bones most often affects adolescents and young adults, CFOS typically occurs in the third or fourth decade of life.[4,5,10-13] Presenting signs and symptoms of CFOS include regional swelling, pain, and paresthesia. Patients may complain about changes in tooth position, loose teeth, or notice a change in the fit of a dental prosthesis. Many of these signs and symptoms are nonspecific, and there is often a considerable delay before the correct diagnosis is made.[14,15]

Osteosarcoma involving the maxillofacial region challenges many clinical specialists. With the exception of skull tumors, CFOS metastasizes less frequently than OSs of other sites.[4,10,11] Similar to OS of the extremities, adequate surgical resection is considered a mainstay of treatment. Local recurrences and intracranial invasion have been reported as the main causes of treatment failure due to anatomical complexity, which sometimes makes tumor resection incomplete.[4,16,17]

Access this article online

Quick Response Code:

Website: www.njms.in

DOI: 10.4103/0975-5950.168232

The introduction of neoadjuvant chemotherapy has revolutionized the treatment of extremity OSs, increasing cure rates from approximately 10% to 60–70%.[18] The role of chemotherapy in CFOS is less clear and meta-analyses of published data have reported conflicting results.[19,20]

A tumor is defined as craniofacial if it is situated in the jaws (mandible or maxilla), or any of the extra gnathic bones. Among the extra gnathic bones, OSs of the skull includes parietal, temporal, and occipital bones and bones of the orbital cavity. Tumors involving several craniofacial bones, the tumor site, were classified according to the region of presumed origin. Recommended procedures used to define the extension of primary tumors of any site includes conventional radiography, whereas the availability of other methods (e.g., computed tomography (CT) and magnetic resonance imaging) varied with time. The minimum requirement for the exclusion of primary metastases is a negative chest X-ray.

Report of Case

A 35-year-old woman was referred by a private practitioner for evaluation and treatment of an asymptomatic enlarging mass of 4 years duration in the left mandible. Oral examination disclosed sessile, strawberry-like hard mass on the lingual mucosal surface of the first molar region of the left mandible extending to the attached gingival margin which was pushing the retromolar tissue toward midline obliterating the left side of the soft palate and anterior faucial pillar [Figure 1a and b]. The overlying mucosa appeared to be hyper vascular but was not ulcerated. There was neither tenderness nor paresthesia.

Radiographic examination

Computed tomography examination showed an erosive bony change in the posterior lingual cortical plate of the left mandible in all multiplanar sections. Large irregular bony growth attached to the medial wall of the left mandibular angle with soft tissue component enveloping exostotic growth. Approximate size of the mass was 55 mm × 25 mm. Mass is seen displacing surrounding soft tissues. Fat planes and adjacent soft tissues were grossly preserved. Distortion of the oropharynx with obliteration of left side vallecula noted. There was periosteal new bone formation with no evidence of medullary involvement [Figure 2a]. Axial section of CT at the level of molar roots showing sunray appearance, which is intruded into lingual tissue [Figure 2b]. Coronal section of CT mandible showing an intruding radiopaque mass in retromolar lingual area obliterating the normal oropharynx. Panoramic radiograph showing mixed radiolucent and radiopaque lesion involving the molar teeth giving the classic feature of sunray appearance [Figure 2c] suggestive of OS.

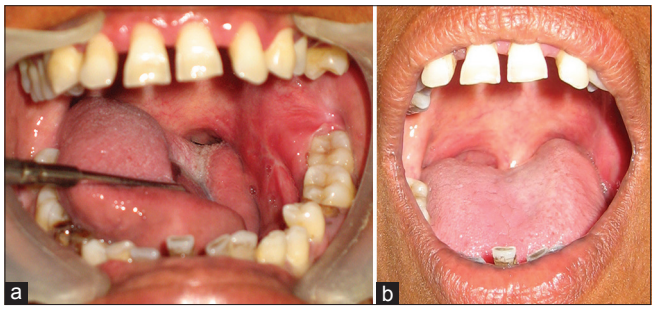

Figure 1: (a) Preoperative intraoral photo showing swelling in the retro molar area pushing posteriorly. (b) Intraoral photo showing lesion pushing the anterior faucial pillar to opposite side

Microscopic examination

Initial microscopic examination of an incisional biopsy specimen revealed that the tumor consisted exclusively of lobules of cartilaginous tissue with small foci of malignant osteoid formation. In addition, mitotic figures were common among the chondrocytic and osteocytic cells.

Diagnosis and treatment

The diagnosis of juxtacortical or parosteal OS was made based on the presenting clinical features, correlated histologic findings, and the CT images. Subsequently chest X-ray and abdominal ultrasonogram showed no evidence of metastatic disease. The patient underwent a partial mandibulectomy under general anesthesia [Figure 3a-c] using modified lip split incision and immediate reconstruction using stainless steel reconstruction plate [Figure 3d]. The patient has been followed up regularly for 48 months. To date, there has been no evidence of recurrence. Postsurgical adjuvant chemotherapy was recommended.

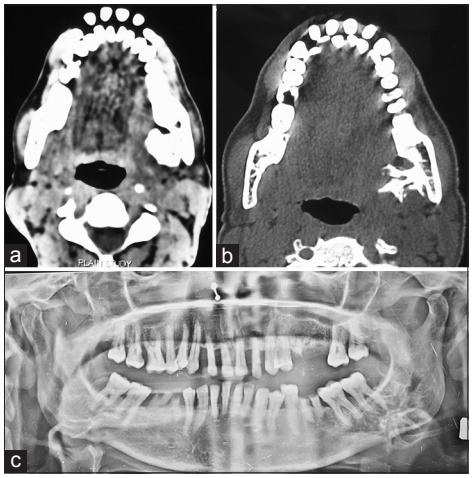

Figure 2: (a) Axial section of computed tomography (CT) mandible showing extruding lesion on the medial aspect of the mandible below the last molar. (b) Axial section of CT at the level of molar roots showing sunray appearance, which is intruded into the lingual tissue. (c) Panoramic radiograph showing the radiolucent lesion in relation to left side molar teeth

Gross examination of surgical specimen

Cut sections of the resected mandible and tumor revealed a grayish-white nodular mass located primarily within the external surface of the lingual bone cortex. The tumor was responsible for the focal destruction of the alveolar bone crest with minimal extension into the periodontal ligament space. However, no evidence of medullary involvement was noted. The left-sided partial mandibulectomy specimen with attached soft tissue, and part of submandibular salivary gland measured 6.4 cm × 4 cm × 7.5 cm in size [Figure 4]. The lingual surface of mandible showed 1.8 cm × 1.9 cm × 0.9 cm sized tumor. The radiographic examination of gross specimen clearly showed classic "sunburst" or "sunray" appearance [Figure 5].

Histopathological examination

The resected tumor demonstrated histologic features identical to those seen in the initial biopsy specimen; namely chondroid matrix admixed with malignant osteoid sheets showing foci of calcification. Pleomorphic spindle, epitheloid, plasmacytoid, fusiform, ovoid cells with increased nuclear-cytoplasmic ratio and hyperchromatism with nuclear atypia. The cells exhibit a moderate pleomorphism and exhibit 3–4 MF/10 HPF. The adjacent bone cortex was free of tumor. Photo micrographic features of H and E stains are conclusive of OS with a grade of 2/4.

Immune histochemistry

The tumor area exhibiting chondroid differentiation shows S-100 positivity, p-53, and human epidermal growth factor receptor 2 (HER-2)/neu negativity [Figure 6a-f].

Discussion

Osteosarcoma of the jaws is uncommon; it represents only 6–8% of all OSs.[2] In the jaws, OS has different clinical and biologic characteristics than its counterpart in the long bones.[3] OS of the jaws tends to occur at an older age, and the prognosis for jaw OS is better than that of OS arising in other sites.[1] The more favorable prognosis may be due to lower mitotic activity of tumor cells and is found less often in OS of the jaws.[3] In addition, jaw OSs have less of a tendency to metastasize than OSs of the long bones.[2]

Osteosarcomas are divided into intramedullary and surface types on the basis of their individual clinical, histopathologic, and radiographic characteristics[4,5] and further classified into three subgroups: Parosteal, periosteal, and high-grade surface OS.[5] Parosteal and periosteal types tend to have a better prognosis than conventional OS or high-grade surface OS.[3,5] Both parosteal and periosteal OSs are uncommon neoplasm

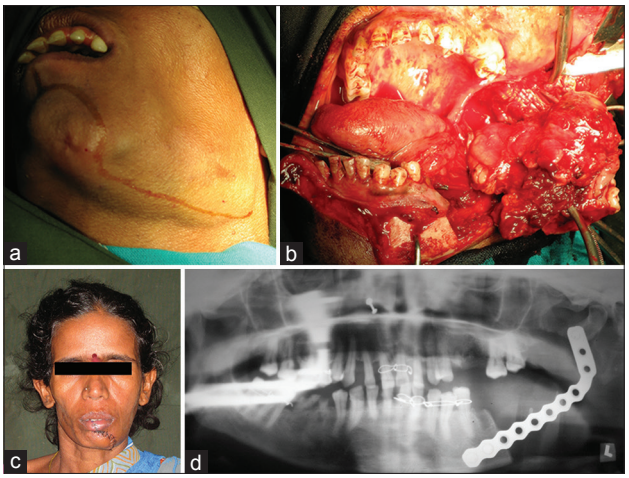

Figure 3: (a) Intraoperative skin marking for modified lip split incision for partial mandibulectomy. (b) Intraoperative photo is showing lesion *in situ* after distal mandibular osteotomy. (c) Postoperative photo is showing healed wound. (d) Postoperative panoramic radiograph showing immediate reconstruction with stainless steel reconstruction plate

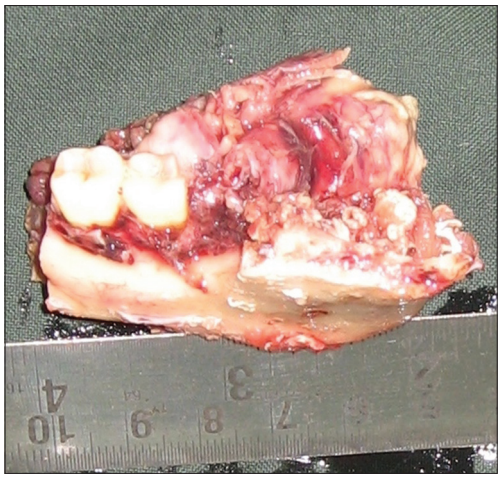

Figure 4: Excised lesion with the partial mandible

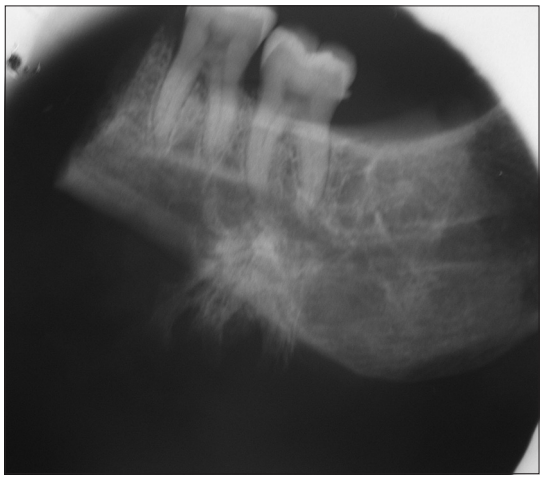

Figure 5: Specimen radiograph showing the sunburst appearance related to base of mandible in relation to last molar tooth

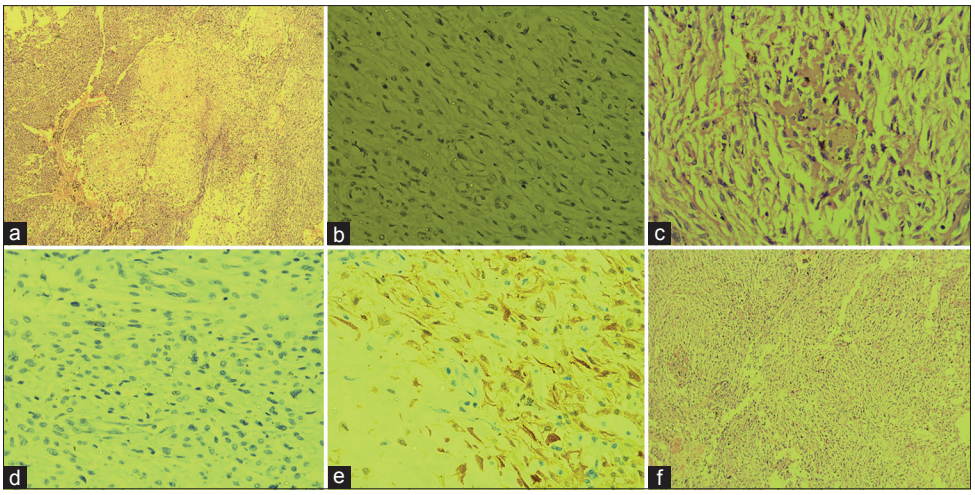

Figure 6: (a) Immunohistochemistry (IHC) microphotograph showing cartilage differentiation under ×5 magnification. (b) IHC microphotograph showing human epidermal growth factor receptor 2 negativity under ×40 magnification. (c) IHC microphotograph showing osteoid production by tumor cells under ×40 00003. (d) IHC microphotograph showing p53 negativity under 40×00008. (e) IHC microphotograph showing S-100 positivity in areas of cartilage differentiation 40×00006, (f) IHC microphotograph showing spindle tumor cells ×10

and comprise approximately 5% of all OSs. These are confined to the jaws exclusively.[2-5] In our case a parosteal high-grade tumor was seen with a better prognosis.

Osteosarcoma is a malignancy of mesenchymal cells that have the ability to produce osteoid or immature bone. The origin of these tumors is largely unknown. Jaw OS are even rarer, accounting for 4% of all documented cases.[14] In the jaws, the biologic behavior of OS differs from that of tumors involving other skeletal bones. The average onset age of OS of the jaw is 10–20 years later than that reported for skeletal lesions, and survival rates are higher.[21,22] However, in our case the age predilection is third to fourth decade.

S-100 positivity shows the malignancy is of mesenchymal origin. HER-2/neu is epidermal growth receptor also known as ErbB2.[23] It is tyrosine kinase oncogene. Overexpression of this oncogene is related to poorer prognosis and overall poor survival rate.[23] The HER-2/neu presence is still controversial in OS;[23] similarly p-53 overexpression correlated with a worse prognosis in CFOSs.[24] However, in our case tumor is negative for HER-2/neu and p-53 expression which shows the good prognosis. At the time of presentation, we successfully followed the case without any recurrence for over 4 years. Careful attention to clinical, radiographic, and microscopic findings is necessary to ensure the establishment of the correct diagnosis, thus helping to prevent incorrect treatment and unreliable and sometimes dire outcomes.

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.

Acknowledgments

Dr. Sanjay Navani surgical pathologist and immunohistochemist LAB SURGPATH, Mumbai, India for IHC study with report.

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How to cite this article: Chakravarthi PS, Kattimani VS, Prasad LK, Satish PR. Juxtacortical osteosarcoma of the mandible: Challenges in diagnosis and management. Natl J Maxillofac Surg 2015;6:127-31.

Source of Support: Nil. **Conflict of Interest:** None declared.