Osteosarcoma of Mandible

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ABSTRACT
Osteosarcomas (OS) are malignant neoplasms of the bone that commonly affect the long bones with rare presentation in jaws. Osteosarcomas of jaws represent about 6 to 8% of all OS, with an incidence of approximately 1 in 1.5 million persons per year. Although the exact cause of OS is still unknown, defects in the retinoblastoma (RB) and p53 genes play an important role in the process. It is characterized histologically by anaplastic stroma with direct osteoid production. Here, we report a case of OS in a 30-year-old female, who came with a massive bony swelling in the right mandibular region.

Keywords: Osteoblastic variant, Osteosarcoma, Sunburst appearance.

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INTRODUCTION
The term “osteosarcoma”, also known as osteogenic sarcoma (OS), refers to a heterogeneous group of primary malignant neoplasms affecting bone forming or mesenchymal tissue that is characterized by formation of osteoid tissue.1 It occurs most commonly in long bones of extremities near metaphyseal growth plate. Osteogenic sarcoma of jaw is rare and represents only 6 to 8% of all OS. Jaw OS usually presents themselves in the 3rd and 4th decades of life, almost a decade after their presentation in long bone tumors with a slight predilection for the mandible.2 The exact etiology is unknown. Three main factors generally may play an important role in their development—irradiation, preexisting benign bone disorders, and genetic predisposition. Biologically, OS of the jaw is considered to be less aggressive with a lower incidence of metastasis and hence better prognosis than that occurring in long bones.3 Despite modern treatment protocols that combine chemotherapy, surgery, and sometimes radiotherapy, the 5-year survival rate for patients diagnosed with OS remains at 50 to 70%.4,5 Here, we report a case of a 30-year-old female with mandibular bony swelling diagnosed as OS.

CASE REPORT
A 30-year-old female reported to Department of Oral Pathology and Microbiology, MGM Dental College and Hospital, Navi Mumbai, with a chief complain of swelling in lower right back region since 8 months. Patient was asymptomatic 8 months ago but started experiencing a swelling with respect to lower right half of the face. Swelling was small initially. Patient visited a local dentist who extracted a tooth from lower right back region. Swelling had gradually increased to its present size. No history of trauma was noted. There was no history of any other disease affecting the jaw or other bones. Medical and family history were noncontributory.

On extraoral examination, a diffuse swelling was present on the right side of face, extending from the zygoma inferiorly 2 to 3 cm below the chin. Anteriorly, it extended from just beyond the midline (left side) to the right preauricular region (Fig. 1). Anteroposteriorly, it measured about 12 × 9 cm in size. Overlying skin was normal but tensed and not associated with any sinus or fistula. Borders of the swelling were poorly demarcated (Fig. 2). There was no evidence of extraoral draining sinus. Temperature of the overlying skin was normal. An intraoral examination revealed an ill-defined, diffuse, ulceroproliferative lesion showing bicortical expansion obliterating the buccal and lingual vestibular spaces on the right side of the mandible. The soft tissue growth showed indentations of the maxillary teeth. Mucosa over the swelling was inflamed (Fig. 3). Floor of the mouth was raised on the right side. On palpation, the swelling was tender and hard to firm in consistency. Grade II mobility was seen in all teeth from 33 to 47. Orthopantomograph (OPG) showed a diffuse radiopacity with a sunburst appearance on right side of the mandible extending from 45 to 48 region (Fig. 4). Periodontal widening of the lower anterior teeth along with two root pieces and two missing teeth was seen. Computed tomography (CT) scan showed bone forming malignant mass of 8 × 4 cm approximately arising from right hemimandible with sunburst periosteal reaction and not breaking the lingual border of the mandible. Externally, it extended up to subcutaneous tissue. Superiortly, the lesion extended up to the level of maxillary alveolus and inferiorly up to submandibular.
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Fig. 1: Extraoral view shows a diffused swelling present on right side of face with no surface ulceration

Fig. 2: Extraoral view shows swelling extending from zygoma to 2 to 3 cm below the chin. Anteriorly it extends from just beyond the midline (left side) to right preauricular region

Fig. 3: Intraoral view shows an ill defined, diffused ulceroproliferative lesion showing indentation of maxillary posteriors, bicortical expansion obliterating the buccal and lingual vestibular spaces

Fig. 4: Orthopantomography view shows diffuse area of increased radiopacity in relation to right side of the mandible having radiating appearance at the periphery (sunburst appearance)

Fig. 5: Computed tomography scan shows bone forming malignant mass arising from right hemimandible with sunburst periosteal reaction

soft tissue (Fig. 5). Three-dimensional (3D) reconstruction image confirmed the extent of the lesion (Figs 6A and B).

Considering the clinical features, the sudden increase in size of the swelling, consistency of the lesion, and the patient's age, a provisional diagnosis of OS, fibrosarcoma, and malignant ameloblastoma were made. An incisional biopsy was performed and sent for histopathological examination.

Hematoxylin and eosin-stained soft tissue section showed hypercellular areas of spindle shaped osteoblasts with malignant tumor osteoid (Fig. 7). Malignant osteoid showed variable areas of mineralization interspersed among the tumor cells (Fig. 8). The tumor cells exhibited marked pleomorphism, hyperchromatism, and increased mitotic activity (Fig. 9). Thus, the diagnosis of osteoblastic OS was given. Hemimandibulectomy of right mandible was planned but patient refused to undergo treatment.

DISCUSSION

According to World Health Organization (WHO) 2005, OS is defined as a primary malignant tumor of bone in which the neoplastic cells produce osteoid or bones. It accounts for approximately 20% of all sarcomas and are the most common primary bone tumors excluding hematopoietic neoplasms. Approximately 6 to 8% of OS occur in the jaws. Osteosarcoma are classified as primary and secondary (Table 1). Our present case belongs to conventional osteoblastic variant of OS.
The exact cause of OS is unknown. However, a number of risk factors do exist. Osteosarcomas can arise de novo or in several preexisting bone abnormalities, such as Paget’s disease, fibrous dysplasia, multiple osteochondromas, bone infarct, chronic osteomyelitis, and osteogenesis imperfect. Exposure to radiation is an environmental risk factor. Genetic mutations in tumor suppressor gene p53 and mutated retinoblastoma (RB) gene are the other etiological factors. In patients with RB, OS occurs 500 times more frequently than in the general population.

The most common sites of OS include femur (42%), tibia (19%), humerus (10%). In jaw OS, mandible is more commonly involved than maxilla, with the mandible accounting for 44 to 73% of cases and that of in maxilla is about 27 to 56% of cases. Mandibular tumors arise more frequently in body of mandible accounting 55 to 75% of cases followed in order of frequency by the angle, the ramus, and the symphysis. Jaw OS commonly presents itself in the 3rd and 4th decades of life. A comparison of gnathic and extragnathic OS is given in (Table 2).
et al.15 did a study on 81 cases of OS and found that maxillary OS occurred in females with the ratio of 4:1, whereas mandibular lesions occurred only in males. The present case is rare, as it occurred in a 30-year-old female. The most common symptom of OS in the head and neck region is pain, swelling, mucosal ulceration, and loosening of teeth, which were also observed in our patient.

Radiographically, OS of the jaw has a purely lytic and destructive pattern (35–45%), a sclerotic pattern (5–65%), and mixed pattern of lysis and sclerosis (22–50%).12 A sunburst pattern with radiating spicules of bone is considered a characteristic feature of OS of the jaw. It occurs only in 7 to 27% of cases.12 The present case showed the classic sunburst pattern.

Histopathologically, on the basis of the amount of osteoid cartilage or collagen fibers produced by tumor, they are classified as osteoblastic, chondroblastic, and fibroblastic.12 The osteoblastic variety consists of tumor osteoid surrounded by bizarrely arranged fibroblast-like cells.16 In chondroblastic OS, tumor cells lie in the lacunae and form lobules. The center of the lobule has bony trabeculae producing a feathery appearance, and toward the periphery, the tumor becomes hypercellular. Most of the times, an area of atypical chondroid tissue is also seen with large chondrocytes. Fibroblastic OS is the least common variant where the tumor cells are spindle-shaped. According to Garrington's study on 56 cases of jaw OS, 60% were osteoblastic, 34% were fibroblastic, and less than 10% were of chondroblastic variant.10 The present case exhibits features of osteoblastic variant of OS.

Surgery and adjuvant chemotherapy radiotherapy may be required sometimes. The need of adjuvant therapy depends on the presence of micrometastases. In mandible, hemimandibulectomy is commonly preferred. A subtotal inferior maxillectomy for selected malignancies located on the alveolar ridge, palate and involving the antral floor have been described in literature.17 Overall, 5-year survival rate of 50% is reported for jaw OS.4 Patients with mandibular tumors generally fair better than those with maxillary tumors. Recurrence rate of OS of jaw is about 40 to 70% with a metastatic rate of 25 to 50%.7 Osteosarcomas are more likely to metastasize to lung and to brain than to regional lymph nodes.7 In the present case, the patient refused to undergo treatment and was lost to follow-up.

CONCLUSION

Osteosarcoma is a very aggressive neoplasm of OS origin with a potent risk of metastasis. Early diagnosis of this lesion may have a bearing on better prognosis and survival rate. Hence, a triple diagnostic approach, i.e., clinical, radiological, and histopathology, is essential for an accurate, and timely diagnosis.

REFERENCES