

# Osteosarcoma of maxilla mimicking neurofibroma

Mounesh Kumar C. D., Suresh K. V.<sup>1</sup>, Pramod R. C.<sup>2</sup>, Rajendra Baad<sup>2</sup>, Sreeja P. Kumar<sup>3</sup>, B. Karthik<sup>4</sup>

Departments of Oral and Maxillofacial Surgery, <sup>1</sup>Oral Medicine and Radiology, and <sup>2</sup>Oral Pathology and Microbiology, School of Dental Sciences, Krishna Institute of Medical Sciences, Deemed University, Satara, Maharashtra, <sup>3</sup>Department of Oral Medicine and Radiology, KMCT Dental College, Calicut, Kerala, <sup>4</sup>Department of Oral Pathology, Rishiraj Dental College, Bhopal, India

## ABSTRACT

Osteosarcoma (OS) is a primary malignant bone tumor producing osteoid or bone. It occurs less frequently in the jaws and in maxilla occurrence is rare. Jaws are less affected compared to long bones (4-13%). The mean age of occurrence is third to fourth decade. The histopathologic features of OS are highly variable. Herein, we report a rare case of a 72 year old female patient with a swelling in right posterior maxilla, whose clinical, radiographic findings were non-specific, An initial incisional biopsy confirmed neurofibroma. Later, a wide surgical excision was done as there was a spontaneous exacerbation of the growth, and histopathology report came as chondroblastic osteosarcoma.

**Key words:** Chondroblastic sarcoma, jaw, maxilla, osteosarcoma, sarcoma

## INTRODUCTION

Osteosarcomas (OSs) are malignant bone tumors characterized by formation of disorganized immature osteoid bone.<sup>[1-5]</sup> It accounts for 15-35% of all primary bone tumors, and is the second most common malignant bone tumor after multiple myeloma.<sup>[1,2]</sup> OS of jaw bones are uncommon, representing 4-8% of all osteosarcomas.<sup>[2,4-7]</sup> Bones with fastest rates of growth have the highest frequency of occurrence. It may occur inside the bones (intramedullary), or on the surfaces of bones, and in extraosseous sites. Rarely, extraskeletal osteosarcoma arise in soft tissues, commonly in thigh, upper extremity, and retroperitoneum, with no case reports of any in the oral cavity.<sup>[8]</sup>

Men develop OS more frequently than women.<sup>[4,5,9]</sup> Among osteosarcomas of jaws, 60% were men.<sup>[10]</sup> The

age-specific frequencies and incidence rates of conventional osteosarcoma is bimodal, whereas the peak incidence for jaw osteosarcomas were in the third and fourth decade.<sup>[1,5,9]</sup> OS of jaws show a predilection for mandible, however few studies found that it affects the mandible and maxilla almost equally.<sup>[4,5,9-11]</sup> Mandibular tumors arise more frequently in posterior horizontal ramus whereas maxillary lesions are commonly discovered in alveolar ridge, sinus floor and palate.<sup>[5,11,12]</sup>

The most common presenting features are swelling, pain, ulceration, and neurological disorders. The average time between presentation of symptoms and diagnosis range from 3 months to 5 months. This article presents an unusual case of a 72-year-old female patient which was initially misdiagnosed as a neurofibroma on histopathologic examination.

## CASE REPORT

A 72-year-old female patient reported to the department with the complaint of a growth on her right upper back tooth region since 1 week. Growth started 1 week after the extraction of tooth in the same area. It was associated with severe pain and difficulty in opening the mouth. She also reported excessive discharge in her right eye, paresthesia over right infra orbital region, and bleeding from right

### Access this article online

#### Quick Response Code:



#### Website:

www.ccij-online.org

#### DOI:

10.4103/2278-0513.106274

**Address for correspondence:** Dr. K. V. Suresh, Department of Oral Medicine and Radiology, School of Dental Sciences, Krishna Institute of Medical Sciences, Deemed University, Karad, Satara, Maharashtra, India. E-mail: dr.suri88@gmail.com

nose on forceful expiration and sneezing. Her systemic examination did not reveal any abnormal clinical findings.

Intraoral examination revealed a fixed hard, tender ulceroproliferative growth measuring  $4 \times 3$  cm with ill-defined margins involving the right half of the maxillary region. It was extending from distal part of 15 to distal part of maxillary tuberosity and from buccal side of residual ridge to 1 cm medial to midpalatine raphe. An unhealed extraction socket in 16 region was noted. No cervical lymphadenopathy was detected.

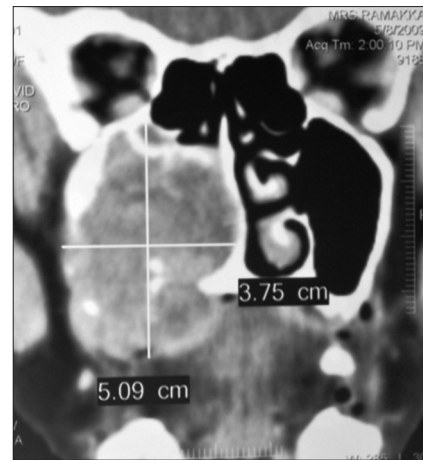
Computed tomography showed irregular mixed dense mass, causing destruction of the alveolar margins of the maxilla invading floor and anterior wall of the right maxillary sinus, inferior wall of the right orbit and lateral wall of nasal fossa [Figure 1]. Chest radiographs, blood tests, and abdominal ultrasonography were normal.

An incisional biopsy of the lesion revealed the diagnosis of Neurofibroma of the maxilla [Figure 2]. One week after the biopsy, patient noticed massive exophytic growth at the operated site. Surgical exploration was performed and the patient underwent a wide excision of the tumor with hemimaxillectomy of the right side. The histopathologic examination of the specimen revealed osteoid consisting of homogenous irregularly distributed material, chondroid areas showed abundant pleomorphism, atypical binucleated cells, and large hyperchromatic nuclei with prominent nucleoli [Figure 3], that was reported as chondroblastic osteosarcoma. The margins of the surgical resection were negative for the tumor. Patient is kept on periodic follow-up, and there was an improvement in her symptoms and no recurrence of the lesion was noted.

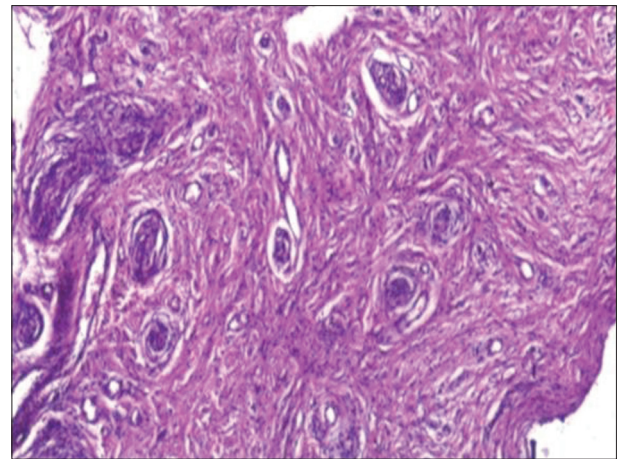
## DISCUSSION

OS is the most common primary malignant tumor of bone. Majority of primary bone malignancies arise *de novo*, but some apparently develop in association with recognizable precursors such as Paget disease, fibrous dysplasia, bone infarcts, chronic osteomyelitis, trauma, and exposure to radiation.<sup>[1,6,9]</sup> It may be associated with genetic predisposition such as the Li-Fraumeni or Beckman-Wiederman syndrome.<sup>[1,6,9,10]</sup> Our case appears to have developed *de novo* as no history of any predisposing factors could be elicited from this case. The peak incidence for jaw OS is third and fourth decade, which is about 10-15 years later than the mean age of long bone osteosarcomas.<sup>[4,5,9-11]</sup> Clinical findings are usually nonspecific, which includes swelling, pain, and general discomfort.<sup>[2,5,9,11,13]</sup> This was in accordance with the present case.

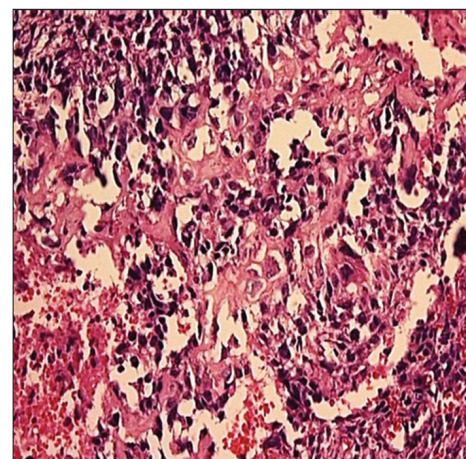
Radiographic findings vary from mixed sclerotic to radiolucent lesions. "Classic" sunray or sunburst appearance



**Figure 1:** Computed tomography showing irregular mixed density mass involving right maxillary sinus



**Figure 2:** Photomicrograph showing highly cellular areas, Schwann cells with fibroblasts arranged in the form of fascicles



**Figure 3:** Photomicrograph showing osteoid production with chondroid areas having typical binucleate appearance

due to osteophytic bone production on surface is noted in 50% of jaw osteosarcoma, best demonstrated in occlusal radiographs and computed tomography (CT) scan.<sup>[2,5,6,9,11]</sup>

Panoramic radiograph may show Garrington's sign—widening of the periodontal ligament space around affected teeth, with tapered resorption of tooth roots due to tumor infiltration.<sup>[2,5,6]</sup> The radiographic evaluation of present case revealed irregular ill-defined radiolucent areas without calcification. Although sun-ray appearance and codman triangle are less common in jaws (5, 18) combination of sun-ray appearance, widening of Periodontal ligament space and mandibular canal are pathognomonic for jaw OS. CT scanning and magnetic resonance imaging (MRI) can also be effective in tumor diagnosis and determination of its invasion to surrounding tissues.

The histopathology of osteosarcoma is highly variable, ranging from the more commonly seen osteoblastic and Chondroblastic type to the rare variants like myxomatous and telangiectatic types.<sup>[11,13,14]</sup> Chondroblastic osteosarcoma is the most common variant of OS in jaw bones, whereas osteoblastic osteosarcoma are common variant reported in the long bones.<sup>[1,5,6,11]</sup> Our case was mainly composed of lobular areas of malignant cartilage, with only few areas of tumor osteoid. Other areas of atypical fibroblast, myxoid areas, and bizarre giant cells were identified. Most authorities currently believe that even though the lesion is composed chiefly of malignant cartilage, it should be designated as osteosarcoma if malignant tumor osteoid can be identified, rather than chondrosarcoma.<sup>[14,15]</sup>

Treatment of this lesion is radical surgery consisting of complete resection with a margin of normal surrounding tissue which usually accompanies radiotherapy or chemotherapy. Anatomical limitations in face cause some difficulties in achievement of uninvolved margins and for this reason local recurrence of the lesion is high (between 33% and 69%).<sup>[15]</sup> The rate of metastasis in this lesion is less than long bone osteosarcomas. Present case had undergone complete excision of the tumor along with hemimaxillectomy.

Its prognosis is dependent not only on clinical and histologic parameters, but also on its anatomic site. Clark *et al.* reported that the patients with chondroblastic osteosarcomas of head and neck had a better overall survival rate than the patients with osteoblastic or fibroblastic tumors. The prognosis of jaw osteosarcomas is better than that of long bone osteosarcomas. This could be due to better histological differentiation of jaw osteosarcomas than long bone osteosarcomas. As jaw osteosarcomas occurs at higher mean age, the patients have less chance of developing metastases.<sup>[14]</sup>

## CONCLUSION

OS of the maxilla have an aggressive biological behavior and are difficult to diagnose clinically alone. Clinicians

and pathologists should be aware of its characteristics and main differential diagnoses to avoid late recognition. Early diagnosis and radical surgery with wide surgical margins are the keys to a good outcome. It has a better prognosis if diagnosed and treated at an early stage.

## REFERENCES

1. Raymond AK, Ayala AG, Knuutila S. Conventional osteosarcoma. In: Fletcher CD, Unni KK, Mertens F, editors. Pathology and Genetics of Tumors of Soft Tissue and Bone. 1<sup>st</sup> ed. IARC Press: Lyon 2002. p. 264-70.
2. Fernandes R, Nikitakis NG, Pazoki A, Ord RA. Osteogenic sarcoma of the jaw: A 10-year experience. J Oral Maxillofac Surg 2007;65:1286-91.
3. Amaral MB, Buchholz I, Freire-Maia B, Reher P, de Souza PE, Marigo Hde A, *et al.* Advanced osteosarcoma of the maxilla: A case report. Med Oral Patol Oral Cir Bucal 2008;13:E492-5.
4. Warnock G. Malignant neoplasms of the gnathic bones. In: Thompson LD, Goldblum JR, editors. Head & Neck Pathology. 1<sup>st</sup> ed. Elsevier: Amsterdam 2006. p. 492-7.
5. Angela C. Bone pathology. In: Neville BW, Damm DD, Allen CM, Bouquot JE, editors. Oral & Maxillofacial Pathology. 3<sup>rd</sup> ed. Saunders: St Louis 2009. p. 660-4.
6. Ogunlewe MO, Ajayi OF, Adeyemo WL, Ladeinde AL, James O. Osteogenic sarcoma of the jaw bones: A single institution experience over a 21-year period. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2006;101:76-81.
7. August M, Magennis P, Dewitt D. Osteogenic sarcoma of the jaws: Factors influencing prognosis. Int J Oral Maxillofac Surg 1997;26:198-204.
8. Bane BL, Evans HL, Ro JY, Carrasco CH, Grignon DJ, Benjamin RS, *et al.* Extraskelatal osteosarcoma. A clinicopathologic review of 26 cases. Cancer 1990;65:2762-70.
9. Rajendran R. Benign and malignant tumors of the oral cavity. In: Rajendran R, Sivapathasundaram B, editors. Shafer's Textbook of Oral Pathology. 6<sup>th</sup> ed. India: Elsevier; 2009. p. 169-73.
10. McGuff HS, Heim-Hall J, Holsinger FC, Jones AA, O'Dell DS, Hafemeister AC. Maxillary osteosarcoma associated with a dental implant: Report of a case and review of the literature regarding implant-related sarcomas. J Am Dent Assoc 2008;139:1052-9.
11. Zarbo RJ, Carlson ER. Malignancies of the jaws. In: Regezi JA, Sciubba JJ, Jordan RK, editors. Oral Pathology. 5<sup>th</sup> ed. Philadelphia: Saunders; 2008. p. 315-21.
12. Soares RC, Soares AF, Souza LB, Santos AL, Pinto LP. Osteosarcoma of mandible initially resembling lesion of dental periapex: A case report. Braz J Otorhinolaryngol 2005;71:242-5.
13. Junior AT, de Abreu Alves F, Pinto CA, Carvalho AL, Kowalski LP, Lopes MA. Clinicopathological and immunohistochemical analysis of twenty-five head and neck osteosarcomas. Oral Oncol 2003;39:521-30.
14. Devi Charan Shetty, Puneet Ahuja, Aadithya B. Urs, Rupinder Kaur. Histopathological variants of jaw osteosarcoma. Int J Pathol 2009;7:98-101.
15. Baghale K, Motahhary P. Osteosarcoma of the jaws: A retrospective study. Acta Med Iran 2003;4:113-21.

**Cite this article as:** Mounesh Kumar CD, Suresh KV, Pramod RC, Baad R, Kumar SP, Karthik B. Osteosarcoma of maxilla mimicking neurofibroma. Clin Cancer Investig J 2012;1:233-5.

**Source of Support Nil, Conflict of Interest:** No.