



Osteosarcoma of Mandible: Report of Two Cases with Review of Literature

¹P Wankhedkar Divyesh, ²N Chaphekar Akansha, ³R Patankar Sangeeta, ⁴S Tripathi Nidhi

ABSTRACT

Primary neoplasms of human skeleton are rare, accounting for 0.2% of overall human tumor burden. Osteosarcoma (OS) accounts for 15 to 35% of all primary bone tumors, in jaw bones, it is even, rarer representing 4 to 8% of all OS. Peak incidence for jaw OS is in 3rd to 4th decades while in long bones, it shows a bimodal age distribution. It may occur inside the bones (in the intramedullary or intracortical compartment), on the surfaces of bones and in extraosseous sites. Dental professionals may be first to detect jaw OS in their initial stages. Regardless of the favorable biologic behavior, the patients of jaw OS usually exhibit advanced tumor as it often goes unnoticed thus it is important for early diagnosing this lesion. Here, we report 2 cases of OS with different histological subtypes (chondroblastic and osteoblastic) and review of literature.

Keywords: Osteosarcoma, Chondroblastic, Osteoblastic.

How to cite this article: Divyesh PW, Akansha NC, Sangeeta RP, Nidhi ST. Osteosarcoma of Mandible: Report of Two Cases with Review of Literature. *Oral Maxillofac Pathol J* 2015;6(2):625-627.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

The term osteosarcoma (OS) refers to a heterogeneous group of primary malignant neoplasms affecting bone forming or mesenchymal tissues having a histopathological evidence of osteogenic differentiation.¹

It is a spindle cell neoplasm and accounts for almost 40 to 60% of all bone sarcomas. About 60% of all OS occur in the second decade of life in children and adolescents, and about 10% occur in the third decade.² Approximately, 5% of OS start in maxillary bones and the mandible is the most involved site.³ The World Health Organization (WHO) lists several variants that differ in location, clinical behavior and level of cellular atypia. The conventional or classical OS is the most frequent variant, which develops in the medullary region of the bone and can

be subdivided in osteoblastic and chondroblastic (CB) histological types, depending on the type of extracellular matrix produced by tumor cells.⁴

The classical OS is the most frequent variant, which develops in the medullary region of the bone and can be subdivided as osteoblastic, CB and fibroblastic types, depending upon the extracellular matrix produced by tumor cells. Here, we report two cases of chondroblastic OS while discussing its differential diagnosis.

CASE REPORTS

Case 1

A 50-year-old male patient reported to the outpatient department (OPD) with a swelling in the left posterior region of the lower jaw since the past 4 months.

Radiographic details showed a mixed radiopaque radiolucent lesion extending from the symphysis up to the left angle region anteroposteriorly and from alveolar crest to inferior border superoinferiorly.

Hematoxylin and eosin (H&E) section showed irregular chondroid tissue with atypical chondrocytes in a basophilic hyaline matrix. Eosinophilic areas suggestive of tumor osteoid were present lined by plump osteoblasts, in the surrounding fibrocellular connective tissue. Based on these findings, final diagnosis of CB variant of OS was given.

Case 2

A 15-year-old male patient reported to the OPD with an ulcer in the left posterior region of the lower jaw since the past 15 days.

Radiograph revealed a unilocular radiolucency with respect to 37 and extending upto the distal aspect of 38.

Hematoxylin and eosin section showed cellular stroma with disorderly arranged neoplastic cells. Areas of tumor osteoid surrounded by atypical osteoblasts could be appreciated. Dysplastic features, like cellular pleomorphism, hyperchromatism and abnormal mitosis, were present in the osteoblasts. A final diagnosis of osteoblastic variant of OS was given.

DISCUSSION

The term 'sarcoma' was introduced by the English surgeon John Abernathy in 1804 and was derived from Greek roots meaning 'fleshy excrescence'. In 1805, the French

^{1,2}MDS (3rd Year), ³Head and Guide, ⁴MDS (2nd Year)

^{1,4}Department of Oral and Maxillofacial Pathology, YMT Dental College and Hospital, Navi Mumbai, Maharashtra, India

Corresponding Author: P Wankhedkar Divyesh, 1st Floor Shri Niketan, 19th A Road, Anand Vihar Society, Khar West Mumbai-400052, Maharashtra, India, Phone: 9004664096 e-mail: divyeshpw@gmail.com

surgeon Alexis Boyer (personal surgeon to Napoleon) first used the term 'osteosarcoma'.⁵ The term 'osteosarcoma' as opposed to 'osteogenic sarcoma' is preferred by the World Health Organization (WHO).

Although the number of craniofacial OS is very low, the prevalence of jaw OS is in fact 10 times greater than that of OS in the total body skeleton, considering that jaws represent only 0.86% of total body volume.⁶ Jaw OS have been reviewed by many authors.

They commonly present in the third to fourth decades,⁷ which is about 10 to 15 years later than the mean age of long bone OS. Males slightly outnumber females. But the patient in first case was 50-year-old male and in second case the patient was 15-year-old boy. The mandible is involved more frequently than the maxilla with posterior body, horizontal ramus and the ascending ramus being the most commonly affected sites as seen in our cases. The average duration of symptoms before diagnosis is 3 to 4 months.⁸

Swelling, pain and general discomfort are the unusual nonspecific clinical findings.⁹ In first case, the patient reported with a swelling in the left posterior region of mandible since 4 months, while the second case the patient reported with an intraoral ulcerative growth since last 15 days. Osteosarcoma may arise *de novo* or several epidemiological risk factors have been related to the development of OS, including a history of ionizing radiation exposure, fibrous dysplasia, retinoblastoma or history of trauma.¹⁰ Risk factors were negative in our cases.

Aggressive ossifying fibroma and osteoblastoma (OB) are considered most likely clinical differential diagnosis which can resemble OS in clinical presentation and aggressive behavior. Recording an accurate clinical history is important in such cases, radiographic features serves as an adjunct in diagnosis.¹¹ The radiographic appearance of OS depends on the interaction of three factors: bone destruction, bone production with mineralization and periosteal new bone formation. Lesions can, thus, appear radiolucent, radiolucent with fluffy cloud-like radiopaque areas or entirely radiopaque. The most common presentation is a mixed radiolucency with radiopaque pattern and poorly defined irregular borders.¹² The tumor commonly perforates the cortical plates and extends into the soft tissue as seen in our case. A classical sunray/sunburst radiographic pattern has been described for OS. This occurs due to the periosteal reaction producing bone perpendicular to bone surface.¹³ This presentation is not pathognomonic as it can be seen with other neoplastic and reactive lesions too. Other radiographic findings include root resorption and a symmetrical widening of the periodontal ligament space around the teeth in the area of the lesion as seen in our cases.

Nakayama et al proposed a classification of CT pattern found in OS of jaws based on osteogenesis and signs of bone destruction. He proposed four types which were osteogenic with or without bone destruction and osteolytic with or without bone destruction. He reported better prognosis for osteogenic type without bone destruction.¹⁴

Histologically, the diagnosis of OS is made when sarcomatous stroma is seen to directly elaborate osteoid or primitive bone. The predominant histological variants described are osteoblastic, fibroblastic and CB types. The jaw lesions are predominantly CB.¹⁵

Although histopathological subtypes have not been shown to have prognostic significance, Broder's grading of the tumor based on the degree of cellular anaplasia including cellular pleomorphism, hyperchromatic nuclei, bizarre mitosis, multinucleated giant cells is prognostically significant. High grade OS of jaws has been reported to be always associated with poorer prognosis.¹⁶

Chondroblastic variant of OS can be confused with chondrosarcoma (CS) in small biopsy specimens and differential diagnosis can be difficult. Osteoid formation within the tumor is the most important histomorphologic diagnostic feature. It is not always possible to visualize osteoid in the small biopsy specimens. As a consequence CB variant of OS may be misdiagnosed as CS. False diagnosis rate has been reported to be as high as 44%.¹⁷

Chondrosarcoma of the jaws is extremely uncommon. It occurs primarily in the anterior maxilla. Although CS occurs in patients of all ages, most of those affected are over 50 years of age.

Immunohistochemistry (IHC) plays an important role in the differentiation between CS and CB variant of OS. Immunohistochemistry will show CS to be positive for S100 and vimentin and negative for cytokeratin and epithelial membrane antigen (EMA). Chondroblastic variant of OS will be positive for vimentin, EMA, S100 and rarely cytokeratin.^{18,19}

The diagnosis of chondroblastic OS may not be possible on the basis of clinical examination and roentgenographic investigations. Biopsy for histological diagnosis is essential before definitive therapy is undertaken. A recently found gene encoding an intranuclear osteocalcin promoter—'Cbfa1' appears to be a potential marker in the definitive diagnosis of malignant bone tumors. In addition, the detection of alkaline phosphatase activity in imprint preparations obtained from the cut surface of OS before fixation, is regarded as diagnostic of OS, when used in combination with radiographs.²⁰ Chondrosarcoma is negative for this enzyme.

According to Garrington's series of 45 cases, the overall 5-year survival rate for maxillary OS was 25% and, for mandibular osteosarcoma, 41%. There was no



correlation between histological characteristics of the tumor and prognosis.²⁰ However, according to Bennett et al, chondroblastic variant has better prognosis than others. In case of CS, the overall 5-year survival rate is approximately 90% for grade I lesions, 81% for grade II lesions and 43% for grade III lesions.²⁰ Chondroblastic OS is sensitive to chemotherapy and radiotherapy while CS is resistant to both. In the reported cases, wide surgical excision was carried out with removal of the adjacent healthy tissue around the tumor. Follow-up for 6 months showed no recurrence in both cases.

CONCLUSION

Osteosarcoma presents a wide spectrum of clinical, histological and radiological features. Therefore, all these features need to be correlated to reach a conclusive diagnosis. Initial diagnosis of OS is difficult to arrive clinically and radiographically. Diagnosis can be missed, if adequate and proper site is not chosen for incisional biopsy. It has a better prognosis if diagnosed and treated at an early stage.

REFERENCES

1. Yesilova E, Akgunlu F, Dolanmaz D, Yasar F, Sener S. Osteosarcoma: a case report. *Eur J Dent* 2007 Jan;1(1):60-63.
2. Sheikh S, Pallagatti S, Aggarwal A, Gupta D, Puri N, Mittal A. Osteosarcoma of maxilla. *J Clin Exp Dent* 2010;2(3):117-120.
3. Rosilene C, et al. Osteosarcoma of mandible initially resembling lesion of dental periapex: a case report. *Braz J Otorhinolaryngol* 2005 Mar-Apr;71(2) (Part 1):242-245.
4. Bennett JH, Thomas G, Evans AW, Speight PM. Osteosarcoma of the jaws: a 30-year retrospective review. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2000 Sep;90(3):323-332.
5. Tamgadge A, Tamgadge S, Gotmare S, Bhalerao S. Osteosarcoma—an osteolytic lesion in the posterior mandible: a case report with review of literature. *J Dent Med Sci* 2014;13(4): 8-13.
6. van den Berg H, Schreuder WH, et al. Clinical study osteosarcoma: a comparison of jaw versus non jaw localizations

and review of the literature. Hindawi Publishing Corporation Sarcoma volume 2013, Article ID 316123. p. 9.

7. George A, Mani V, Sunil S, Sreenivasan BS, Gopakumar D. Osteosarcoma of maxilla: a case of missed initial diagnosis. *Oral and Maxillofac Pathol J* 2010 Jan-Jun;1(1):33-38.
8. Garrington GE, Scofield HH, Cornyn J, Hooker SP. Osteosarcoma of the jaws: analysis of 56 cases. *Cancer* 1967 Mar;20(3):377-391.
9. Fernandes R, Nikitakis NG, Pazoki A, Ord RA. Osteogenic sarcoma of the jaw: a 10-year experience. *J Oral and Maxillofac Surg* 2007;65(7):1286-1291.
10. Augustine D, Ramesh M, Murali S, Sekar B, Paul G. Osteosarcoma of mandible: a rare report with literature review. *World J Dent* 2014;5(3):190-192.
11. Petrikowski CG, Pharoah MJ, Lee L, Grace MG. Radiographic differentiation of osteogenic sarcoma, osteomyelitis, and fibrous dysplasia of the jaws. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1995;80(6):744-750.
12. Lindqvist C, et al. Osteosarcoma of the mandible: analysis of nine cases. *J Oral and Maxillofac Surg* 1986 Oct;44 (10):759-764.
13. Bojan A, Christy W, Chanmougananda S, Ashokan K. Osteosarcoma of mandible: a case report and review of literature. *J Clin Diag Res* 2012 May (Suppl-2);6(4):753-757.
14. Nakayama E, et al. The association between the computed tomography findings, histologic features, and outcome of osteosarcoma of the jaw. *J Oral and Maxillofac Surg* 2005;63(3): 311-318.
15. Kumaravelu C, et al. Chondroblastic osteosarcoma of maxilla: a case report and review of literature. *J Maxillofac Oral Surg* 2009 Sep;8(3):290-293.
16. Okada K, Unni KK, Swee RG, Sim FH. High grade surface osteosarcoma: a clinicopathologic study of 46 cases. *Cancer* 1999 Mar 1;85(5):1044-1054.
17. Akpolat N, et al. Sacral chondroblastic osteosarcoma misdiagnosed as chondrosarcoma and chordoma. *Turk J Med Sci* 2007;37(4):243-249.
18. Hasegawa T, et al. Immunophenotypic heterogeneity in osteosarcomas. *Human Pathology* 1991 Jun;22(6):583-590.
19. Yamamura H, Yoshikawa H, Tatsuta M, Akedo H, Takahashi K. Expression of the smooth muscle calponin gene in human osteosarcoma and its possible association with prognosis. *Int J Cancer* 1998 Jun 19;79(3):245-250.
20. Vasudeo KS. Chondroblastic osteosarcoma of jaw bone. *J Case Reports* 2013 Dec;3(2):480-484.