Fibroblastic Osteosarcoma in Mandible: A Rare Case Report and Review of Literature

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Osteosarcoma is a malignancy of mesenchymal cells able to produce osteoid or immature bone. It is uncommon and represents 6-8% of all osteosarcomas. It occurs most often in the third and fourth decades of life, i.e. is 10-15 years older than the mean age for osteosarcomas of the long bones.

A fibroblastic osteosarcoma in the symphyseal region of mandible of a 58 years old man is reported. The specific characteristic of this case is its histological features and its site, because fibroblastic osteosarcoma is rare in jaws and most cases are chondroblastic osteosarcoma.

Key Words: osteosarcoma, malignant bone tumor, fibroblastic type.



Fig. 1. Clinical picture showing the lesion.

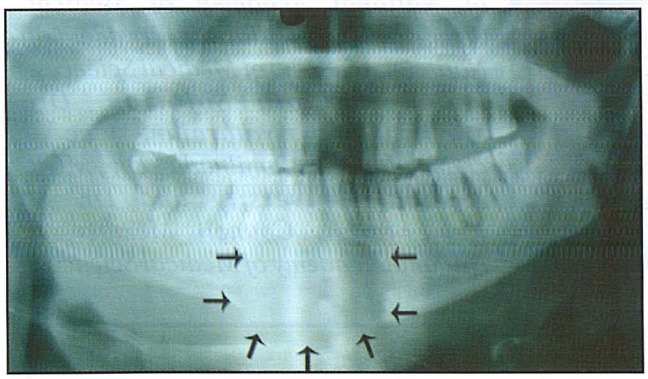


Fig. 2. Radiograph showing large irregular radioluscent lesion in the symphsis region showing destruction of inferior cortical margin.

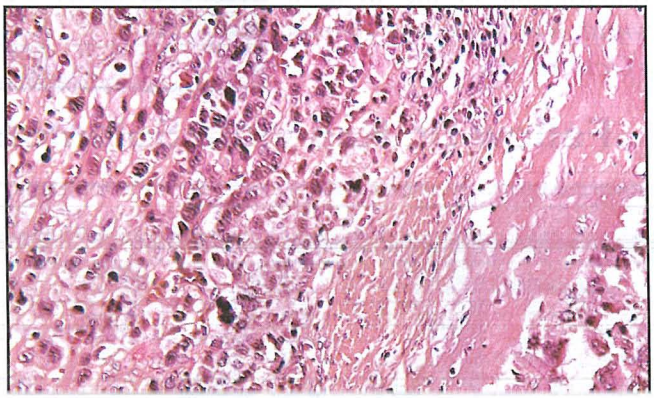


Fig. 3. Histologic slide showing mesenchymal cells with spindle to ovoid shaped nuclei with variable degree of atypical features and minimum amount of bone production.

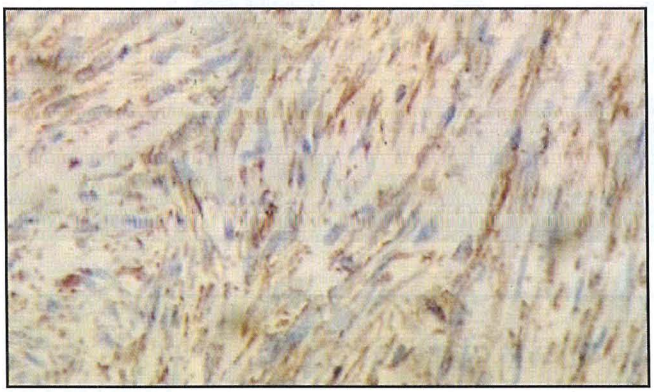


Fig. 4. Immunohistochemical staining showing vimentin positivity in spindle cells.

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Osteosarcoma is a primary malignant tumor of the skeleton characterized by the direct formation of immature bone or osteoid tissue by the tumor cells.[1] 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 surgeon Alexis Boyer first used the term "Osteosarcoma. It is the most common non-hemopoeitic primary tumor of bones. Intraoral sarcoma is a very rare disease and may constitute approximately 1% of all head and neck cancers and only 0.14% of intraoral malignancies.[2] The gender distribution of jaw osteosarcoma is a controversial issue. Men seem to be more commonly affected. In a review of jaw osteosarcoma in the medical literature by Mardinger et al there was a male predilection with a male : female ratio of 1.2:1.0. This has been attributed to the longer period of skeletal growth and additional volume of bone in men, though neither has been confirmed.[3] In the jaws, the biologic behavior of osteosarcoma differs from tumors occurring elsewhere; the average age of onset is 10-20 years later than for skeletal lesions, the histopathologic variables are more favorable, distant metastasis occur less frequently; and survival rates are higher.[4] The most common places of occurrence of this lesion are alveolar ridge and body in maxilla and mandible.[5] The most common symptoms of osteosarcoma include persistent pain, swelling and paresthesia/anesthesia. General manifestations of disease, such as fever or considerable weight loss are rare. Notably, a significant proportion of patients with osteosarcoma may present without any pain except jaw swelling. The age range of cases of osteosarcoma appears variable in diverse studies worldwide. Among the 30 cases presented by August et al. the age range was 4-84years. Notably, Chindia et al presented a 1 week old female patient in their series of 14 cases of osteosarcoma. Thus disease may manifest at any age.[6]

In the histopathologic findings, depending on the relative amounts of osteoid, cartilage or collagen fibers produced by the tumor, osteosarcoma is classified into the: osteoblastic, chondroblastic, fibroblastic and small round cell types. The tumor cell may vary from relatively uniform round or spindle shaped cells to highly polymorphic cells with bizarre nuclear and cytoplasm shapes.[6] The essential microscopic criterion is the direct production of osteoid by malignant mesenchymal cells in a sarcomata stoma.[6]

In addition to osteoid, the cells of the tumor may produce chondroid materials and fibrous connective tissue.[6]

The diagnostic evaluation of osteosarcoma should delineate the extent of the primary tumor and the presence or absence of metastatic disease. Thus, in addition to a definitive incisional biopsy, basic diagnostic evaluation should include: a clear history, physical examination, plain radiographs of the affected jaw including those of chest: and computerized tomography or magnetic resonance imaging of the affected area. Radiologically, the findings may be the same as those of osteosarcoma in other sites: osteolytic, osteoblastic or a mixture of the two with poorly defined irregular margins that are characteristic of malignant lesion. It is important to note that some cases may present with no evident radiological changes, while others may exhibit variable appearances including sclerotic, laminated and the classic sunburst patterns.[4]

The management of osteosarcoma can include preoperative chemotherapy; complete surgical excision of the tumor; postoperative chemotherapy, radiation, and combination of treatment modalities.[7]

We present here a unique case of jaw osteosarcoma in symphysial region in 60 yr old male with histopathology revealing fibroblastic variant which is again a rare entity. The aim of this article is to describe a case of osteosarcoma at a rare site with rare histopathologic representation.

Case Report

A 58-year-old man came with complain of a gradually enlarging painful mass in his lower right side of jaw of 2 months duration. Pain was gradual in onset, intermittent and throbbing in nature. Facial asymmetry due to presence of extraoral swelling in the

symphysis region of size 6x4 cm approximately was extending superoinferiorly from lower lip to 5 cm below the lower border of mandible and mediolaterally from corner of the mouth extending towards the right side of face approximately 2 cm short of angle of mandible. Intraorally, exophytic growth was present in the symphyseal region extending anteroposteriorly from lower left lateral incisor to distal of lower right first molar with obliteration of labial vestibule. Swelling was soft to firm in consistency and tender on palpation (Fig 1). There was no lymphadenopathy noted. Habit of betel nut chewing 10-12 times a day since 30 years was present. History of tingling and numbness on the lower right side of jaw was also present. Orthopentomogram(OPG) shows large irregular radioluscent lesion present at mandibular symphysis region extending mediolaterally from lower left canine to lower right first molar and superoinferiorly from apical area of anterior teeth to inferior cortical margin destroyed by pathology. No sclerotic margin was seen adjacent to radiolucent area. Destruction of inferior cortical margin is irregular. The OPG also shows generalized bone loss with periapical lesion involving lower right first, second and third molar.(Fig 2)

Incisional biopsy was taken under local anesthesia, fixed in formalin and was sent for histopathological examination. The diagnosis was given as osteosarcoma. Complete excision of the lesion was done with was done with segemental mandibulectomy. One margin was close on histopathological examination so the patient was subjected for intense radiotherapy. The excisional specimen was fixed in formalin, paraffin embedded and stained by hematoxylin and eosin. Microscopically the lesional tissue revealed proliferating fibroblasts cells with various spindle to few ovoid shaped nuclei and indistinct cytoplasmic membranes along with bundles of collagen fibers. The cells exhibit a variable degree of atypical features, including hyperchromatism, pleomorphism, abnormal mitoses, and increased nuclear/cytoplasmic ratio. The tumor cells exhibited only minimal bone matrix production. There was a presence of abundant inflammatory cells along with few reactionary giant cells, interspersed with a myxoid background (Fig. 3). Immunohistochemical staining was performed with vimentin markers and S-100 protein. Vimentin positivity [Fig: 4] was seen in spindle cell population indicating myofibroblastic origin and S-100 was negative, thus ruling out neural tumors. Based on the clinical picture, rate of growth of the lesion and histopathological findings, a final diagnosis of fibroblastic osteosarcoma was given.

The patient was followed for 9 months and had a normal postoperative course of healing with no recurrence and distant metastasis.

Discussion

Osteosarcoma is the most common non haemopoietic primary malignant tumor of bone. Majority of primary bone malignancies arise de novo, but some apparently develop in association with recognizable precursors (precancerous conditions) such as Paget's disease, fibrous dysplasia, bone infarcts, chronic osteomyelitis, trauma, and exposure to radiation. It also has been associated with metallic implants, joint prostheses, and in some genetic syndromes – Li-Fraumeni syndrome, hereditary retinoblastoma and Rothmund-Thomson syndrome.[8] The present case appears to have developed de novo.

Osteosarcoma is a common primary tumor of jaws. The prevalence of jaw osteosarcoma is 10 times greater than that of osteosarcoma in the total body skeleton, considering that jaws represent only 0.86% of the total body volume and 6.5% of all osteosarcomas. This highlights the importance of understanding the nature of jaw osteosarcoma.[1]

Males are affected more frequently in most series (male: female ratio; 1.4:1), though the rate for girls up to about age 13 years are roughly 30% higher than those for boys.[2] In mandible osteosarcomas, the body of the mandible is usually the primary site, followed by the symphysis and ascending ramus. The alveolar ridge and antrum are the most common sites in the maxilla.[9] The usual presentation is of a rapidly growing swelling with loosening and displacement of teeth which may cause dysaesthesias.[9]

Radiography is the initial imaging modality in the evaluation of bone tumors.[10] The diagnosis of osteosarcoma is typically suspected by the radiographic appearance of the affected bone. Ossification in the soft tissue component of the bone, manifesting as "sunburst" pattern is classic for osteosarcoma but is not a specific feature. Periosteal new bone formation with lifting of the cortex leads to the appearance of a Codman's triangle. Garrington et al. (1967) mentioned that roentgenographic evidence of a symmetrically widened periodontal membrane space is a significant early finding in Osteosarcoma of jaw, although the same features have been seen in some chondrosarcomas[2] In an analysis of osteosarcoma of the mandible, Lindquist et al reported that the widening of periodontal ligament space and inferior dental canal, together with sunburst effect are almost pathognomonic of jaw osteosarcoma. This highlights the importance of histopathologic analysis in the diagnosis of osteosarcomas. [11]

Histopathologically, osteosarcoma of long bones and jaw bones share common features. The diagnosis of osteosarcomas is based on the recognition of osteoid production by tumor cells .[12]

In present case lesion was a non ulcerated swelling covered by an inflamed oral mucosa, and clinically resembling a benign intrabony tumor. On subsequent visits it drastically grew to an enormous size causing facial asymmetry extraorally and also obliteration of labial vestibule intraorally along with tingling and numbness on the lower right side of jaw. Microscopically the lesional tissue revealed characteristic features of fibroblastic variant of osteosarcoma. Radiographically the lesion showed large irregular radiolucent lesion present in mandibular symphysis region extending mediolaterally from lower left canine to lower right first molar and superoinferiorly from apical area of anterior teeth to inferior cortical margin destroyed by pathology with no sclerotic margin adjacent to radiolucent area. Destruction of inferior cortical margin is irregular. The OPG also shows generalized bone loss with periapical lesion involving lower right first, second and third molar. [Fig 2]

Recent data suggest that in patients with osteosarcoma of head and neck, treatment with surgery and radiation therapy improves the prognosis of a subset of patients whose surgical specimens exhibited tumor at the resection margins.[7]

There is no absolute consensus as to the classification of osteosarcoma with respect to projected outcomes and treatment recommendations. However, there is a small group of early stage tumors that are cured by surgery alone. The majority of tumors though, presents in more advanced stages require combination treatment.[7] In the present case complete surgical excision of lesion was done with segmental mandibulectomy which was followed by intense radiotherapy.

The mandibular lesions have better prognosis than the maxilla with the survival rate of 25-40%. Osteosarcomas of the jaws are locally aggressive and tend to recur than to metastatise.[7] Local uncontrolled disease is more often the cause of death for patients with jaw osteosarcoma than are the effects of distant metastasis. Although regional lymph nodes may be infrequently involved, most often metastasis affects the lung and brain. [13]

However the following factors improve 5 years survival rate: [14]

1. Early detection of tumor and performing surgical excision
2. Early age of onset
3. Radical surgical excision
4. Low histological grade
5. Complete surgical removal without recurrence.

In conclusion, in fibroblastic osteosarcoma, the key to a good outcome is complete and exact attention to the radiographic changes and early diagnosis.

References

1) Nissanka EH, Amaratunge EAPD, Tilakaratne WM. Clinicopathological analysis of osteosarcoma of jaw bones. Oral Dis 2007; 13:82-87.

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mal adjustment as compared to guidance ramp of acrylic resin which can be periodically adjusted.

Conclusion

Prosthodontic modifications can be accomplished to compensate for defects that are not correctable with surgical reconstruction. Hemimandibulectomy patients suffer numerous problems with mastication, speech, and deglutition due to anatomical compromise resulting from tumor resection. Rehabilitation of rudimentary function of speech and swallowing should be of paramount importance for all Prosthodontist. Prosthodontist should strive to maintain existing mandibular dentition as they will help to retain prosthesis and support the lips and stabilize the mandible for swallowing. Anterior dentition is important to assist in air flow during speech, especially when tongue movements are compromised. While prosthesis can replace dentition to aid in mastication, the effect on mastication is a secondary to prosthetic benefit compared with rehabilitation of the rudimentary function of speech and swallowing that the prosthesis offer.

Success of prosthesis for resected edentulous mandible is directly related to the amount of remaining bone, the number and integrity of remaining teeth and continuity and discontinuity of the bone defect.

References

1. Master S.B. Prosthodontist and cancer patient – A monograph. Journal of Pierre Fauchard Academy 1987 Nov;4:1:111-141
2. John Beumer, Thomas A Curtis, Mark T Marunic and Eleni Roumanas. Acquired Defects of the Mandible: Etiology, Treatment, and Rehabilitation. In. Maxillofacial Rehabilitation Prosthodontic and surgical considerations. Editor, Brian W. Cochran. Ishiyaku EuroAmerica-Inc St. Louis. Tokyo 1996:113-224
3. Cantor & Curtis. Prosthetic management of edentulous mandibulectomy patients. Part-1. Anatomic, physiologic, and psychologic considerations. J Prosthet Dent 1971 Apr ;25(5): 446-457
4. Thomas D Taylor. Clinical Maxillofacial prosthetics. Quintessence Publishing Co. Inc. Carol Stream Illinois 2000.
5. Alan B Carr, Glen p McGivney , David T Brown. Removable partial denture considerations in maxillofacial prosthetics. In. McCracken's Removable partial Prosthodontics eleventh edition. Mosby Elsevier St. Louis Missouri 2005:397-420

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2) Khorate MM, Goel S, Singh MP, Ahmed J. Osteosarcoma of Mandible: A Case Report and Review of Literature. J of Cancer Sci and Ther 2010;2(4):122-125.

3) Mardinger O, Givol N, Talmi YP. et al . Ostcosarcoma of the jaw – The Chaim Sheba Medical Center Experience. Oral Surg Oral Med Oral Path Oral Radiol Endod. 2001; 91: 445–451.

4) Chindia L. Mark. Osteosarcoma of Jaw bones. Oral Onco 2001; 37: 545-547.

5) Baghaie F. Motahhary P. Osteosarcoma of the Jaws: A retrospective study. http://journals.tums.ac.ir/on Wednesday, June 27, 2012.

6) Neville B, Damm D, Allen C.M, Bouquot J.E. Oral & Maxillofacial Pathology. 2nd ed. W.B Saunders an Imprint of Elsevier Science Philadelphia London New York St. Louis Sydney Toronto; 2002.

7) Padilla J R, Murrah A M. The spectrum of Gnathic Osteosarcoma: Caveats for the Clinician and the Pathologist. Head and Neck Pathol 2011; 5: 92-99.

8) George Antony, Varghese Mani, Sunil S, Sreenivasan BS, Devigopakumar. Oral and Maxillofac Pathol J. 2010; 1(1):976-1225.

9) Cawson RA, Binnie WH, Barrett AW, Wright JM. Oral Disease. 3rd ed. St. Louis: C.R. Mosby co.; 2001.

10) Massengill AD, Seeger LL, Eckardt JJ. The role of plain radiography, computed tomography and magnetic resonance imaging in sarcoma evaluation. Hematolo Oncol Clin North Am . 1995; 9:571-604.

11) Lindquist C, Teppo L, Sane J, Holmstrom T, Wolf J Osteosarcoma of the mandible: analysis of 9 cases. J Oral Maxillofac Surg. 1986; 44: 759–764.

12) Schajowicz F (1993). Histological typing of tumours of bone; WHO international histological typing of tumours. Springer-Verlag: Berlin.

13) August M, Magennis P, Dewitt D: Osteogenic sarcoma of the jaws: factors influencing prognosis. Int J Oral Maxillofac Surg. 1997; 26: 198-204.

14) Kassir RR, Rassekh CH, Kinsella JB, Segas J, Carrau RL, Hokanson JA. Osteosarcoma of the head and neck: Meta analysis of non-randomized studies. Laryngoscope 1997; 107:56-61.