

Osteosarcoma of maxilla: A case report

Dr M.Vijaya Sree M.D¹, Dr I.V.Renuka. M.D,² Dr.G. Saila bala. M.D,³

Dr K. Nageswara rao M.D,⁴

¹ Associate Professor of pathology, Guntur Medical college, Guntur, Andhra Pradesh, India.

² Professor of Pathology, Guntur Medical college, Guntur, Andhra Pradesh, India

³ Professor of Pathology, Guntur Medical college, Guntur, Andhra Pradesh, India

⁴ Assistant Professor of pathology, Guntur Medical college, Guntur, Andhra Pradesh, India

Abstract: Craniofacial Osteosarcoma is a relatively rare entity comprising about 6-10% of all sarcomas. We present a rare case of maxillary OS early diagnosed in a 17 year old girl through clinical, radiological and histological examination. The patient was much younger than the average age of presentation of a jaw tumor, which typically presents in the third to fourth decade of life.

I. Introduction

Osteosarcoma is one of the most common primary malignant neoplasms, which predominantly occur in the long bones and rarely in the maxilla, facial area (6-10% of all sarcomas)⁽¹⁾ Within the craniofacial region the mandible is usually reported as the most common site of the involvement, followed by the maxilla and skull.^(1,2) Jaw lesions are diagnosed on average two decades later than sarcomas of long bones, with a peak incidence between 20 and 40 years⁽³⁾. Head and neck OS (HNOS) are associated with a lower metastatic rate than long bone OS, and they have a better 5-year survival, ranging between 27% and 84%. Approximately 80% of HNOS originate from soft tissue, while 20% arise from bone. Although a male predilection has been reported, some authors have observed a slight predominance in females⁽⁴⁾. The chief clinical features are swelling, pain and ulceration⁽⁵⁾. Radiologically, the findings may include radiolucency, radiopacity, or a mixture of both with poorly defined irregular margins.⁽⁵⁾ HNOS differ from Osteosarcoma of the long bones in its biological behaviour, presenting a lower incidence of metastasis and a better prognosis. Early diagnosis and adequate surgical resection are the keys to high survival rates⁽⁶⁾.

II. Case report

A 17-year-old girl presented with swelling over the left side of the cheek since three weeks. It started intraorally and gradually increased in size and was exhibited extraorally. There was no discharge from the swelling. It was associated with difficulty in swallowing solids. There was no history of other medical problems and she was otherwise in a state of good health. The hematological and biochemical parameters were within normal limits. All the vital signs like blood pressure, pulse, respiratory rate and temperature were within normal limits. On inspection a diffuse swelling was noted on the left side of the cheek over the body of the maxilla measuring 4cm x 3cm x 2cm in dimensions. The skin over the swelling was normal in appearance. The surface over the swelling showed no secondary changes like scar formation, sinus or fistula with no discharge of any kind. There was no localized rise in temperature. The swelling was firm, immovable and was fixed to the underlying bone. On internal examination a solitary localized swelling over the upper left molar tooth extending into the maxilla and retromolar area. The surface of the swelling was red in color. On palpation the swelling was firm in consistency, non-tender and fixed to the underlying bone. All these features gave a clinical impression of a neoplastic swelling. To confirm the nature of the swelling various investigations were carried out. The patient was first subjected to radiographic investigations. A CT scan of the paranasal sinus region showed a 3cm x 3cm lesion in the body of the maxilla with bony erosion in the lateral wall. An incisional biopsy was carried out under anesthesia and sent for histopathological examination. We received multiple grey-white to grey-brown bits of total size 2.5cm x 2cm x 1cm. The histological examination revealed areas of osteoid surrounded by sheets of neoplastic cells. Microscopic diagnosis was osteosarcoma. Immunohistochemistry for pancytokeratin was also done to differentiate it from a malignant odontogenic tumor. The pancytokeratin was negative in neoplastic cells. Thus the final diagnosis of osteosarcoma was confirmed. The patient was referred to the oncology hospital for further treatment.



Figure 1: Intraoral view of the tumor



Figure 2: C T scan showing extensive maxillary tumor

III. Discussion

Os involves the head and neck region and approximately 10% of all sarcomas. The mandible and maxilla are the most frequently affected sites, followed by the paranasal sinuses and skull⁽⁶⁾. The HNOS occur in a population older than the one in which sarcoma of the long bones occur, with a peak incidence from 20 – 40 years. Males are affected slightly more often than females. Approximately 80% of HNOS originate from soft tissue, while 20% arise from bone.⁽⁷⁾ Soft tissue sarcomas arise from the mesenchyme, including muscle, endothelial cells, cartilage and supporting elements.⁽⁸⁾

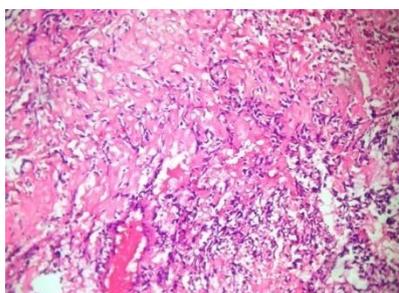


Figure 3: Hematoxylin and eosin stains Magnification 100x. Note areas of osteoid formation pleomorphism and hyperchromasia

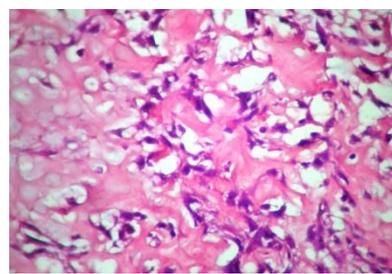


Figure 4: Hematoxylin and eosin stains Magnification 400x. Note moderate nuclear pleomorphism and hyperchromasia

Most head and neck sarcomas appear at presentation as localized disease. Regional metastasis occur in 10-15% of overall HNOS; most of these arise from high grade primary lesions. At presentation, distant metastasis are rare in absence of regional metastasis; the presence of nodal metastasis showed a prompt search for distant metastasis. Most common site of distant metastasis is the lung, followed by liver and bone. The diagnosis of osteosarcoma is usually dependent on clinical, radiologic and histopathological features⁽⁹⁾. The affected patients usually present with pain and swelling of the concerned area, along with occasional ulceration.⁽¹⁰⁾ Other clinical features include mobility of tooth, numbness with the lips {or} other parasthesia.⁽¹¹⁾ Radiological evaluation is necessary for diagnosis because clinical symptoms such as pain, parasthesia, swelling etc, are non specific⁽¹²⁾. Radiographically may present with an expansion of bones, it may present as radio opacity, radio lucency {or} of mixed nature. There is a need to be more conscious while diagnosing such patients as it often goes undiagnosed for a significant period of time. This is so because some of the signs and symptoms can be produced by a number of different developmental, infections, benign neoplastic diseases {or} malignancies. Further its presentation is similar to that of osteomyelitis, ossifying fibroma and fibrous dysplasia which has often causes an osteosarcoma to be delayed in its diagnosis. A tissue biopsy is the only means of making a definite diagnosis and such cases presenting clinically of osteosarcoma requires a biopsy as soon as possible.⁽¹³⁾ The biopsy specimen from superfaecial {or} peripheral aspects of the tumour from the advancing edge is least likely to be representative of the tumour and frequently fails to demonstrate osteoid formation. The deeper biopsy should be taken from the centre of the lesion. This will avoid missing the diagnostic portion of the tumour because superfaecial part of the specimen include the immature part of the lesion. Early and small lung metastatic deposits are of great concern in osteosarcoma. So its complete screening requires a chest radiography {or} a chest CT scan.⁽¹³⁾ The histopathological evaluation of the biopsy specimen showed a cellular stroma with evidence of osteoid and chondroid formation. The former is a diagnostic feature for the diagnosis of osteosarcoma. The tumor classification as osteoblastic (or) chondroblastic was not performed, since the specimen was collected by an incisional biopsy. More ever no association between histological classification JOS and

survival has been reported. Regardless of favourable biologic behaviour in relation to other skeletal bone osteosarcomas patients with JOS may exhibit advanced tumors mainly early diagnosis is not performed.

IV. Conclusion

It is concluded that misdiagnosis is very common in osteosarcoma of the jaw although JOS presents a better diagnosis than osteosarcoma of the long bones, patients with JOS can exhibit advanced tumors mainly early diagnosis is not performed. Clinicians and pathologists should be aware of its characteristics and main differential diagnosis to avoid late recognition. IHC in our case proved to be useful in confirmation of diagnosis. Although the histological picture of OS long bones and jaw bones exhibit similarity the management modalities may not necessarily be the same. Since local regional recurrence is the main cause of mortality in OSJ, meticulous wide resection seems to be the mainstay of treatment. Surgery with wide margins is an important factor in successful therapy for patient with JOS. Therefore early diagnosis and radical surgery with wide surgical margin are the key to a good outcome. In our view the presented case is interesting in terms of relatively early age of onset of tumour and diagnosis. CT has an important role in early diagnosis and the evaluation of its extent for the surgical planning.

References

- [1]. Barnes EL, Peel RL, Verbin RS, Goodman MA, Apple BN, Diseases of the bones and joints. In: Barnes EL, editor, surgical pathology of the head and neck. NEW YORK: Marcel Dekker Inc; 1985. P.985-99.
- [2]. Mardinger O, Givol N, Talmi Yp, Taicher S, Saba K, Hashomer T. Osteosarcoma of jaw oral surg oral med oral pathol oral radiol Endod 2001;91:445-51.
- [3]. Tommaso cuttilli, secondo scarsella, Desiderio Di Fabio, Antonio oliva, pasqualino Cargini. Annals of maxillofacial surgery. July-December 2011; volume1: Issue-2:176-80.
- [4]. Bennett JH, Thomas G, Evans AW; Speight PM. Osteosarcoma of the jaw; a 30 year Retrospective review. Oral surg oral med oral pathol oral radiol endod. 2000 sep;90(3):323-32.
- [5]. August M, Magennis P, Decoitt D. Osteosarcoma of the jaws: Factors influencing prognosis. Int J oral maxillofac surg. 1997 june;26(3):198-204.
- [6]. Kassir RR, Rassekh CH, Kinsella JB, Segas J, Carrau RL, Hookanson JA. Osteosarcoma of the head and neck: meta-analysis of non randomized studies. Laryngoscope 1997;107:56-61.
- [7]. Yamaguchi S, Nagasawa H, Suzuki T, Fujii E, Iwaki H, Jakagi M, et al. Sarcomas of the oral and maxillofacial region; A review of 32 cases in 25 yrs. Clin oral investi of 2004;8:52-5.
- [8]. Seidl RO, Gerein V, Vogel HJ, Ernst A. mesenchymal chondrosarcoma of the facial skull. HNO 2001;49:744-9.
- [9]. Bianchi SD, Boccardi A. Radiological aspects of osteosarcoma of the jaws. Dento maxillofac Radiol 1999;28:42-7.
- [10]. Amaral MB, Buchholz-J, Freire - maia B, Reher P, desouza PE, Marigo Hde A, et al. Advanced osteosarcoma of the maxilla; a case report. med oral pathol, oral cir Buccal; 2008;3E492-5.
- [11]. Yesilova E, Akgunlu F, Dolanmaz D, Yasar F, Sener S. Osteosarcoma; a case report. EUR J dent 2007; 1:60-3.
- [12]. Forteza G, Colmenero B, Lopez-barea F. Osteosarcoma of maxilla and mandible. oral surg oral med oral pathol 1986;62:179-84.
- [13]. Soheyl sheikh, shambulingappa pallagatti, Amit Aggarwal, Deepak Gupta, Nidhi pari, Amit mittal. Osteosarcoma of maxilla. A case report J clin exp dent. 2012;2(3); e117-20.