CASE REPORT

Maxillary Osteosarcoma Diagnosed from a Limited Biopsy: A Diagnostic Challenge

Sabu Paul[1], Srevya Rajeev[2], Latha Mary Cherian[3], Mahsoonabi B K[4], Gautam Kamal[5], Aparna Thomas[6]

Abstract

Background: Jaw osteosarcomas (JOS) are rare malignant bone tumours, accounting for 6.5% of all osteosarcomas. It presents with swelling, pain, tooth mobility or paresthesia, often mimicking odontogenic infections. Limited biopsy samples or lack of characteristic osteoid formation pose diagnostic challenges, leading to delays that impact survival.

Case presentation: A 63-year-old female presented with persistent swelling for one month following extraction of the upper right premolar. Swelling involved the edentulous alveolus and labial vestibule from tooth 12 to 15. CT scan revealed a lytic lesion with a soft tissue component and periosteal reaction in the right maxillary alveolar process, extending to maxillary sinus walls. Incisional biopsy revealed a pleomorphic tumour with a lace-like eosinophilic area but no frank osteoid formation. Immunohistochemical (IHC) markers Vimentin and SATB2, with a high Ki-67 index, were used to confirm the diagnosis of maxillary osteosarcoma.

Conclusion: Maxillary osteosarcoma may be misdiagnosed in limited biopsy specimens because sampling error and scant osteoid can obscure its characteristic histopathological features. A multidisciplinary approach integrating clinical, radiological, and histopathological findings, supported by immunohistochemistry, is crucial for accurate diagnosis and timely management.

Keywords: Jaw osteosarcoma, Malignant bone tumour, Osteoid, SATB2, Vimentin.

Introduction

Osteosarcoma (OS) is the most common primary malignant bone tumour and constitutes about 1% of all head and neck malignancies. Jaw osteosarcomas (JOS) are rare and account for approximately 6.5% of all OS.[1] Despite their rarity, JOS frequently poses a diagnostic challenge because its clinical and radiological features often mimic benign odontogenic and fibro-osseous lesions.[2]

The most common presenting complaint is painless swelling, often mimicking odontogenic pathologies. Two-thirds of patients have their teeth extracted as a result of misdiagnosis, and half of those individuals receive antibiotic treatment.[3] Histologically, diagnosis depends on identifying malignant osteoid; however, incisional biopsy specimens may be non-representative because of superficial sampling and scant osteoid, making definitive diagnosis challenging. Furthermore, distinguishing malignant osteoid from hyalinized collagen in limited biopsy specimens can be difficult. In such cases, SATB2 has emerged as a valuable adjunctive marker of osteoblastic differentiation.[4]

This case describes a maxillary osteosarcoma presenting after tooth extraction, in which significant intraoperative bleeding and limited biopsy material with scant osteoid contributed to diagnostic uncertainty. It highlights the importance of representative tissue sampling, multidisciplinary clinicoradiologic-pathologic correlation, and adjunctive immunohistochemistry in establishing an accurate diagnosis.

Department and Institution Affiliation: [1-6]Department of Oral Pathology & Microbiology, Government Dental College Kottayam, Kerala, India.

Corresponding Author: Dr. Srevya Rajeev, Department of Oral Pathology and Microbiology, Government Dental College, Kottayam, Kerala 686008. Email: srevyarajeev@gmail.com.

How to cite the article: Paul S., Rajeev S., Cherian L M., B K Mahsoonabi, Kamal G., Thomas A.Maxillary Osteosarcoma Diagnosed from a Limited Biopsy: A Diagnostic Challenge. Oral MaxillofacPathol J 2026; 17(2); 330-333.

Source of Support: Nil

Conflict of Interest: None

Case Report

A 63-year-old female presented to dental OPD with chief complaint of swelling on upper right back tooth region for past one month. She stated that the symptoms began the day after the extraction of her carious upper right premolar at a local dental clinic. The patient described the extraction as traumatic. She was prescribed analgesics and antibiotics, which alleviated the pain, however the swelling persisted. Two weeks later, she began experiencing a sensation of tightness over the right cheek area and subsequently reported to dental OPD. The patient's medical history was non-contributory, with no history of tobacco or alcohol use and no history of facial trauma.

Extraoral examination revealed diffuse swelling on right

side of zygomatic area of face. On palpation, the swelling was tender, firm in consistency, with a local rise in temperature. Intraorally, a localized swelling measuring approximately 3x2.5 cm was observed, extending anteriorly from the vestibule of teeth 12 to 15 posteriorly and inferiorly involving edentulous alveolar crest region (Figure 1). The swelling appeared slightly erythematous, with obliteration of the nasolabial fold. On palpation, swelling was firm in consistency with tenderness noted. No cervical lymphadenopathy was detected.

CT scan showed a lytic lesion with a soft tissue component and periosteal reaction measuring 4.4 x 3.0 x 3.1 cm involving the right half of alveolar process of maxilla extending to anterior and medial wall of right maxillary sinus. These findings raised suspicion of malignancy. Therefore, our provisional diagnosis was malignant bone tumour, with osteomyelitis and fibro-osseous lesions considered as differential diagnoses.

Histopathological examination of the incisional biopsy revealed parakeratinised stratified squamous epithelium overlying connective tissue infiltrated by a highly cellular neoplasm composed of pleomorphic round to spindle-shaped cells arranged in sheets (Figures 2 and 3). The tumour cells exhibited marked nuclear pleomorphism and hyperchromatism, with occasional vesicular nuclei. Scattered mitotic figures were identified. Focally, a lace-like eosinophilic extracellular matrix was present at the deeper aspect of the biopsy specimen (Figure 4). However, because of its limited extent and resemblance to hyalinized collagen, it was difficult to determine whether this represented malignant osteoid. Areas of haemorrhage with extravasated red blood cells were present within the tumour, correlating with the significant intraoperative bleeding encountered during biopsy. These features also raised the possibility of a vascular neoplasm in the differential diagnosis.

Immunohistochemistry (IHC) was performed for a confirmatory diagnosis. PanCK and CD31 were negative, ruling out an epithelial or vascular origin. Vimentin and SATB2 were positive, confirming mesenchymal and osteoblastic origin, respectively (Figure 5). The tumour also demonstrated a high Ki-67 proliferative index, supporting its malignant nature. Based on the histomorphological features and immunohistochemical findings, a diagnosis of maxillary osteosarcoma, most consistent with the osteoblastic variant, was rendered. The patient was referred to a higher centre for further management.

Discussion

Jaw osteosarcoma (JOS) differs from long-bone osteosarcoma in its clinical presentation, biological behaviour, and radiographic appearance. Patients are generally older at presentation, have a lower incidence of distant metastasis, and demonstrate better overall survival following complete surgical resection. Although the mandible is more commonly affected, Yildiz et al. reported a higher frequency of maxillary osteosarcomas in females, consistent with the present case.[5]

Clinically, LBOS typically presents with exercise-related bone pain, whereas JOS most commonly manifests as swelling, with pain developing later in the course of the disease.[6] Other presenting features include tooth mobility or loss, tooth displacement, muscle spasm, and paraesthesia, often leading to an initial misdiagnosis as chronic pulpitis, chronic periodontitis, or osteomyelitis, particularly when presenting as a non-healing extraction socket.[1] In the present case, the swelling developed following tooth extraction and was initially

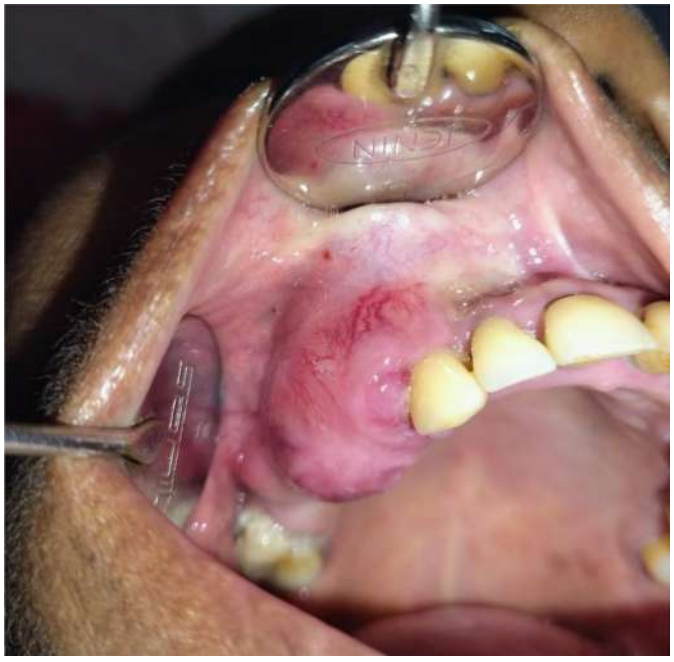

Fig. 1: Intraoral photograph showing a swelling in the anterior vestibule and edentulous alveolar ridge with obliteration of the nasolabial fold.

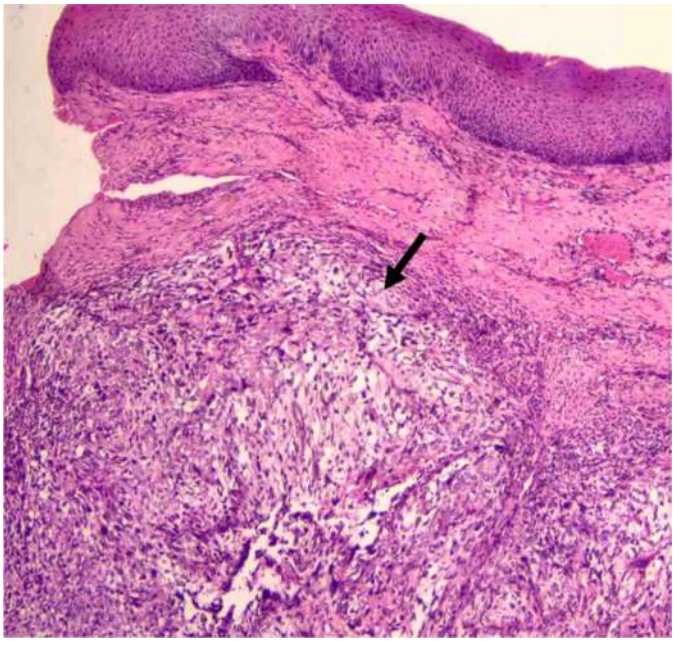

Fig. 2: Photomicrograph showing parakeratinised stratified squamous epithelium overlying connective tissue infiltrated by sheets of pleomorphic tumour cells (arrow). (H&E, x10).

considered a post-extraction complication, underscoring the importance of maintaining a high index of clinical suspicion and prompt biopsy to avoid delayed diagnosis and facilitate timely treatment.

Radiologically, jaw osteosarcomas frequently lack the classical features described in long-bone osteosarcoma, such as a sunburst periosteal reaction or Codman's triangle. Although widening of the periodontal ligament space may represent an early sign, it is not specific and may occur in other jaw lesions.[7,8] Nakayama et al. observed that maxillary osteosarcomas are predominantly osteogenic. In the absence of significant bone destruction, they may resemble benign cemento-osseous lesions, making radiological diagnosis challenging.[9,8] In the present case, CT demonstrated an aggressive lesion suggestive of malignancy but lacked definitive radiological features of osteosarcoma.

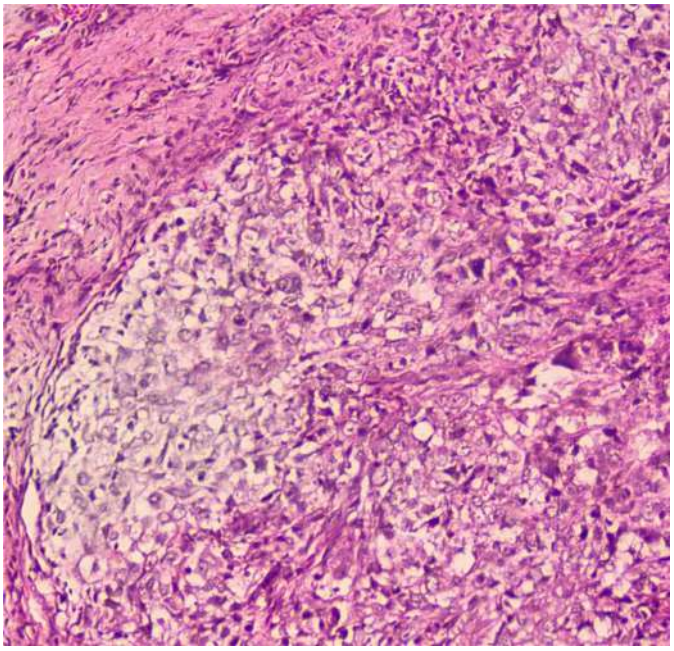

Fig. 3: Photomicrograph showing pleomorphic round and spindle-shaped tumour cells arranged in a disorganised pattern. (H&E, x40).

Histopathological features may vary greatly, but Lichtenstein stated the essential criteria as "the presence of a frankly sarcomatous stroma and the direct formation of tumour osteoid and bone by this malignant connective tissue".[10] However, tumour heterogeneity, superficial sampling, and limited biopsy material may result in minimal or absent osteoid, making definitive diagnosis difficult.[11] Furthermore, incisional biopsies may sample only the superficial soft tissue component, leading to under-representation of the malignant bone-forming component.[12] In the present case, biopsy sampling was further limited by significant intraoperative bleeding, resulting in a predominantly soft tissue specimen containing only focal lace-like eosinophilic matrix. Because this material closely resembled hyalinized collagen, the diagnosis of osteosarcoma could not be established confidently on routine H&E sections alone.[4]

In such challenging cases, immunohistochemistry serves as a valuable adjunct to routine histopathological examination. SATB2 (Special AT-rich Sequence Binding Protein 2) is a sensitive marker of osteoblastic differentiation, particularly in limited biopsy specimens where malignant osteoid is sparse or absent.[4] Several studies have demonstrated that strong SATB2 expression is present in OS, independent of the amount of malignant osteoid.[4,11,13] Other useful adjunctive markers

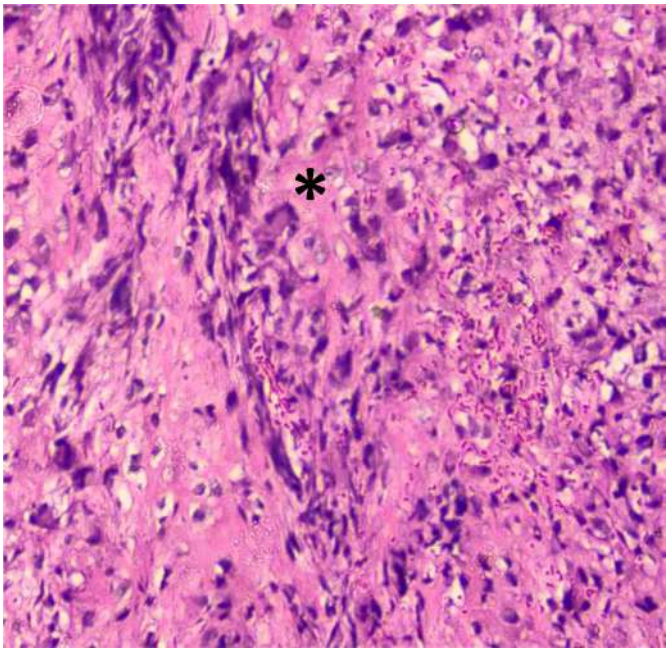

Fig. 4: Photomicrograph showing a focal lace-like eosinophilic extracellular matrix (asterisk), suspicious for malignant osteoid, at the edge of the biopsy (H&E, ×40).

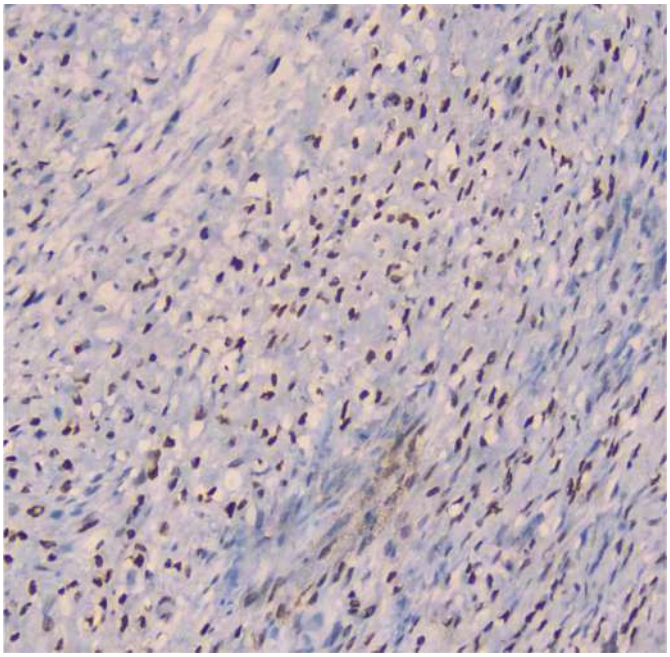

Fig. 5: Photomicrograph showing diffuse nuclear immunopositivity for SATB2 (×40).

include MDM2 and CDK4, particularly in distinguishing low-grade osteosarcomas from benign fibro-osseous lesions.[2,12] In this case, correlation of the patient's post-extraction clinical presentation, aggressive radiological features, limited biopsy morphology, and immunohistochemical findings was crucial in arriving at the final diagnosis.

Similar diagnostic challenges have been reported by Alramadhan et al., who described a mandibular osteosarcoma in which a superficial biopsy lacking representative malignant osteoid resulted in diagnostic uncertainty and necessitated repeat biopsy to establish the diagnosis.[12] In addition, Davis and Horvai emphasised that limited biopsy specimens with scant osteoid may preclude a definitive diagnosis and highlighted the value of SATB2 as an adjunctive marker in such cases.[11] These reports, together with the present case, underscore the importance of adequate biopsy sampling and multidisciplinary clinicoradiologic-pathologic correlation, supplemented by immunohistochemistry, in establishing an accurate diagnosis.

Radical surgical excision with histologically negative margins remains the cornerstone of treatment for jaw osteosarcoma. Because achieving negative margins is more challenging in the maxilla, early diagnosis is essential to facilitate timely definitive surgery and improve outcomes.[9,14] Adjuvant chemotherapy, with or without radiotherapy, may be considered in selected cases to enhance local control and survival.[15]

Conclusion

A biopsy is considered the gold standard for confirming osteosarcoma (OS) diagnoses.[16] ElKordy et al. reported that the incidence of faulty biopsy results of bone tumours ranged from 17 to 25%.[9] Therefore, it is strongly recommended that the incisional biopsies of suspected osteosarcoma lesions be as representative and abundant as possible. A cautious approach is recommended for conditions that mimic malignancy. A pathologic diagnosis alone from biopsy may sometimes be inconclusive, especially if the biopsy is small or non-representative. The diagnosis should be made only after carefully combining clinical, radiographic and microscopic findings.[17]

References

1. Mahajan A, Vaish R, Desai S, Arya S, Sable N, K. D'cruz A. Gnathic Osteosarcoma: Clinical, Radiologic, and Pathologic Review of Bone Beard Tumor. JGO. 2017 Dec;3(6):823–7. doi:10.1200/JGO.2016.006494
2. Owosho AA, Ladeji AM, Adesina OM, Adebiyi KE, Olajide MA, Okunade T, et al. SATB2 and MDM2 Immunoexpression and Diagnostic Role in Primary Osteosarcomas of the Jaw. Dentistry Journal. 2021 Dec 30;10(1):4. doi:10.3390/dj10010004
3. Soufflet E, Thibaut F. Case report of maxillary osteosarcoma with up-to date review. J Oral Med Oral Surg. 2020;26(2):2. doi:10.1051/mbcb/2020008
4. Conner JR, Hornick JL. SATB2 is a novel marker of osteoblastic differentiation in bone and soft tissue tumours. Histopathology. 2013;63(1):36–49. doi:10.1111/his.12138
5. Yildiz FR, Avci A, Dereci O, Erol B, Celasun B, Gunhan O. Gnathic osteosarcomas, experience of four institutions from Turkey. Int J Clin Exp Pathol. 2014 May 15;7(6):2800–8. PubMed PMID: 25031699; PubMed Central PMCID: PMC4097283.
6. Shah S, Mishra B, Tiwari N, Nikunj A. Osteosarcoma of jaws: Challenges in diagnosis. Journal of Oral and Maxillofacial Pathology : JOMFP. 2021 Jan 9;24(3):589. doi:10.4103/jomfp.JOMFP_142_20 PubMed PMID: 33967520.
7. Arora P, Rehman F, Girish KL, Kalra M. Osteosarcoma of mandible: Detailed radiographic assessment of a case. Contemp Clin Dent. 2013;4(3):382–5. doi:10.4103/0976-237X.118355 PubMed PMID: 24124311; PubMed Central PMCID: PMC3793566.
8. Nakayama E, Sugiura K, Ishibashi H, Oobu K, Kobayashi I, Yoshiura K. The clinical and diagnostic imaging findings of osteosarcoma of the jaw. Dentomaxillofacial Radiology. 2005 May;34(3):182–8. doi:10.1259/dmfr/71175262
9. ElKordy MA, ElBaradie TS, ElSebai HI, Amin AAE, KhairAlla SM. Osteosarcoma of the jaw: Challenges in the diagnosis and treatment. Journal of the Egyptian National Cancer Institute. 2018 Mar 1;30(1):7–11. doi:10.1016/j.jnci.2018.02.001
10. Unni KK, Inwards CY. Dahlin's Bone Tumors: General Aspects and Data on 10,165 Cases. Philadelphia: Wolters Kluwer; 2015. 410 p.
11. Davis JL, Horvai AE. Special AT-rich sequence-binding protein 2 (SATB2) expression is sensitive but may not be specific for osteosarcoma as compared with other high-grade primary bone sarcomas. Histopathology. 2016;69(1):84–90. doi:10.1111/his.12911
12. Alramadhan SA, Vyas R, Cohen DM, Bhattacharyya I, Islam MN, Reith JD. An Unusual Histopathological Presentation of Mandibular Osteosarcoma. Head Neck Pathol. 2024 Nov 19;18(1):121. doi:10.1007/s12105-024-01724-4 PubMed PMID: 39560895; PubMed Central PMCID: PMC11576681.
13. Machado I, Navarro S, Picci P, Llombart-Bosch A. The utility of SATB2 immunohistochemical expression in distinguishing between osteosarcomas and their malignant bone tumor mimickers, such as Ewing sarcomas and chondrosarcomas. Pathology - Research and Practice. 2016 Sep 1;212(9):811–6. doi:10.1016/j.prp.2016.06.012
14. Nthumba PM. Osteosarcoma of the jaws: a review of literature and a case report on synchronous multicentric osteosarcomas. World J Surg Onc. 2012 Dec;10(1):240. doi:10.1186/1477-7819-10-240
15. Mohanavalli S, Viswanathan MS, Karthikeyan R, Gnanaguru V, Sree Vijayabala G, Rani LJ. Osteosarcoma of the mandible and its management: A rare case report. Surgery Case Reports. 2024 Mar 1;1:100009. doi:10.1016/j.sycrs.2024.100009
16. Misaghi A, Goldin A, Awad M, Kulidjian AA. Osteosarcoma: a comprehensive review. SICOT-J. 2018;4:12. doi:10.1051/sicotj/2017028
17. Amini Shakib P, Foroughi R, Seyedmajidi M. Osteosarcoma of the Maxilla: A Rare Case with Unusual Clinical Presentation. J Dent Res Dent Clin Dent Prospects. 2013;7(3):177–81. doi:10.5681/joddd.2013.029 PubMed PMID: 24082991; PubMed Central PMCID: PMC3779379.