Indian Journal of Surgical Oncology (March 2024) 15 (Suppl 1):S172–S178
https://doi.org/10.1007/s13193-023-01846-1

CASE REPORT

A Rare Case Report of Osteosarcoma of Maxilla with Double Free-Flap Reconstruction

Surendra K. Dabas[1] · Nandini N. Menon[1] · Reetesh Ranjan[1] · Himanshu Shukla[1] · Bikas Gurung[1] · Sukirti Tiwari[1] · Yash Chaddha[2] · Ajit Sinha[1] · Rahul Kapoor[1] · Vinay Kumar Verma[1] · Saurabh Arora[3] · Ashwani Sharma[1] · Jasbir Singh[4] · Rishu Singal[5] · Sandeep Kumar Mohan[1] · Seema Sachan[6]

Received: 22 August 2023 / Accepted: 11 November 2023 / Published online: 21 November 2023
© The Author(s), under exclusive licence to Indian Association of Surgical Oncology 2023

Abstract

Osteosarcomas are rare and highly malignant bone tumours which are composed of malignant mesenchymal cells producing osteoid or immature bone. Maxillary osteosarcomas are rare tumours accounting for less than 1% of head and neck malignancies. Aggressive surgical resection is the main modality of treatment with good reconstruction. Due to the complex anatomy and location of maxilla as well as its proximity to the skull, resection with negative margins is always a challenge and so is the reconstruction so as to reduce the morbidity of the patient and to also give a good functional and cosmetic outcome. Clinical outcomes can be improved with administration of neoadjuvant or adjuvant chemotherapy in selected cases and radiotherapy in case of positive margins. A 41-year-old male patient presented to the outpatient department with complaints of a bulge over the hard palate for the past 1 year. CT scan showed a $6 \times 5 \times 4$ cm osseous expansile lesion arising from the maxillary bone. Biopsy of the tumour showed features of conventional high-grade osteosarcoma. Plate-preserving maxillectomy with tracheostomy was done followed by reconstruction with a double free flap. On post-operative day 1, the flap showed signs of venous congestion and a new free anterolateral thigh flap was done. Patient was discharged on day 7 with a Ryles tube and a tracheostomy tube in situ. Final histopathological examination showed that the tumour was a high-grade chondroblastic osteosarcoma. After regular post-operative visits in the outpatient department and evaluation with flexible laryngoscopy, patient was started on oral feed by day 10 and decannulated by day 15. He has been on regular follow-up for the past 1 year and shows no signs of recurrence or residual disease on clinical examination as well as imaging. Maxillary osteosarcoma is a rare bone tumour which requires accurate imaging and biopsy for accurate surgical planning. The ideal treatment modality is radical resection with negative margins and appropriate reconstruction. With the advent of microvascular surgery, free flaps form the backbone for reconstruction of such large defects.

Keywords Osteosarcoma · Maxillary neoplasm · Bone tumour · Sun ray appearance · Free flap · Microvascular surgery · Case report

✉ Nandini N. Menon
menonnandini1602@gmail.com

[1] Department of Surgical Oncology, BLK-MAX Super Speciality Hospital, Pusa Road, Rajendra Place, Delhi 110005, India

[2] Department of Head and Neck Oncology, NH-MMI Hospital, Dhamtari Road, Raipur, Chhattisgarh 492001, India

[3] Department of Nuclear Medicine, BLK-MAX Super Speciality Hospital, Pusa Road, Rajendra Place, Delhi 110005, India

[4] Department of Histopathology, BLK-MAX Super Speciality Hospital, Pusa Road, Rajendra Place, Delhi 110005, India

[5] Department of Radiodiagnosis, BLK-MAX Super Speciality Hospital, Pusa Road, Rajendra Place, Delhi 110005, India

[6] Department of Pathology, BLK-MAX Super Speciality Hospital, Delhi 110005, India

Introduction

Osteosarcoma is a malignant tumour in which tumour cells produce osteoid or immature bone [1]. It is the most common primary bone tumour which accounts for 40 to 60% of all primary malignant bone tumours [2]. It commonly occurs in the second to fourth decade. It mostly occurs in patients with genetic predisposition, underlying abnormalities like Paget's disease, fibrous dysplasia, previously irradiated bones, bone infarcts, chronic osteomyelitis, trauma, viral infection, exposure to high-dose radiation, metallic implants, joint prostheses, genetic syndromes such as Li-Fraumeni syndrome, and hereditary retinoblastoma [2].

Jaw osteosarcomas are extremely rare and comprise 7% of osteosarcomas and only 1% of head and neck malignancies [3]. Maxillary osteosarcomas more commonly affect the alveolar ridge, sinus floor, and palate in comparison to orbit and zygoma [4]. Unlike long bone osteosarcomas, jaw osteosarcomas occur more commonly in men [3]. Microscopically, 50% of jaw osteosarcomas are chondroblastic or osteoblastic [5].

Head and neck osteosarcomas have a greater tendency to recur locally and present with swelling, loosening of teeth, and facial dysaesthesia [3]. Distant metastases are less common in head and neck osteosarcomas compared to long bone osteosarcomas [3].

Jaw osteosarcomas can have mixed radiographic features which can be osteogenic, osteolytic, or mixed. Periosteal reaction that deposits bone spicules over the surface of the lesion can give it a typical "sun-ray appearance". Widening of periodontal ligament space and widening of mandibular canal are also important features of jaw osteosarcomas [6].

Prognosis of osteosarcomas mainly depends on the histological subtype, grade, tumour size, patient age, and response to chemotherapy [3]. If untreated, the prognosis of jaw osteosarcoma is very poor. The main modality of treatment is aggressive surgical resection with negative margins and advanced reconstruction [4]. The use of chemotherapy and radiotherapy in select cases is also slowly gaining popularity as they seem to improve the prognosis of patients [4]. Radiotherapy should be considered in patients with positive margins or in high-grade tumours [7] and also in patients who are not surgical candidates for local control.

Case Report

A 41-year-old male patient, with no comorbidities, presented to the outpatient department with complaints of a bulge over the hard palate for the past 1 year which has been gradually increasing in size. He had no other associated complaints like dysphagia, bleeding, weight loss, or anorexia. On examination, there was a 5 × 4 cm smooth bulge over the anterior aspect of the hard palate, extending across both sides of the midline and the soft palate appeared free (Fig. 1a). There was also associated swelling over the right side of the face. Diagnostic nasal endoscopy was done which showed a bulge in the lateral wall of the right nasal cavity and compressing the turbinate.

MRI of paranasal sinuses showed a hypointense lesion measuring 5.3 × 3.8 × 4.3 cm arising from the anterior aspect of the hard palate and anterior maxilla, bulging into the anterior nasal cavity and oral cavity with subtle spiculations seen along the periphery, likely to be an osseous lesion. PET CT scan (Fig. 2a-f) was done which showed a FDG avid lesion measuring 4.6 × 5.6 × 4.1 cm lesion arising from the anterior aspect of the hard palate and anteromedial aspect of both maxilla. The lesion appeared predominantly osseous with dense intralesional tumour matrix with fuzzy ground-glass attenuation. Superiorly, it appeared to extend to the anterior aspect of the right nasal cavity with close proximity to inferior turbinates, inferiorly bulging into the oral cavity with involvement of the root of incisors and canines on both sides of the maxillary alveolus. No evidence of regional or distant metastasis was noted. CBCT of the maxilla was done which demonstrated a 5.7 × 4.1 × 3.2 cm lesion with mixed radiopaque and radiolucent features, crossing the midline on either side. It extends from the alveolar crest involving the alveolar and basal bone and invading nasal fossa bilaterally and also invading anteromedial wall of both maxillary sinuses. A contrast-enhanced CT scan of the face with 3D reconstruction was done which showed an osseous expansile mass measuring 6.1 × 5.3 × 4.1 cm arising from the anterior wall of the maxilla with bony spicules radiating perpendicular to the cortex giving the sunburst appearance. Upper alveoli appeared to be involved and extended posterolaterally to the premolar region on both sides. It was seen eroding the floor of the bilateral nasal fossa and involving the bilateral inferior nasal turbinates. The inferior aspect of the nasal septum also appears involved. There is associated erosion of the anteromedial wall of bilateral maxillary sinuses. The lesion also involves the anterior aspect of the hard palate. Biopsy was performed which showed features consistent with high-grade conventional osteosarcoma of the chondroblastic type.

Patient was planned for upfront surgery in view of the possibility of complete resection of tumour and adequate reconstruction. He underwent bilateral orbital plate–preserving maxillectomy (class 2c maxillectomy according to the modified Brown classification) (Fig. 1b, e) with tracheostomy. The specimen was sent for histopathological examination (Fig. 1c, d). The reconstruction of the defect was done was with a double free flap which were free fibula osseocutaneous flap and a free iliac crest osseocutaneous flap

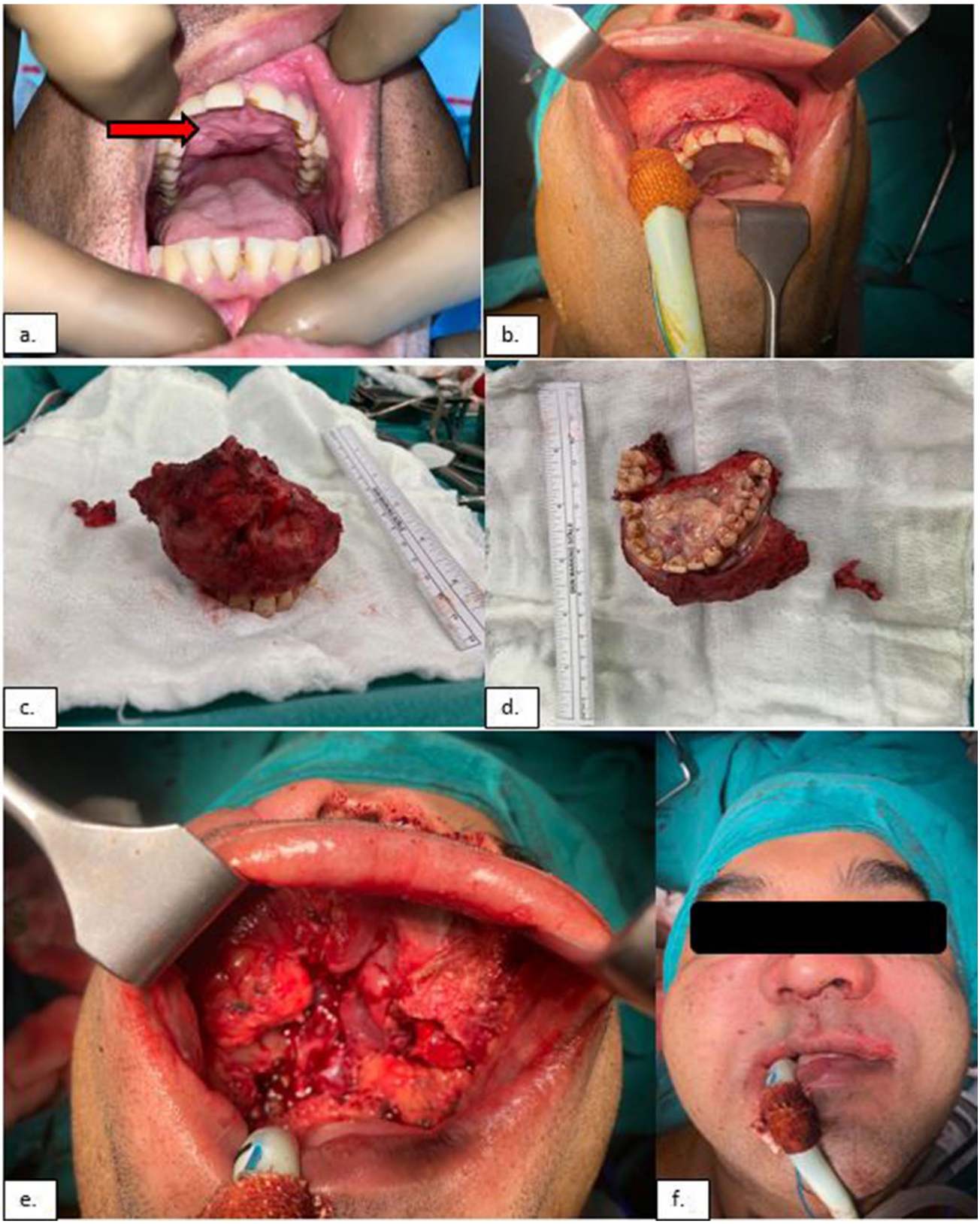

Fig. 1 **a** Preoperative appearance of the lesion over the hard palate marked with a red arrow. **b** Intraoperative appearance after giving intraoral incision and raising the cheek flap as well as the incision marked over the palate. **c, d** Gross appearance of resected specimen. **e** Primary defect after resection with negative margins. **f** Immediate post-operative appearance after reconstruction with double free flap

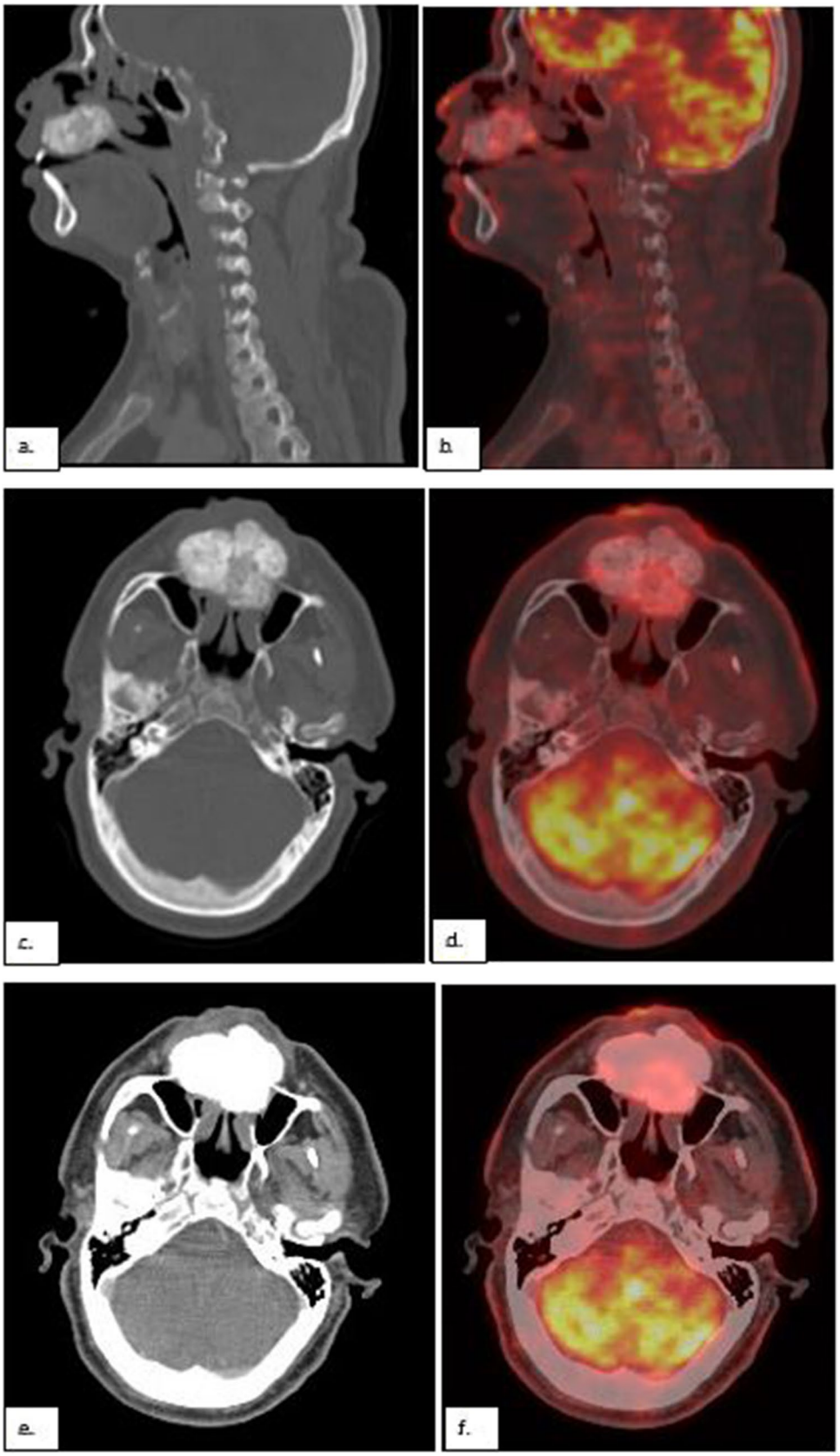

Fig. 2 Sagittal (**a**–**b**) and axial (**c**–**f**) CT and fused PET/CT images showing FDG avid lesion arising from the hard palate and anteromedial aspect of bilateral maxilla

(Fig. 1f). On post-operative day 1, the iliac crest flap showed features of venous congestion. The patient was taken for flap re-exploration during which debridement of internal oblique muscle was done and the iliac crest bone was covered with a free anterolateral thigh flap. During the immediate post-operative period, the patient was kept on Ryles tube feed. The patient was discharged on post-operative day 7 with a Ryles tube and a tracheostomy tube in situ.

Final histopathology report was 6 × 4.5 × 4.5 cm tumour centred at the maxilla with mild extension into the surrounding soft tissue. Maxillary sinus mucosa appears uninvolved by the tumour. Histologically (Fig. 3) it was identified as a high-grade chondroblastic osteosarcoma with no evidence of necrosis. There was no evidence of lymphovascular invasion. All margins were free from tumour involvement and closest soft tissue margin measured 0.5 cm. According to AJCC, it was termed a stage I disease.

Following discharge from hospital, he was on regular follow-up in the outpatient department. Flexible laryngoscopy was done on day 10 and repeated on day 15. Oral feed was started on day 10 and decannulation of the tracheostomy tube was done on day 15. He has been on regular follow-up for the past 1 year and there have been no signs of recurrent or residual disease on clinical examination as well as a PET scan done after 1 year.

Discussion

Osteosarcomas are a heterogenous group of malignancies characterized by osteoid or neoplastic ossification [8]. Jaw osteosarcomas tend to occur in the third or fourth decade [9]. The diagnosis of jaw osteosarcomas is often missed for a long time due to rarity of the tumour and atypical presentations. So thorough clinical examination is a key point which will help in early diagnosis of such cases. Early diagnosis plays an important role because it increases the chances of complete removal of the tumour in comparison to advanced stages. Although craniofacial osteosarcomas are a well-reported entity, maxillary osteosarcomas reported are very few in number. This is one of very few cases where we have been able to successfully operate a case of maxillary osteosarcoma and achieve negative margins along with good microvascular reconstruction giving excellent cosmesis and functional outcome to the patient.

Common presenting symptoms for jaw osteosarcomas include the following—swelling, local pain, facial dysaesthesia, loosening of teeth, trismus, and nasal obstruction.

Radiologically, osteosarcomas present with expansion of bone, as radiolucent, radio opaque, or mixed lesions [10]. Occasionally, widening of periodontal ligament can be seen which is known as the Garrington sign [9]. CT scan shows the irregular endosteal and extracortical bone with an obliterated or destroyed cortex along with sun-ray spiculations in soft tissue [11]. Bianchi and Boccardi classified the radiological appearances of osteosarcomas into three categories: (1) radio transparent with absence of bone formation within the tumour; (2) marbled with areas of amorphous ossification; and (3) lamellar ossification with sunshine pattern [12].

A tissue biopsy is required for definitive diagnosis and it is advised to take a deep biopsy from the centre of the lesion to avoid misdiagnosis [11]. Microscopically, osteosarcomas show a malignant and undifferentiated cellular stroma that produces neoplastic osteoid or bone. Although osteoid formation is pathognomic for the condition, variable amounts of chondroblastic and fibroblastic tissues can also be seen in the stroma. The chondroblastic variant has higher prevalence in osteosarcomas of head and neck and has better prognosis compared to the other variants [6]. Histologically, osteosarcomas are divided into central and peripheral subtypes. The main type of central osteosarcoma is the conventional osteosarcoma which has the following subtypes—osteoblastic, chondroblastic, and fibroblastic [13].

The main differential diagnosis for osteosarcoma includes osteomyelitis with proliferative periostitis, suppurative osteomyelitis, ossifying fibroma, osteoblastoma, and fibrous dysplasia [9]. Jaw osteosarcomas of the jaw are often difficult to diagnose due to vague and delayed presentation of patients, non-specific radiological features, and erroneous pathology reports. The incidence of false reports in bone tumours is as high as 17 to 25% [14]. Galactin 1 estimation can be done to differentiate osteosarcoma from chondrosarcoma; it is elevated in the former but not in the latter with an appositive predictive value of 85.7% [14].

The American Joint Committee on Cancer (AJCC) System for bone sarcomas has four stages: stage I for low-grade tumours, stage II for high-grade tumours, stage III for "skip metastasis", and stage IV for metastatic sarcomas. The Enneking system of classification divides sarcomas into stage I which are low-grade tumours, stage II for high-grade tumours, and stage III for metastatic tumours.

The ideal treatment for craniofacial osteosarcomas is complete surgical resection with negative margins but, due to the anatomical complexity of the maxilla and its proximity to vital structures coupled with the patients' apprehension regarding cosmesis and psychological uncertainty, makes the treatment extremely challenging. The role of adjuvant treatment in craniofacial osteosarcomas is still under debate, unlike osteosarcomas of extremities. Available data shows that adjuvant chemotherapy does not improve survival statistics in jaw osteosarcomas and if the surgical margins are not free of disease, radiation is also ineffective [15, 16].

Chemotherapy and radiotherapy in locoregionally advanced head and neck cancer have shown to improve the overall survival. Chemotherapy in neoadjuvant settings reduces the tumour size prior to surgery and in adjuvant settings it helps in local control and reduces the chances of

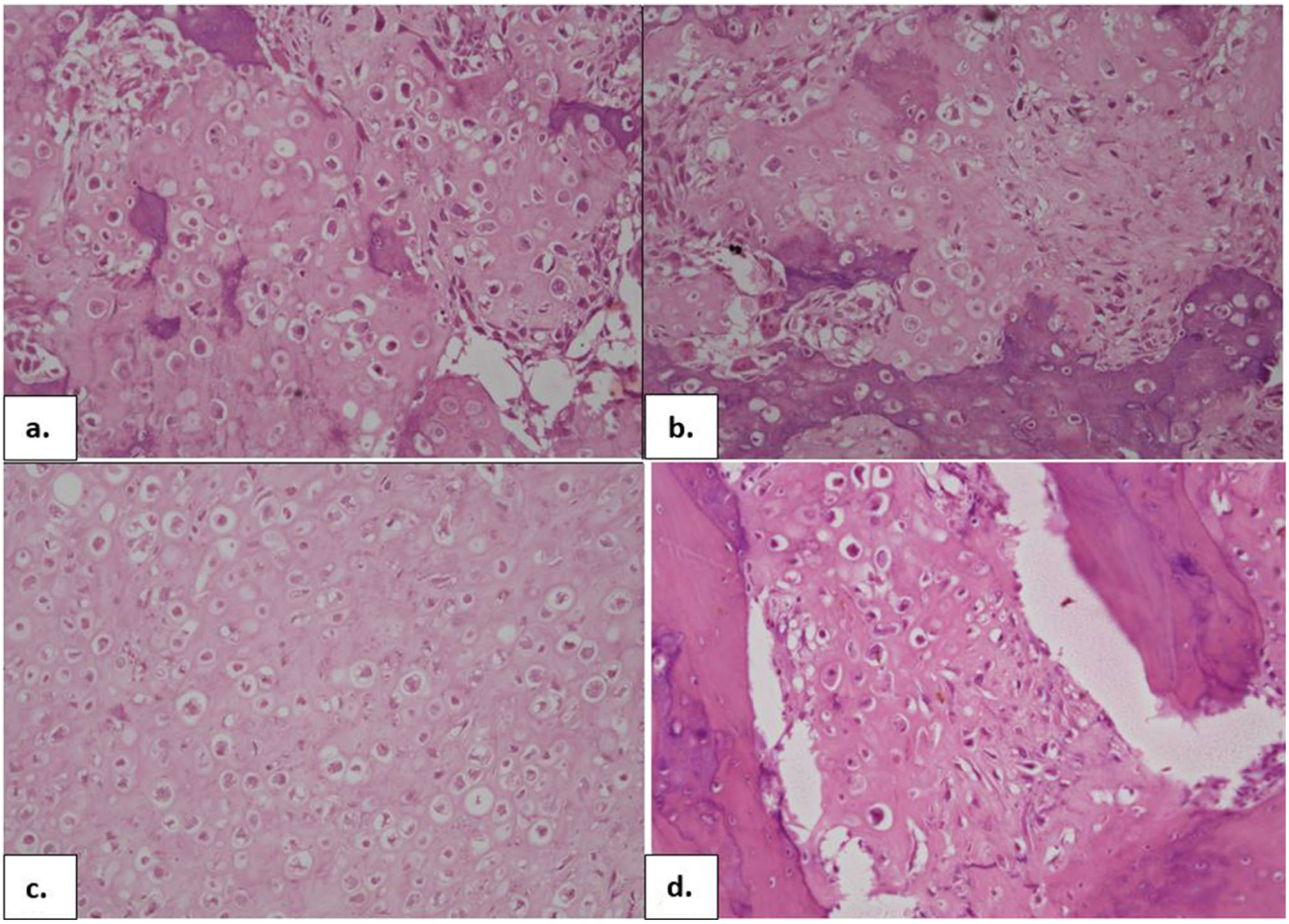

Fig. 3 Haematoxylin and eosin stained, ×40 images. **a**, **b** Showing chondroid areas with minimal scattered osteoid. **c** Section showing sheets of high-grade chondrocytes. **d** Section showing bony trabeculae with chondroid and minimal osteoid

distant metastasis. Radiotherapy in adjuvant settings has the fundamental role of locoregional control [17].

The role of adjuvant radiotherapy with jaw osteosarcoma was studied in 119 patients by Guadagnolo et al. In total, 92 patients underwent surgery alone and in 27 cases surgery was followed by radiotherapy. Stratified analysis by resection margin status demonstrated that the combined use of surgery and radiotherapy gave superior results in terms of better overall survival and disease-free survival in patients with positive or uncertain margins [7]. Two retrospective studies of osteosarcoma jaw have demonstrated the favourable effect of chemotherapy on survival [18, 19]. The roles of CTRT have been discussed in a systematic review of 201 patients from 20 uncontrolled series [20]. The 5-year overall survival and progression-free survival in patients undergoing multimodal therapy (surgery and neoadjuvant/adjuvant chemotherapy) were 80 and 75% respectively. The 5-year overall survival and disease-free survival in patients who were subjected to radical surgery alone were 40 and 33% respectively. In this review, it was concluded that chemotherapy improved survival when combined with surgery more than radiotherapy [21]. A single institutional study conducted by Ferrari et al. showed better 5-year overall survival and disease survival rates for patients who underwent radical surgery and chemotherapy with or without radiotherapy [22].

The most important factor determining prognosis in these cases is completeness of surgery. Although multimodality treatment has shown to improve clinical outcomes, for small T1 lesions, adequate surgery still is the best treatment modality with no requirement of neoadjuvant or adjuvant treatment.

Jaw osteosarcomas have better prognosis than long bone osteosarcomas except those jaw osteosarcomas associated with Paget's disease which are more aggressive and have a poor prognosis [23]. The main prognostic criteria for jaw osteosarcomas include tumour size, possibility of complete resection at the time of presentation, and osteoblastic subtype [24, 25]. Females with predominantly chondroblastic pattern may have poorer prognosis [26]. Complete resection of maxillary osteosarcomas is technically more challenging leading to higher chances of positive margins and local recurrence of disease following surgery [3].

Conclusion

Osteosarcoma maxilla is an aggressive disease and requires early diagnosis and surgical intervention at the earliest so as to attain negative surgical margins. Reconstruction of large defects should be given importance so that it gives the patient less morbidity and good functional and cosmetic outcomes. In higher staged tumours, neoadjuvant and adjuvant therapy may play an important role.

Acknowledgements I would like to acknowledge the efforts of the Department of Robotics and Surgical Oncology, Department of Radiology and Pathology of BLK – MAX Super Speciality Hospital.

Data Availability All materials submitted in the article are original.

Code Availability Not applicable.

Declarations

Informed Consent Written and informed consent has been taken from the patient for the procedure and all procedures have been done ethically.

Conflict of Interest The authors declare no competing interests.

References

1. Soufflet E, Thibaut F (2020) Case report of maxillary osteosarcoma with up-to date review. J Oral Med Oral Surg 26:15. https://doi.org/10.1051/mbcb/2020008
2. Klein MJ, Siegal GP (2006) Osteosarcoma: anatomic and histologic variants. Am J Clin Pathol 125(4):555–581. https://doi.org/10.1177/1066896908319675
3. August M, Magennis P, Dewitt D (1997) Osteogenic sarcoma of the jaws: factors influencing prognosis. Int J Oral Maxillofac Surg 26(3):198–204
4. Jasnau S, Meyer U, Potratz J, Jundt G, Kevric M, Joos UK, Jürgens H, Bielack SS (2008) Craniofacial osteosarcoma experience of the cooperative German-Austrian-Swiss osteosarcoma study group. Oral Oncol 44(3):286–294. https://doi.org/10.1016/j.oraloncology.2007.03.001
5. Nakayama E, Sugiura K, Kobayashi I, Oobu K, Ishibashi H, Kanda S (2005) The association between the computed tomography findings, histologic features, and outcome of osteosarcoma of the jaw. J Oral Maxillofac Surg 63(3):311–318. https://doi.org/10.1016/j.joms.2004.04.033
6. Altuwairgi O, Papageorge MB, Karp DD (1996) Maxillary chondroblastic sarcoma: presentation of two cases and a literature review. J Oral Maxillofac Surg 54:1357–1364
7. Guadagnolo BA, Ashleigh Guadagnolo B, Zagars GK, Kevin Raymond A, Benjamin RS, Sturgis EM (2009) Osteosarcoma of the jaw/craniofacial region. Cancer 115(14):3262–3270
8. Lima Verde MEQ, Nogueira AS, De Negreiros WA, Hirth CG, CarreiroFilho FP, LimaMota MR, Nunes Alves APN (2017) Chondroblastic osteosarcoma of maxilla: a case report. Oral Surg Oral Med Oral Pathol OralRadiol 124:e100. https://doi.org/10.1016/j.oooo.2017.05.204
9. Yeşilova E, Akgünlü F, Dolanmaz D, Yaşar F, Sener S (2007) Osteosarcoma: a case report. Eur J Dent 1:60–63. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2612954/
10. Amaral MB, Buchholz I, Freire-Maia B, Reher P, de Souza PE, MarigoHde A et al (2008) Advanced osteosarcoma of the maxilla: a case report. Med Oral Patol Oral Cir Bucal 13:E492–E495
11. Cabral LA, Werkman C, Brandão AA, Almeida JD (2009) Imprint cytology of osteosarcoma of the jaw: a case report. J Med Case Reports 3:9327
12. Bianchi SD, Boccardi A (1999) Radiological aspects of osteosarcoma of the jaws. Dentomaxillofac Radiol 28:42–47
13. Chaudhary M, Chaudhary SD (2012) Osteosarcoma of jaws. J Oral Maxillofac Pathol 16(2):233–238. https://doi.org/10.4103/0973-029X.99075
14. ElKordy MA, ElBaradie TS, ElSebai HI et al (2018) Osteosarcoma of the jaw: challenges in the diagnosis and treatment. J Egypt Natl Canc Inst 30:7–11
15. Mardinger O, Givol N, Talmi YP, Taicher S, Saba K, Hashomer T (2001) Osteosarcoma of the jaw. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 91:445–451
16. Van Es RJJ, Keus RB, Van der Waal I, Koole R, Vermey A (1997) Osteosarcoma of the jaw bones. Int J Oral Maxillofac Surg 26:191–197
17. Yildiz FR, Avci A, Dereci O, Erol B, Celasun B, Gunhan O (2014) Gnathic osteosarcomas, experience of four institutions from Turkey. Int J Clin Exp Pathol 7(6):2800–2808
18. Link MP, Goorin AM, Horowitz M, Meyer WH, Belasco J, Baker A, Ayala A, Shuster J (1991) Adjuvant chemotherapy of high-grade osteosarcoma of the extremity: updated results of the multi-institutional osteosarcoma study. Clin Orthop Relat Res 270:8–14
19. Doval DC, Kumar RV, Kannan V, Sabitha KS, Misra S, Vijay Kumar M, Hegde P, Bapsy PP, Mani K, Shenoy AM, Kumarswamy SV (1997) Osteosarcoma of the jaw bones. Br J Oral Maxillofac Surg 35(5):357–362
20. Smeele LE, Kostense PJ, van der Waal I, Snow GB (1997) Effect of chemotherapy on survival of craniofacial osteosarcoma: a systematic review of 201 patients. J Clin Oncol 15(1):363–367
21. Ferrari D, Codecà C, Battisti N, Broggio F, Crepaldi F, Violati M, Bertuzzi C, Dottorini L, Caldiera S, Luciani A, Moneghini L, Biglioli F, Cassinelli G, Morabito A, Foa P (2014) Multimodality treatment of osteosarcoma of the jaw: a single institution experience. Med Oncol 31(9):171. https://doi.org/10.1007/s12032-014-0171-9
22. Bertoni F, Dallera P, Bacchini P, Marchetti C, Campobassi A (1991) The Istituto Rizzoli-Beretta experience with osteosarcoma of the jaw. Cancer 68(7):1555–1563
23. Ferrari D, Moneghini L, Allevi F, Bulfamante G, Biglioli F (2017) Osteosarcoma of the jaw : classification , diagnosis and treatment. Osteosarcoma - biology, behavior and mechanisms. Chapter 12:197–219. https://doi.org/10.5772/67564
24. Gadwal SR, Gannon FH, Fanburg-Smith JC, Becoskie EM, Thompson LD (2001) Primary osteosarcoma of the head and neck in pediatric patients: a clinicopathologic study of 22 cases with a review of the literature. Cancer 91(3):598–605
25 Steve M, Ernenwein D, Chaine A et al (2011) Jaw osteosarcomas. Rev Stomatol Chir Maxillofac 112:286–292
26. Paparella ML, Olvi LG, Brandizzi D et al (2013) Osteosarcoma of the jaw: an analysis of a series of 74 cases. Histopathology 63:551–557

Publisher's Note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Springer Nature or its licensor (e.g. a society or other partner) holds exclusive rights to this article under a publishing agreement with the author(s) or other rightsholder(s); author self-archiving of the accepted manuscript version of this article is solely governed by the terms of such publishing agreement and applicable law.