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CASE REPORT

Massive Bilateral Maxillary Osteosarcoma: a Dramatic Clinical Presentation and a Reconstructive Challenge

Arvind Krishnamurthy[1]

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Introduction

Osteosarcomas are the most common primary bone malignant tumors in adults. These tumors commonly arise from the metaphysis of long bones of the extremity; their occurrence in the head and neck region is rare. Head and neck osteosarcomas account for < 10% of all cases of osteosarcomas and about 1% of all the head and neck malignancies [1]. Bilateral maxillary involvement at presentation is extraordinarily rare and to the best of our knowledge has never been reported before. We describe an unusual case of a massive bilateral maxillary osteosarcoma in a 38-year-old gentleman and review the limited literature with regard to its overall management along with the associated reconstructive challenges.

Case Report

A 38-year-old gentleman presented to our center with complaints of painless swelling over the midfacial region for 6 months. Clinical evaluation revealed a large midfacial swelling involving both the maxillary regions, more on the left side, and also associated broadening of the external nose (Fig. 1a, b). There was no significant cervical lymphadenopathy.

A CT scan of the head and neck showed an expansile lytic lesion measuring 8.7 × 6.5 × 5.6 cm involving both sides of the body, the alveolar process, and the palatine part of the maxilla with a central area of dense calcification (Fig. 2a, b). A plain skiagram of the skull further in fact demonstrated the classical sunburst appearance (Fig. 3).

The patient underwent a left total and a right subtotal maxillectomy and reconstruction with a vascularised fibula flap after a biopsy confirmation of osteosarcoma (Fig. 4a, b). The final histopathology confirmed the presence of a high-grade osteoblastic osteosarcoma resected with close but clear margins. The patient subsequently received 60 Gy of adjuvant external beam radiation therapy along with 6 cycles of ifosfamide-, adriamycin-, and cisplatinum-based chemotherapy after discussion in the multispecialty board. The patient is presently disease-free and on regular follow-up for the past 20 months with good esthetic and functional outcomes as well (Fig. 5a, b).

Discussion

Osteosarcomas are rare malignant bone tumors which commonly arise from the metaphysis of long bones. The head and neck region is a rare subsite of osteosarcoma, with less than 10% of all cases of osteosarcomas accounting for about 1% of all the head and neck malignancies [1].

The exact etiology of osteosarcomas is obscure; however, some predisposing factors have been implicated, which include exposure to radiation, Paget's disease of bone, multiple osteochondromatosis, fibrous dysplasias, and even chronic osteomyelitis.

Clinically, osteosarcomas presents with myriad symptoms which include bony swelling, facial deformity, paresthesias, and toothache. These tumors rarely present with symptoms of ulceration, epistaxis, visual problems, or nasal obstruction [2]. Bilateral maxillary involvement causing a gross facial

✉ Arvind Krishnamurthy
drarvindkrishnamurthy@yahoo.co.in

[1] Surgical Oncology, Cancer Institute (WIA), 38, Sardar Patel Rd, Adyar, Chennai 600036, India

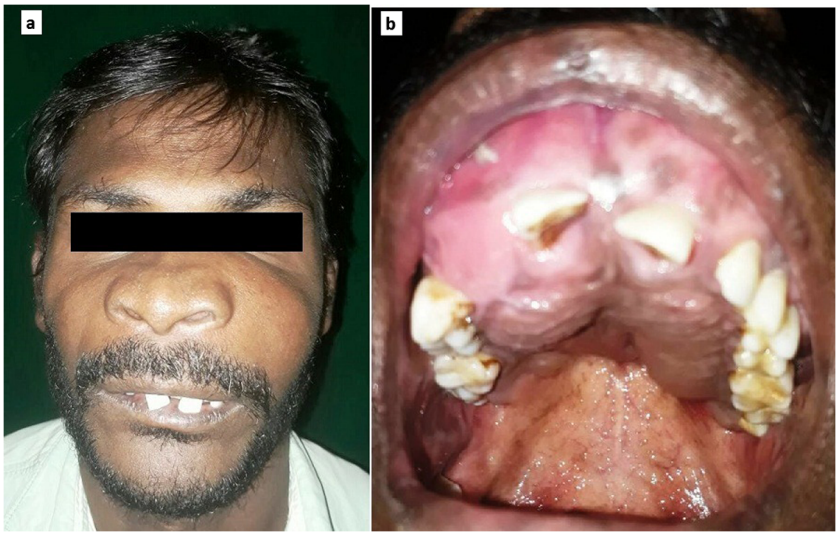

Fig. 1 **a**, **b** Clinical photograph at presentation

deformity was an unusual clinical presentation in our patient (maxillary involvement more on the left side).

The management philosophy of head and neck osteosarcomas is primarily a multidisciplinary approach which primarily consists of radical surgical resection with negative margins [3, 4]. The complex anatomy of the maxillofacial region makes both resection and reconstruction a major challenge [5, 6]. Various flaps are used to reconstruct the defect following radical excision of the tumor; these include the temporal flap or the microvascular free flaps (including the radial forearm flap and the anterolateral thigh flap). With the advent of virtual surgical planning technology, the reconstruction of complex maxillary and midfacial defects can be optimized through preoperative planning with a 3D printing bone model by computer-aided design and manufacturing [5, 6]. Considering the extensive facial deformity caused by the resection of both the maxilla, a decision to reconstruct this complex defect with a free fibula was considered in our patient.

The exact roles of radiotherapy and chemotherapy in the overall management of head and neck osteosarcomas are unclear [4]. Adjuvant radiation therapy is generally recommended for patients with close/positive surgical margins and/or other adverse prognostic factors [4]. The role of chemotherapy in head and neck osteosarcomas also remains unclear, largely due to the conflicting results of published data [7]. The role of chemotherapy, especially neo-adjuvant chemotherapy, in

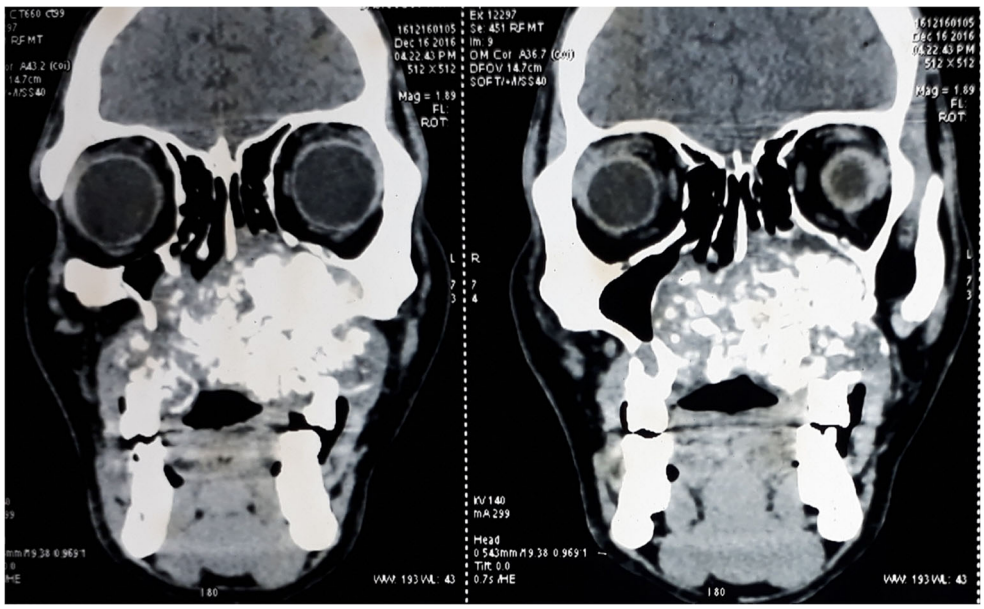

Fig. 2 **a**, **b** CT scan (coronal) of the skull showed an expansile lytic solid lesion of size 8.7 × 6.5 × 5.6 cm, which was seen extending posteriorly up to the level of the root of the third molar tooth on the left side and first molar tooth on the right side with destruction of the floor of the nasal cavity and bilateral maxillary sinuses (left > right) with obstruction of the osteomeatal unit

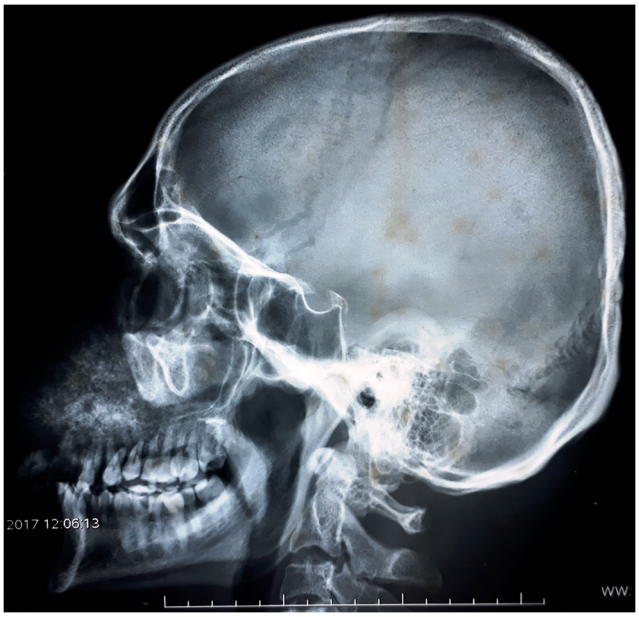

Fig. 3 A plain skiagram of the skull (lateral view) showed a mixed lytic sclerotic lesion involving the whole of the maxilla on the left eroding the anterior, alveolar, and nasal surface, extending to the infratemporal surface and zygomatic process crossing the midline and involving the contralateral maxilla, typically showing a sunburst appearance

head and neck osteosarcomas continues to evolve [8]. Adjuvant radiation and chemotherapy was considered in our patient in view of the unusually large size of the tumor at presentation and the close surgical margins.

Local recurrences are more common in osteosarcomas of the head and neck region with a reported incidence of 17–70% as compared to a local recurrence rate of about 5–7% in extremity osteosarcomas [9]. On the contrary, distant metastases tend to occur less frequently than from osteosarcomas arising from the long bones. There is sparse data on the various prognostic factors influencing the survival outcomes [4, 9]. Adverse outcomes have been reported for tumors > 6 cm, age > 60 years, a non-mandibular tumor location, an osteoblastic histology, non-surgical initial therapy, and for surgical margin positivity [10].

In conclusion, osteosarcomas of the head and neck region are rare malignant bone tumors that occur primarily in the jawbones and seem to have a more aggressive clinical course when compared with their counterparts in the long bones of the extremity. An appreciation and understanding of this unique biology should aid clinicians in better managing these rare tumors.

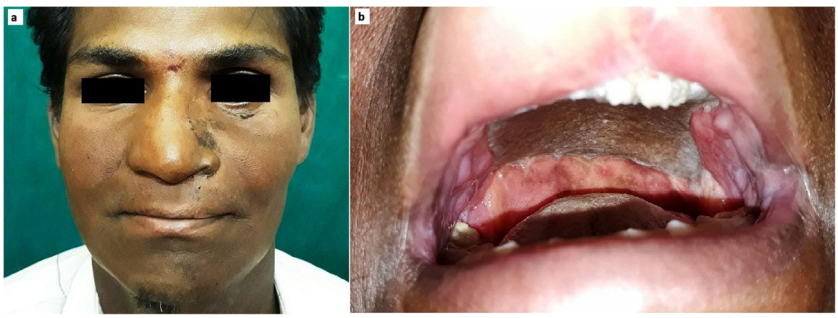

Fig. 4 **a**, **b** Postoperative clinical photograph. The free fibula was fashioned as an arch to reconstruct the facial contour with the help of multiple osteotomies, fixing to a prearched plate and then anchoring onto the zygomatic bone on both sides. The skin paddle was used to reconstruct the hard palate and enable oronasal separation. Posteriorly, the skin edge is folded superiorly to form the nasal floor

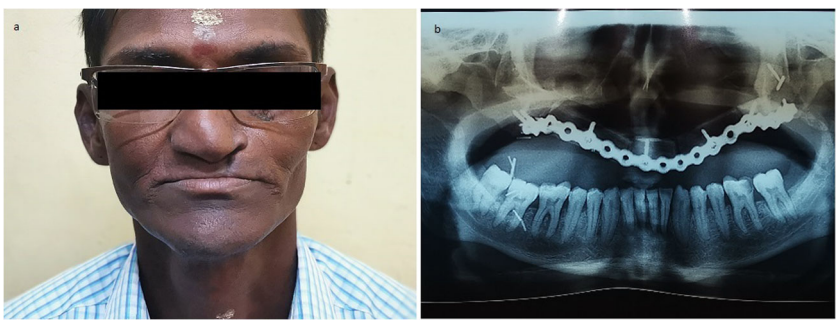

Fig. 5 **a** Clinical photograph after a follow-up of 20 months. **b** Orthopantomogram at follow-up of 20 months showing the prearched plate anchored onto the zygomatic bone

Compliance with Ethical Standards

Declaration of Patient Consent The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of Interest The authors declare that they have no conflict of interest.

References

1. Sturgis EM, Potter BO (2003) Sarcomas of the head and neck region. Curr Opin Oncol 15:239–252
2. Bennett JH, Thomas G, Evans AW, Speith PM (2000) Osteosarcoma of the jaws: a 30-year retrospective review. Oral Surg Oral Med Oral Pathol 90:323–333
3. Canadian Society of Otolaryngology-Head and Neck Surgery Oncology Study Group (2004) Osteogenic sarcoma of the mandible and maxilla: a Canadian review (1980–2000). J Otolaryngol 33: 139–144
4. Krishnamurthy A, Palaniappan R (2018) Osteosarcomas of the head and neck region: a case series with a review of literature. J Maxillofac Oral Surg 17:38–43
5. Melville JC, Manis CS, Shum JW, Alswied D (2018) Single-unit-3D-printed titanium reconstruction plate for maxillary reconstruction: the evolution of surgical reconstruction for maxillary defects-a case report and review of current techniques. J Oral Maxillofac Surg 77(4):874
6. Xue R, Lai Q, Sun S, Lai L, Tang X, Ci J, Zhang Z, Wang Y (2018) Application of three-dimensional printing technology for improved orbital-maxillary-zygomatic reconstruction. J Craniofac Surg 30: e127–e131. https://doi.org/10.1097/SCS.0000000000005031
7. 9. Guadagnolo BA, Zagars GK, Raymond AK, Benjamin RS, Sturgis EM (2009) Osteosarcoma of the jaw/craniofacial region. Cancer 115:3262–3270
8. 10 Chen Y, Shen Q, Gokavarapu S, Lin C, Yahiya Cao W, Chauhan S, Liu Z, Ji T, Tian Z (2016) Osteosarcoma of head and neck: a retrospective study on prognostic factors from a single institute database. Oral Oncol 58:1–7
9. 7. Kassir RR, Rassekh CH, Kinsella JB, Segas J, Carrau RL, Hokanson JA (1997) Osteosarcoma of the head and neck: metaanalysis of nonrandomized studies. Laryngoscope 107: 56–61
10. 8. Mendenhall WM, Fernandes R, Werning JW, Vaysberg M, Malyapa RS, Mendenhall NP (2011) Head and neck osteosarcoma. Am J Otolaryngol 32:597–600