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Case Report

Very Enlarged Mandibular Osteosarcoma

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Abstract

In this study, we aimed to present a huge mandibular tumor. In our study, we presented a 29-year-old patient with mandibular tumor. Osteosarcoma is the most common malignant tumor of bone that occurs mostly in the young age group. It is most common in the second decade. The most important areas of bone involvement are the metaphysis of the load-bearing joints. Osteosarcoma is rarely seen in the head and neck region. The most affected area in the head and neck region is the mandible. No matter how large jaw tumors are, they can be operated on with an experienced surgeon and adequate repair.

Keywords: Mandible; Osteosarcoma; Mesenchymal Tumors

Introduction

Sarcomas are malignant tumors originating from mesenchymal cells such as bone, cartilage, muscle, fat, and nerve. Osteosarcoma generally occurs in the long bones of adolescent patients in the skeletal system. In contrast, jaw osteosarcomas are most common in the 3rd and 4th decades. After the femur, tibia and humerus, the fourth most common location is the jaw bones [1]. It most commonly affects the mandible, maxilla, and calvarium in the head and neck region, respectively [2]. In the our case, the mentioned lesion occurred in an adult patient, which corroborates the majority of reports in the literature, which indicate the greater prevalence of the lesion in individuals in the 1st and 2nd decade of life [3]. Osteosarcoma (OS) is a malignant neoplasm characterised by the formation of osteoid matrix by neoplastic cells. It is the most common primary malignant bone tumor accounting for 20% of all sarcomas, although its occurrence in the jaw is rare. It shows typical clinical behaviour but varied radiological and histopathological features [4]. In this report, we presented the clinical, radiological and pathological findings of a young male patient with severely enlarged osteosarcoma in the mandible.

Case report: A 29-year-old male patient applied to us with the complaint of swelling on his face growing for 7 months. No surgical or medical treatment was given to the patient until he was admitted to us. In the examination, a lesion starting from the right mandible mentum and extending to the condyle and invading the floor of the mouth was observed. A preliminary diagnosis was made through anamnesis, physical examination and imaging methods, and a definitive diagnosis was made after the operation. No biochemical examination was applied to the patient preoperatively, except for anesthesia preparation. A definitive diagnosis was made by pathological examination. No systemic disease was detected in the patient. No metastasis was detected in the patient (Figure 1).

In the neck, tomography imaging was observed that a heterogeneous central calcific mass lesion measuring 10 cm at its widest point invading the mandible and maxilla on the right half of the face, leading to destruction in the right zygomatic arch. This mass also extended to the level of the vascular space and continued up to the medial pterygopalatine fossa (Figure 2).

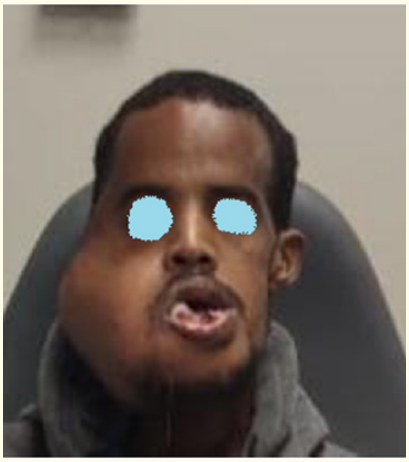

Figure 1: On physical examination, mass starting from the right mandible mentum and extending to the condyle.

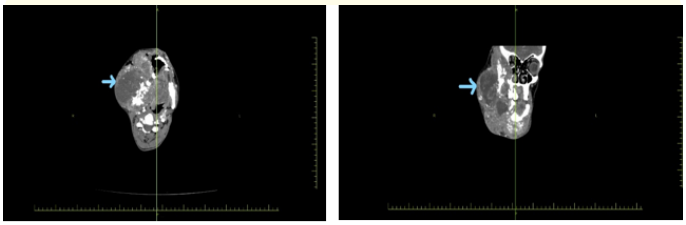

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Figure 2: a: CT axial section. b: CT Coronal section.

On neck magnetic resonance imaging, a heterogeneous mass lesion of 18x15 cm in size of mandibular origin was observed in the right face region. It had necrosis areas and partially occupied the maxillary sinus, partially involving the pterygopalatine fossa and muscles and extending superiorly to the temporal level, periorbital and intraorbital areas. In addition, it was observed that the mass extended to the base of the tongue and oral cavity, invading the oropharynx, nasopharynx, and partially inferiorly to the hypopharynx (Figure 3).

In FDG Positron emission tomography, there was an intense hypermetabolic mass lesion in the right half of the face, but no metastasis was detected in other parts of the body (Figure 4).

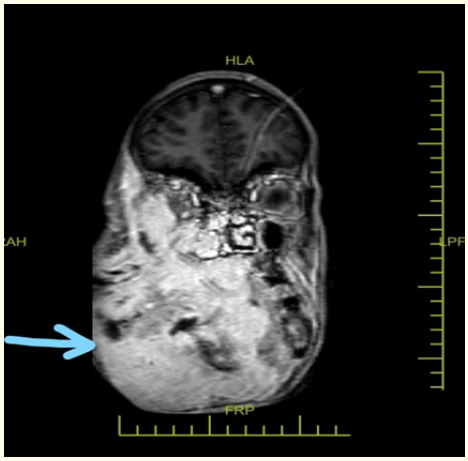

Figure 3: Neck MRI (Magnetic resonance imaging).

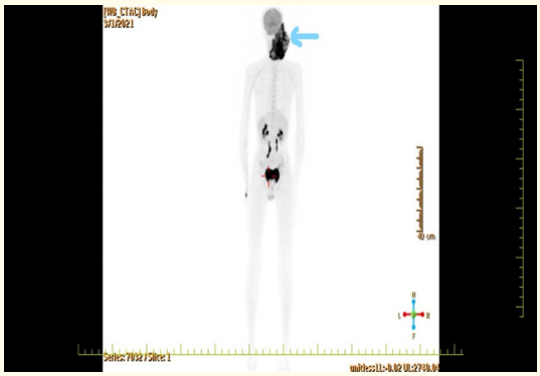

Figure 4: Positron emission tomography (PET).

Surgical procedure

Under general anesthesia, the Y incision extending from the mastoid type to the middle of the lower lip was extended to the clavicle. The subplastymal flap was extended to the zygomatic arch and elevated. The mass was dissected from the tragus and reached to the tragal pointer. The mass was retracted from the tragus to the anterior area, the facial nerve trunk was found, and the nerve was sacrificed because the nerve was surrounded by the tumor 360 degrees. It was observed that the coronoid process and the zygomatic arch were destroyed. The temporomandibular joint was disarticulated from the glenoid fossa and the mass was released

inferiorly. The mandible was cut from the left lateral part of the midline by performing a full-thickness osteotomy. The tumor had invaded the right part of the floor of the mouth up to the base of the tongue and anterior fold. The mass was lateralized by making an anterior to posterior incision on the floor of the mouth. Infero-lateral maxillectomy was performed due to tumor invasion in the postero-superior and tubel region of the maxilla. The tumor was resected en-bloc, together with the ramus and corpus of the mandible. The defect was repaired with a pectoralis major myocutaneous flap (Figure 5a-5b).

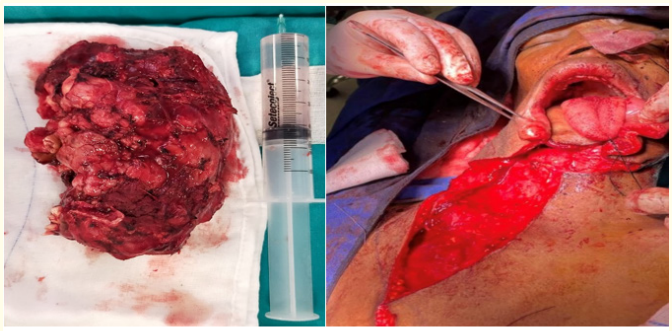

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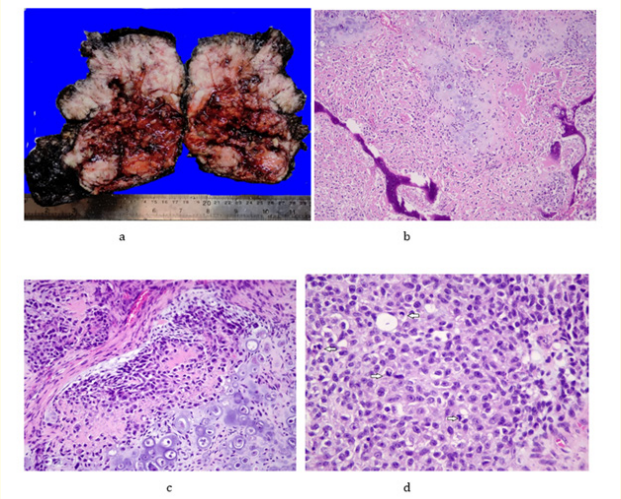
Figure 5: a: Tumoral lesion; b: Myocutaneous flap.



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c d

Figure 6: a: Gross pathology photos of the surgical specimen, the cut-surface of the resected tumor. b: Replacement of bone marrow by the tumor cells and the lace-like osteoid deposition. Note the tumor component with chondroblastic differentiation and the high-grade atypia, H&E, x100. c: Atypical osteoblasts with plasmacytoid morphology producing both osteoid and chondrogenic matrices, H&E, x200. d: Increased mitotic activity (arrows) and the high-grade atypia of the tumor cells, H&E, x400.

The patient received nutrition and wound care in our service for 2 months in the postoperative period.

On histopathological examination, a high-grade malignant neoplasm producing osteoid matrix and showing extensive areas of chondroblastic differentiation was revealed. Neoplastic cells were high-grade in morphology by being possessed of hyperchromatic, pleomorphic large nuclei with increased nucleo-cytoplasmic ratio and eosinophilic cytoplasms. Mitotic activity was brisk (15 per 10 hpf) and necrosis was present (15% of the tumor).

Discussion

The clinical and biological behaviour of long-bone and jaw osteosarcomas slightly differs. Head and neck osteosarcomas have a tendency to recure locally and frequencey sypmtoms are swelling site of disease facial disesthesia and loosining of the teeth [5]. There were clinical findings in our case, caused by a malignant lesion such as facial anesthesia, pain, and swelling. Although histological subtypes have no effect on treatment and prognosis in conventional osteosarcomas, they are important in the differential diagnosis of osteosarcomas. The differential diagnosis of osteosarcoma includes benign lesions such as osteoblastoma, chondroblastoma, and fibrous dysplasia, as well as some other malignancies invading bone tissue such as chondrosarcoma and Ewing's sarcoma. The presence of cellular atypia and surrounding tissue invasion is important in the differential diagnosis from benign lesions. In cases of osteosarcoma containing chondroblastic areas, it is necessary to see the osteoid matrix structure in the tumor cells in order to make the differential diagnosis of chondrosarcoma. Chondrosarcoma, which is almost always seen in advanced ages, but osteosarcoma shows bimodal age distribution. Chondrosarcoma of the jaw bones has been reported very rarely [6]. The age of our patient is compatible with the literatüre. Histologically, osteosarcoma is divided into the central and peripheral subtypes. The main type of central osteosarcoma is the conventional osteosarcoma, which is represented by a wide spectrum of morphologies. Depending on the predominant type of extracellular matrix present, conventional osteo-

sarcoma is classified histopathologically in osteoblastic, chondroblastic and fibroblastic subtypes. The osteoblastic subtype consists of osteoid or immature bone surrounded by haphazardly arranged fibroblast-like or epithelioid cells. The chondroblastic variant shows areas of atypical hyaline chondroid tissue [7]. In our case, high-grade conventional osteosarcoma was detected. The tumor contained chondroblastic and osteoblastic areas. Difficulties may be encountered when diagnosing chondroblastic OS and can be mistaken as chondrosarcoma especially in the cases when the osteoid is absent in incisional biopsy and/or in unrepresentative tissue samples. Chondrosarcomas are relatively rare in jaw bones and show better prognosis than osteosarkoma [8]. Osteosarcomas in the jaw relatively rare and the majority of them arise de novo but some of them may develop in bone affected by Paget's disease, fibrous dysplasia, trauma, viral infection, exposure to high-dose radiation, joint prostheses and genetic syndromes such as Li-Fraumeni syndrome, hereditary retinoblastoma. No specific risk factor was found in the history of our case [9]. In a study by Mardinger., *et al*. the histological subtype with the highest prevalence in jaw tumors are chondroblastic osteosarcoma, followed by osteoblastic osteosarcoma [10]. In some studies, it has been shown that the osteoblastic pattern is more dominant, followed by the chondroblastic pattern [11]. There is no consensus on differentiation patterns in jaw tumors and more often it has heterogeneous histology [12]. Rare but important histological subtype of central jaw osteosarcoma is the low-grade central osteosarcoma (LGOC). High-grade, Grade 3 tumor was detected in our case. This is a well-differentiated osteosarcoma consisting of spindle cell fibroblastic proliferation with low cellularity, no significant atypia, low mitotic figures and a variable osteoid production. The most important feature of LGCO in long bones, and also in the jaw, is its similarities with benign lesions, first of all with fibrous dysplasia [13]. Yoshida., *et al*. reported that immunohistochemical analysis shows 100% sensitivity and 97.5% specificity for the diagnosis of low-grade osteosarcoma [14]. However, Tabareau-Dalanlande., *et al.* noted discordant results, with 33% of ossifying fibromas and 12% of fibrous dysplasias exhibiting MDM2 amplification by qRT-PCR but no cases exhibiting MDM2 overexpression by immunohistochemistry. These investigators also showed amplification of an MDM2 neighbor, RASAL1, in all the fibro-osseous lesions with MDM2 amplification but in none of the low-grade osteosarcomas studied [15]. We did not perform an immunohistochemical study in our case. After the anamnesis and physical examination, the imaging method should first be a direct X-ray. Later, whole body bone scintigraphy, magnetic resonance imaging (MRI), computed tomography (CT), and positron emission tomography (PET) may be requested. In scintigraphy, osteoblastic activity of the lesion on direct X-ray, skip metastases in the same bone and bone metastases in the skeleton can be revealed. CT can be used to obtain detailed information about cortex invasion in osteosarcoma. PET-CT is valuable in the treatment of osteosarcoma in terms of evaluating the response to chemotherapy and showing whole body metastases [16]. CT scanning is rarely used as a diagnostic tool in patients with chondroblastoma. This imaging modality is usually reserved for more severe or recurrent tumours. CT scans can depict matrix mineralization, an extension of the tumour into the soft tissue, and erosion of cortical bone. Coronal and sagittal reconstructions can be used to assess the extension of the tumour across the epiphyseal plate into the metaphysis of the bone [17]. The diagnosis of osteosarcoma can be made by fine needle aspiration biopsy and incisional biopsy after anamnesis, clinical examination and imaging [18]. As a result of anamnesis, physical examination, and imaging methods, we operated on the patient and confirmed the diagnosis by performing an excisional biopsy. The first step in the treatment of osteosarcoma is chemotherapy. After induction chemotherapy, the operation is complated with wide excision margins and negative surgical margins [19]. We applied the treatment plan as stated in the literature. Osteosarcomas in the head and neck region have high morbidity and mortality due to the complex anatomy and difficulty of complete resection. The best treatment results can be achieved with neoajjuvan chemotherapy and adjuvant chemotherapy after surgical resection. The effectiveness of chemotherapy in the treatment of osteosarcoma in the jaw bones is not clearly known. Although radiotherapy is recommended in inoperable cases, it is not effective in osteosarcoma of the jaw bone. Elective neck dissection is not recommended because lymph node metastasis is rare. Tumor grade and surgical margin are the most important parameters in determining prognosis in osteosarcoma. While survival reaches 75% in patients with a negative surgical margin, this rate regresses to 32% in the presence of residual tumor. 5-year survival is 74% for low-grade tumors but 42% for high-grade tumors. In the jaw bones, the anterior mandible has the best prognosis, while the anterior maxilla has the worst prognosis because of its difficult of resection [20]. The two main prognostic criteria of JOS are tumor size and resectability at presentation [21]. In the high-grade osteosarcomas, the best curative option is represented by a multimodal

treatment. On the other hand, the treatment of low-grade central and parosteal osteosarcomas can rely on surgery alone, provided a complete assessment of their metastatic potential [22]. Wide surgical resection is an effective treatment because it provides a clean surgical margin, a lower risk of local recurrence, and a higher overall survival [23]. Although adequate margins being the first goal of head and neck surgery in osteosarcoma, it requires a careful balance between effective surgery and function-saving procedures. Surgical planning and the technical execution should be based on the expectation of performing a functionally effective reconstructive surgery [24]. In a study by Bertoni., *et al.* it was reported that poor results were probably due to inadequate surgery, the presence of positive surgical margins, and the inefficiency of surgery [25]. Death is usually secondary to local tumor extension with neural and vascular infiltration [26]. Our case had lymphovascular invasion and a negative surgical margin. Preoperative and adjuvant kemoterapi has become the standard of care in long bone osteosarcomas, its role in jaw tumors is still controversial [27]. Excellent results have been obtained with the application of surgery, Neo-adjuvant chemotherapy and adjuvant chemotherapy [28]. In our study, following the surgical treatment, the patient received chemotherapy and radiotherapy. Adding CT or RT to surgery has demonstrated improved survival in locoregionally advanced head and neck cancer. The aim of chemotherapy is to reduce tumor size ameliorating surgical outcome, improve local control and reduce distant metastases. RT is usually employed in the adjuvant setting and has the fundamental role of decreasing locoregional relapse [29].

Conclusion

In conclusion, the achievement of osteosarcoma treatment is directly related to the grade and surgical margin of the tumor. In places such as the head and neck region, the morbidity and mortality of the operation result are high. However, no matter how large the tumor is in this region, the operation can be performed successfully with an experienced surgeon and adequate reconstruction.

Author Contributions

The first author (A.B.), corresponding author (A.B.), and the co-author (Ş.Ö.F.M.D.-S.T.) contributed to the writing and preparation of this article.

Declaration of Conflicting İnterests

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Ethical Approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed Consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

Bibliography

1. Pardhe ND., *et al*. "Giant Osteosarcoma of the Mandible". *Journal of College of Physicians and Surgeons Pakistan* 27.9 (2017): S117-S119.

2. Kyle S and Ettinger KA. "Sarcomas of the Jaws". In: Fonseca RJ (ed) Oral and Maxillofacial Surgery, Volume 2, Third Edition. (2018): 643-669.

3. Etanaiara Almeida., *et al*. "Chondroblastic osteosarcoma". *Journal of Oral and Maxillofacial Pathology* 18.3 (2014): 464-468.

4. Bajpai M., *et al*. "Osteogenic Sarcoma of Mandible". *Journal of College of Physicians and Surgeons Pakistan* 27.3 (2017): S26-S27.

5. "Osteosarcoma-Biology Behavior and Mechanism". Edited by Kanya Honoki and Kurt Richard Weiss. Intech Open (2017).

6. The WHO Classification of Tumours Editorial Board. WHO Classification of Tumours Soft Tissue and Bone Tumours, 5th ed. Lyon: IARC Press (2020).

7. Chaudhary M and Chaudhary SD. "Osteosarcoma of jaws". *Journal of Oral and Maxillofacial Pathology* 16.2 (2012): 233-238.

8. Bajpai, M and Pardhe N. "Chondroblastic osteosarcoma: the wolf in sheep's clothing". *Cukurova Medical Journal* 43 (2018) 761-762.

9. George A and Mani V. "Gnathic osteosarcomas: review of literature and report of two cases in maxilla". *Journal of Oral and Maxillofacial Pathology* 15.2 (2011): 138-143.

10. Paparella ML., *et al*. "Osteosarcoma of the jaw: an analysis of a series of 74 cases". *Histopathology* 2013;63 (4): 551-557.

11. Argon A., *et al*. "Osteosarcomas of jaw: experience of a single centre". *Journal of Plastic Surgery and Hand Surgery* 49.1 (2015): 13-18.

12. Bennett JH., *et al*. "Osteosarcoma of the jaws: a 30-year retrospective review". *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology* 90.3 (2000): 323-332.

13. Tabatabaei SH., *et al*. "Diagnostic challenges of low-grade central osteosarcoma of jaw: a literature review". *Journal of Dentistry* 16.2 (2015): 62-67.

14. Yoshida A., *et al*. "MDM2 and CDK4 immunohistochemical coexpression in high-grade osteosarcoma: correlation with a dedifferentiated subtype". *The American Journal of Surgical Pathology* 36.3 (2012): 423-431.

15. Tabareau-Delalande F., *et al*. "Chromosome 12 long arm rearrangement covering MDM2 and RASAL1 is associated with aggressive craniofacial juvenile ossifying fibroma and extracranial psammomatoid fibro-osseous lesions". *Modern Pathology* 28.1 (2014): 48-56.

16. Greanspan A., *et al*. "Differential Diagnosis in Orthopaedic Oncology". Bone Forming (Osteogenic) Lesions. 2nd ed. Philadelphia: Lippincott Williams and Wilkins (2007): 88-89.

17. Greenspan A., *et al*. "Differential diagnosis of tumours and tumour-like lesions of bones and joints". Lippincott Williams and Wilkins (1998).

18. Murat Hız. "Osteosarcoma, malignant fibrous histiocytoma clinical radiology, pathology and treatment strategies". *TOTBİD Dergisi* 13 (2014): 227-239.

19. Picci P., *et al*. "Risk factors for local recurrence after limb-salvage surgery for highgrade osteosarcoma of the extremities". *Annals of Oncology* 8.9 (1997): 89903.

20. Shah JP., *et al*. "Bone Tumors and Odontogenic Lesions". Jatin Shah's Head And Neck Surgery And Oncology, Fıfth Edıtıon. (2020): 701 - 753.

21. Gadwal SR., *et al*. "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 (2001): 598-605.

22. The ESMO/European Sarcoma Network Working Group. "Bone sarcomas: ESMO clinical practice guidelines for diagnosis, treatment and follow-up". *Annals of Oncology* 23 (2012): vii100-vii109.

23. Ketabchi A., *et al*. "Sarcomas of the head and neck: a 10-year retrospective of 25 patients to evaluate treatment modalities, function and survival". *British Journal of Oral and Maxillofacial Surgery* 49.2 (2011): 116-120.

24. Picci P., *et al*. "Relationship of chemotherapy-induced necrosis and surgical margins to local recurrence in osteosarcoma". *Journal of Clinical Oncology* 12.12 (1994): 2699-2705.

25. Bertoni F., *et al*. "The Istituto Rizzoli-Beretta experience with osteosarcoma of the jaw". *Cancer* 68.7 (1991): 1555-1563.

26. Mardinger O., *et al*. "Osteosarcoma of the jaw. The Chaim Sheba Medical Center experience". *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology* 91.4 (2001): 445-451.

27. Bacci G., *et al*. "Neoadjuvant chemotherapy in the treatment of osteosarcoma of the extremities: preliminary results in 131 cases treated preoperatively with methotrexate and cis-diamminoplatinum". *Italian Journal of Orthopaedics and Traumatology* 14.1 (1988): 23-39.

28. Thiele OC., *et al*. "Interdisciplinary combined treatment of craniofacial osteosarcoma with neoadjuvant and adjuvant chemotherapy and excision of the tumour: a retrospective study". *British Journal of Oral and Maxillofacial Surgery* 46.7 (2008): 533-536.

29. “Osteosarcoma of the Jaw: Classification, Diagnosis and Treatment”. Daris Ferrari, Laura Moneghini, Fabiana Allevi, Gaetano Bulfamante and Federico Biglioli. Additional information is available at the end of the chapter.