

Case Study**Osteogenic Osteosarcoma of the Orbit: a case report****1. Abstract**

Osteogenic osteosarcoma of the orbit is a rare and aggressive type of osteosarcoma. Head and neck osteosarcomas occur most commonly in the third and fourth decades and have equal sex predisposition. A 56-year-old male patient was admitted to our clinic with proptosis on the left side and ptosis that had been present for five months. Ophthalmic examination revealed normal eye movements and vision. There was no pain and no paresthesia or any other significant complaint or clinical finding. Magnetic resonance imaging studies showed a hyperintense, destructive extraconal mass lesion adjacent to the lateral wall invading retrobulber fat tissue of the left orbita. The patient underwent a lateral orbitotomy. Histopathological examination revealed osteogenic osteosarcoma of the orbit. Herein, we report a case with osteogenic osteosarcoma arising from the orbit is presented. The differential diagnoses of this tumor and the surgical results of radical resection are also presented.

Keywords: Osteosarcoma, orbit, tumor, exenteration.

INTRODUCTION

Osteogenic osteosarcoma of the orbit is an extremely rare osteogenic tumor (1-4). Osteogenic osteosarcoma is a malignant bony tumor mesenchymal cell origin and because of the relative rarity of this condition, there are no clearly defined evidence-based treatment guidelines for the management of these tumors (3). In this report, we describe the clinical, radiologic, and pathologic records of a rare case of primary osteogenic osteosarcoma of the orbit in a middle-aged man, with no previous history of trauma or surgery.

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27 CASE REPORT

28 A 56-year-old male patient was admitted to our clinic with proptosis on the left side
29 and ptosis that had been present for five months. At presentation at our clinic, his visual
30 acuity was 10/10 in the right eye and 10/10 in the left eye. Ophthalmic examination revealed
31 normal eye movements and vision. There was no pain and no paresthesia or any other
32 significant complaint or clinical finding. Physical examination was otherwise unremarkable.

33 Magnetic resonance imaging studies showed a hyperintense, destructive extraconal
34 mass lesion adjacent to the lateral wall invading retrobulbar fat tissue of the left orbita. The
35 mass was lying extraconally and measuring 4 x 3 x 3 cm.

36 The patient underwent a lateral orbitotomy. Exposure of the mass was achieved
37 through a lateral orbitotomy. The zygomatic arch was also removed to obtain more exposure
38 of the tumor. Intraoperatively, the tumor was found attached to the lateral wall of the orbit.
39 The tumor appeared lobulated with a smooth capsule (Fig 1).

40 Macroscopically, the tumor had a well-circumscribed cut surface, was grayish white,
41 and had a solid nodular pattern. Histopathologic examination demonstrated extremely cellular
42 and contained irregular cells accompanied by high mitotic activity and mineralized regions
43 containing osteoid and the tumor cells had large hyperchromatic nuclei (Fig 2). Tumor
44 invasion into the adjacent tissue was observed. With regard to these histopathological
45 findings, the diagnosis of osteosarcoma was established.

46 Due to the invasive nature of the mass, the patient was referred to another center for
47 chemotherapy, radiotherapy or exenteration without any further intervention. He underwent
48 further excision of this mass. No metastasis was found during chest, abdominal, and pelvis CT
49 scans. No tumor recurrence or distant metastasis occurred during one year of follow-up.



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51 **FIGURE 1:** Gross apperance of the mass, The tumor appeared lobulated with a smooth
52 capsule

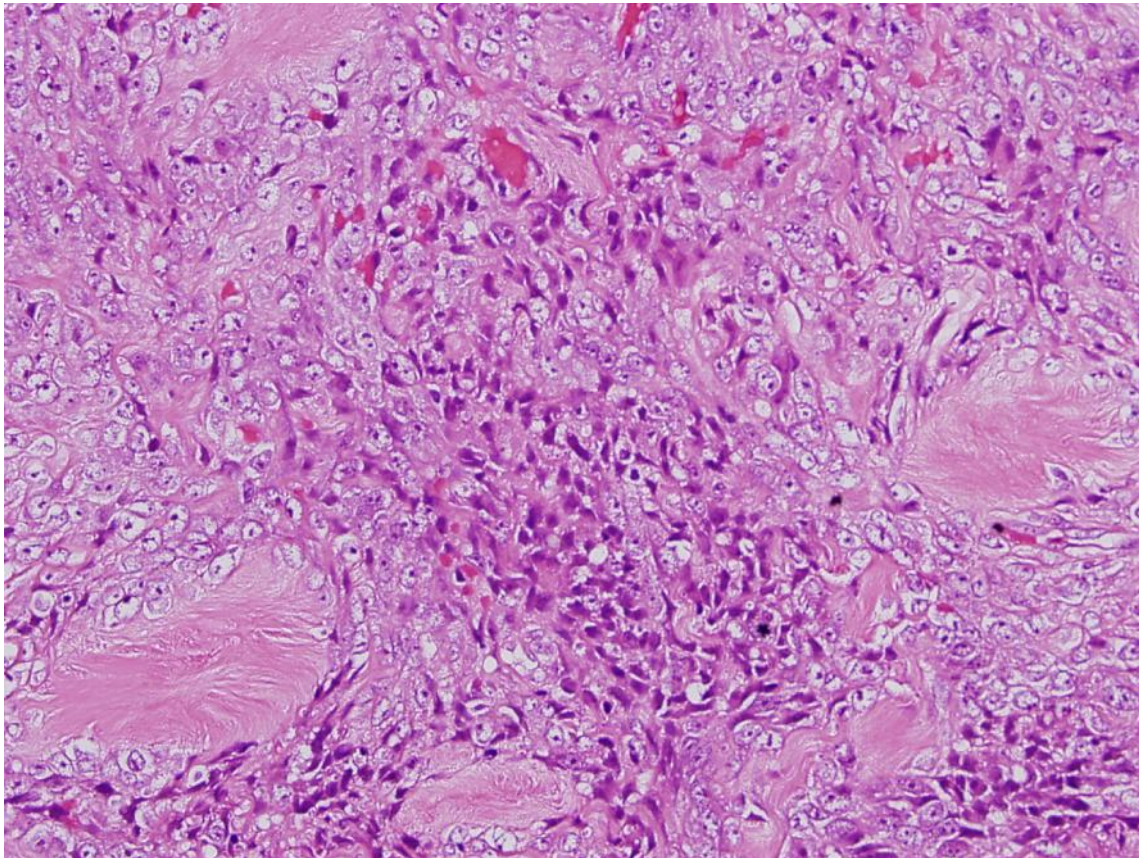


FIGURE 2: Hematoxylin-eosin stain, original magnification x400, Osteogenic osteosarcom with extremely cellular and contained irregular cells accompanied by high mitotic activity and mineralized regions containing osteoid.

DISCUSSION

Osteogenic osteosarcomas of the head and neck origin are rare and aggressive malignancies (3-6). Approximately 6% to 13% these tumors occur in the head and neck, and less than 2% occur in the cranium (2, 4). Predisposing conditions are Paget's disease of the bone, exposure to radiation, fibrous dysplasia, multiple osteochondromatosis, chronic osteomyelitis, myositis ossificans, and trauma (7).

In the previous studies some histopathological subtypes were defined as chondroblastic, osteoblastic, fibroblastic and telangiectatic osteosarcoma. Chondroblastic and osteoblastic osteosarcoma were the most common subtypes (3, 8).

66 Histologic diagnosis of parosteal osteosarcoma is often difficult, with the differential
67 diagnosis including diverse conditions such as myositis ossificans and metaplastic bone
68 formation in carcinoma, melanoma, synovial sarcoma and epithelioid sarcoma (9-12).

69 In our patient, the malignant cells and the absence of 'zonation' excluded a benign
70 reactive condition such as myositis ossificans. Immunostains with melanocyte and epithelial
71 markers were negative (S100, HMB-45, MELAN-A, EMA, LMWCK, HMWCK,
72 Kromogranin respectively), thus excluding melanoma and carcinoma. In contrast, osteogenic
73 sarcoma is characterized by a highly cellular pleomorphic anaplastic stroma, which forms
74 amorphous islands of osteoid in addition to cartilage and fibrous tissue (13-15).

75 Although the optimal management of patients with osteosarcoma of the head and neck
76 is still unclear, adequate surgical resection is the mainstay treatment (4, 13-15) stated that the
77 prognosis in osteosarcoma of the skull is poor and identified late diagnosis, early intracranial
78 extension, and limited surgical excision as responsible factors. Osteogenic osteosarcomas
79 occasionally invade the surrounding tissue, as in our case.

80 Surgeons might be reluctant to perform wide resections because of the complex
81 anatomy of the fronto-orbital region and the very high rate of recurrence. When non-negative
82 surgical margins are obtained, re-resection or adjuvant radiotherapy should be added as
83 additional treatment. Smeele et al (16) concluded that chemotherapy improves survival and
84 advocated the use of similar chemotherapy protocols as used for osteosarcoma of the long
85 bones.

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87

88 **CONCLUSION**

89 Optimal management of patients with osteosarcoma of the head and neck is still
90 unclear because of the relative rarity of this condition. There are no clearly defined evidence-

91 based treatment guidelines for the management of these tumors. Adequate surgical resection
92 is the most important step in treatment. Recurrence would be lower or the tumor-free period
93 would be longer if negative margins could be obtained.

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