Case Report

# Orbital Metastatic Osteosarcoma

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**Abstract**

At an estimated incidence of 2 cases per million persons per year, osteosarcoma is the most common primary malignant bone tumor in children and adults, excluding hematopoietic intraosseous tumors. Orbital metastases of osteosarcoma are very rare. Only 5 cases of orbital metastasis of osteosarcoma previously reported in the literature. We report the case of a 19-year-old man with known history of osteosarcoma of right distal femur who presented with acute visual loss and progressive protrusion of his left eye. Orbital CT scan and MRI revealed orbital mass eroding orbital walls and intracranial invasion. He underwent superotemporal orbitotomy for debulking of orbital mass. Histopathological examination (HPE) of the specimen was reported as metastatic osteosarcoma with extensive tumor necrosis. Then he underwent adjuvant chemotherapy and palliative radiotherapy. Although orbital metastasis of osteosarcoma is a rare event, it seems it has had an increasing trend recently. so, making efforts to palliate the patient's symptoms by multidisciplinary teamwork and proper interaction among ophthalmologist, orthopedic surgeons and oncologists is necessary.

**Keywords:** Osteosarcoma, Orbit, Metastasis, Palliative chemoradiotherapy

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## Introduction

At an estimated incidence of 2 cases per million persons per year, osteosarcoma is the most common primary malignant bone tumor in children and adults, excluding hematopoietic intraosseous tumors. It occurs primarily in the long bones and typically metastasizes to the lungs.[1,2]

The most common tumors to metastasize to the orbit are adenocarcinomas in adults and embryonal tumors or sarcomas, especially neuroblastoma, in children.[3] Orbital metastases of osteosarcoma are very rare. A review of literature in MEDLINE revealed only 5 cases of orbital metastasis of osteosarcoma.[4–8]

Here, we intend to present the sixth case of orbital metastases of osteosarcoma and compare it with previous reports.

## Case Report

A 19-year-old man was referred to our hospital because of acute visual loss and progressive protrusion of the left eye (Figure 1). He was admitted for detailed ophthalmologic examination and treatment of severe proptosis and visual loss.

His medical history revealed osteosarcoma of the right distal femur diagnosed 20 months earlier for which he had received neoadjuvant chemotherapy. Six months later, he had undergone limb-sparing resection (partial resection of right distal femur) which was replaced with allograft and intramedullary nail placement. Then, he was on adjuvant chemotherapy. The postoperative follow-up was uneventful until 2 months ago when he presented with severe frontal headache. Brain and orbital computed tomography (CT) and magnetic resonance imaging (MRI) demonstrated metastasis to the frontal lobe and left orbit. He underwent partial excision of metastatic brain lesion by craniotomy.

At presentation, on ophthalmic examinations, the visual acuity was NLP (left eye) and RAPD was positive in the left eye. External examinations revealed left proptosis. Ocular movement was limited in all directions. Intraocular pressure (IOP) by applanation tonometry was 35 mmHg.

Fundus examination revealed cherry red spot appearance that was compatible with diagnosis of central retinal artery occlusion. The right eye was normal.

Orbital CT scan and MRI revealed orbital mass eroding superior, lateral, and posterior orbital walls. Also, intracranial large lesion within the temporal fossa could be seen (Figures 2 and 3).

He underwent superotemporal orbitotomy via supralateral lid crease approach for debulking of orbital mass. Histopathological examination (HPE) of the specimen was reported as metastatic osteosarcoma with extensive tumor necrosis (Figure 4). Since then, he has been on adjuvant chemotherapy and palliative radiotherapy.

## Discussion

One of the infrequent causes of adult proptosis is orbital metastasis; approximately 2.5%–13% of orbital biopsies have demonstrated a metastatic tumor, and this diagnosis is often unexpected.[9] the most common primary tumor sites for orbital metastases include breast, prostate and lung carcinoma. Osteosarcoma is a very rare primary tumor site accounting for orbital metastasis. A review of literature in MEDLINE showed that only five other cases of orbital metastatic osteosarcoma have been reported. Table 1 describes these cases including ours.[1]

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**Table 1.** Previously reported cases of osteosarcoma metastasis to the orbit.

| Case | Authors | Age/sex | Primary tumor | Orbital metastasis | Symptoms | Histopathology | management |
|---|---|---|---|---|---|---|---|
| 1 | Misra, et al.[4] | 8/M | Right tibia | right | Proptosis<br>Normal VA<br>Normal VF | N/A | Palliative chemotherapy |
| 2 | Mohadjer, et al.[5] | 26/M | Right pubis | bilateral | Proptosis<br>Decreased VA<br>VF defect<br>3rd nerve palsy | Telangectatic | Chemotherapy/<br>radiotherapy |
| 3 | Lin, et al.[6] | 15/F | Right tibia | right | Proptosis<br>Decreased VA | Osteoblastic | Chemotherapy/<br>Local irradiation/<br>Embolization/<br>Partial resection |
| 4 | Suresh, et al [8] | 16/m | Right tibia | right | Painful protrusion<br>Decreased VA | Osteoblastic | He was offered enucleation, local radiation, and chemotherapy |
| 5 | Rajabi, et al.[7] | 55/M | Left scapula | Left | Proptosis<br>Decreased VA | N/A | Partial resection<br>Palliative chemotherapy |
| 6 | Rajabi, et al.<br>(this report) | 19/M | Right femur | left | Proptosis<br>High IOP<br>Vision loss<br>CRAO (central retinal artery occlusion) | Conventional | Neoadjuvant chemotherapy/<br>Partial resection/<br>Adjuvant hemotherapy/<br>Palliative radiotherapy |

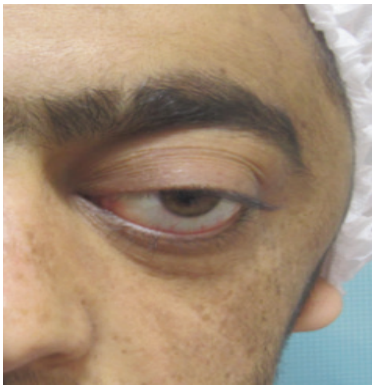

**Figure 1.** Patient's photograph showing left eye proptosis and inferolateral displacement of the globe.

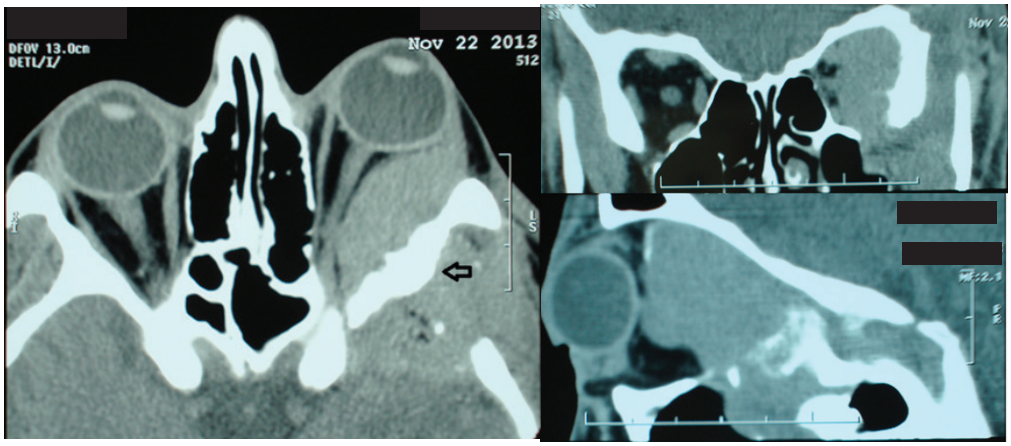


**Figure 2.** Orbital CT scan: Showing large homogeneous intraorbital mass pushing the globe anteriorly. Destruction of the temporal bone and erosion of greater wing of sphenoid (arrow) and large intracranial mass are visible.

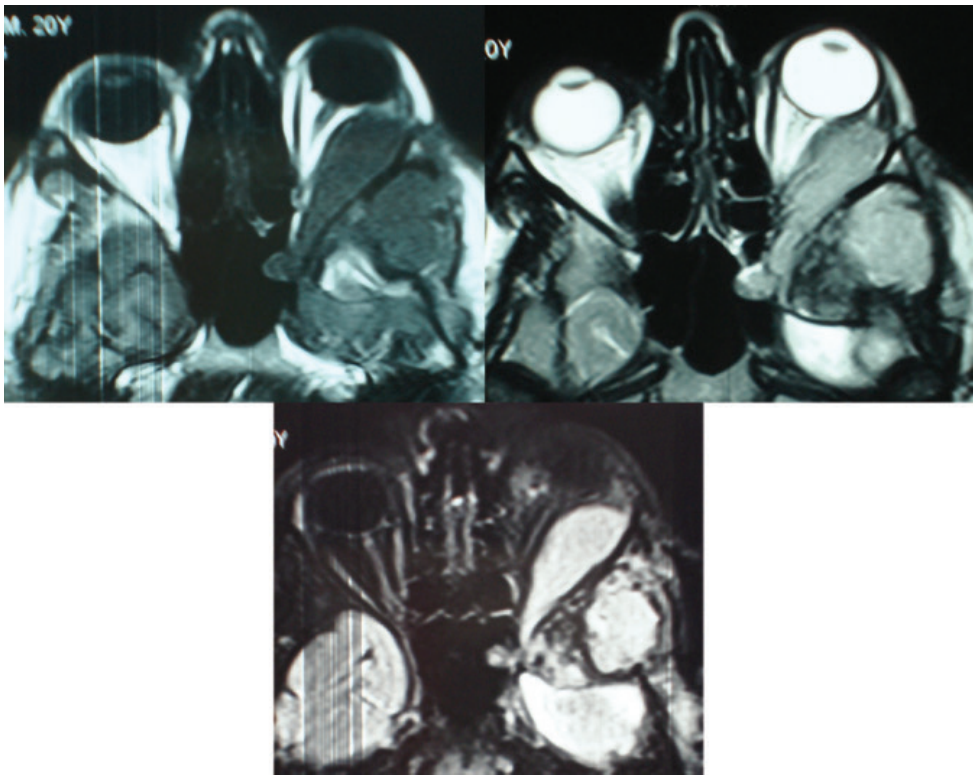

**Figure 3.** Orbital MRI: T1-weighted axial MRI showing the lesion with hypo signal intensity (left upper). T2-weighted axial MRI shows the lesion with hyper intensity (Right upper).T1-weighted with contrast and fat suppression shows contrast enhancement of the mass within the orbit and brain (lower).

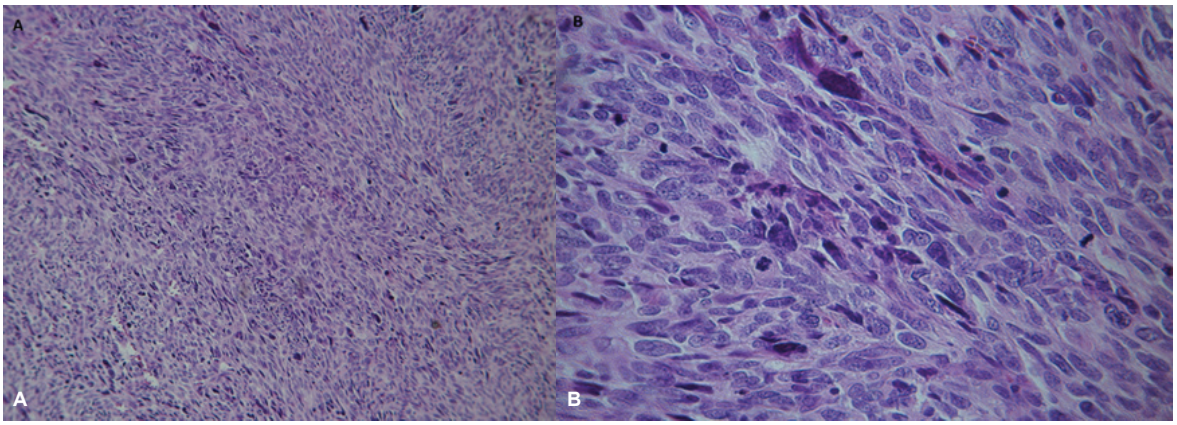

**Figure 4.** Hematoxylin and eosin stained section of tissue from orbit at ×100 magnification shows a high-grade spindle cell sarcoma with nuclear pleomorphism **(A)**; At higher magnification (×400), high-grade spindle-cell sarcoma, numerous mitoses are seen. The tumor shows no specific pattern of differentiation **(B)**.

It is more common for osteosarcoma to appear in long tubular bone, although it is possible to arise in a flat bone in rare circumstances (about 20 times less frequent)[1] Amongst these six cases, two arose in flat bones (pubis and scapula) and four in long bones. Three metastases were derived from tibia and our case is the first case which is of femoral bone origin.

Typical patients with central osteosarcomas are often a decade or younger than patients with surface osteosarcoma.[1] Our case, similar to others with osteosarcoma of long bone origin, is in a younger age group compared to patients with osteosarcoma of flat bone origin.

Proptosis is the most common sign of orbital metastasis and all these six cases had painful proptosis. One case had normal Visual Acuity and Visual Field while five others had decreased vision. Our patient is the only case who presented with Central retinal artery occlusion and High IOP.

In all five previous cases, osteosarcoma metastasized to the ipsilateral orbit (one had bilateral orbital involvement). Our patient is the first one whose osteosarcoma metastasized to the contralateral orbit (from the right femur to the left orbit).

It seems that the orbital metastases appear late in the course of osteosarcoma and their prognosis is generally poor. According to our literature review, three cases including ours had a long duration (>20 month) from the presentation of the primary tumor to

the occurrence of orbital metastasis. In one case with duration of more than 36 month, the patient died of his disease.

All patients received palliative therapy and three including ours underwent partial resection of the metastatic lesion. Regarding the long course of the disease, palliative chemoradiotherapy and partial resection would be a good choice in its management.

Although orbital metastasis of osteosarcoma is a rare event, it seems it has had an increasing trend recently; therefore, ophthalmic manifestations should always be kept in mind during the follow up period. Also, we should make efforts to palliate the patient's symptoms as much as possible. These patients need multidisciplinary management; so, organized teamwork and proper interaction among ophthalmologists, orthopedic surgeons and oncologists is necessary.

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