INTRODUCTION

Metastatic orbital lesion due to osteosarcoma is a rare condition and manifestations include orbital mass, pain, diplopia, chemosis and ocular motility disturbance depending on the location of the mass and its effect on adjacent structures. There is no effective treatment, but the combination of treatment modalities including chemotherapy, radiotherapy and surgery for tumor debulking may delay disease progression. Herein, we report a case of orbital metastasis from scapular bone osteosarcoma.

CASE REPORT

A 55-year-old man was referred to the Oculoplastics Clinic at Farabi Eye Hospital, Tehran, Iran with acute onset painful decrease of vision and protrusion of his left eye. He was a known case of left scapular bone osteosarcoma and had undergone surgical excision of the original tumor and chemotherapy, approximately 4 months before. Visual acuity was counting fingers and 20/20 in the left and right eyes, respectively. Ophthalmic examination revealed proptosis, severe lid swelling, chemosis and conjunctival injection in his left eye [Figure 1]. There was profound gaze limitation in all directions in the left eye. Dilated fundus examination was normal in both eyes.

In the next step, orbital CT-scan revealed a large heterogeneous mass lesion in the left intraconal orbital space with tiny foci of hyperintensity [Figure 2]. For further investigation, MRI was done in which an intraconal lesion, hypointense in some planes and hyperintense in others, was noted on T1 images [Figure 3a]. The intensity of the lesion was heterogeneous to hyperintense on T2 images [Figure 3b and c] which also displayed...
Orbital Metastatic Osteosarcoma; Rajabi et al

ring enhancement on gadolinium enhanced T1 images [Figure 3d].

The patient underwent incisional biopsy through a lateral orbitotomy. Histopathological examination of the tumor revealed malignant spindle cells with osteoid formation and mineralization which was compatible with metastatic osteosarcoma [Figure 4].

The orbital metastasis being confirmed, the patient underwent systemic evaluation for any other site of metastasis; however, there was no other organ involvement. The patient was referred to his oncologist for palliative chemotherapy. Unfortunately, after 2 months of chemotherapy, he deceased in the clinical setting of sepsis probably due to systemic immunosuppression.

DISCUSSION

Osteosarcoma is the most common malignant bone tumor and may involve the long bones of young adults primarily.[1] It can be seen in association with hereditary retinoblastoma, previous exposure to ionizing radiation or Paget disease.[2] The most common site of involvement is the medulla of the metaphysis in long bones, particularly the distal femur, proximal tibia and proximal humerus.[3] It has been classified conventionally into osteoblastic (50%), chondroblastic (25%) and fibroblastic (<25%) variants; a small percentage of them are categorized into telangiectatic, small cell multifocal, parosteal, periosteal, and extra-osseous variants.[2]

These tumors are very invasive and metastasize in approximately 20% of patients.[4] Factors affecting prognosis include tumor histopathology, size, raised pre- and post-operative serum alkaline phosphatase levels, involvement of lymph nodes and treatment modality. However, age, gender, history of trauma or bone fracture and the site of primary osteosarcoma do not influence prognosis.[5] The most common sites for metastases for tumors originating from long bones is the lung and the second one are bones.[6]

By advances in systemic chemotherapy regimens, the long-term survival of patients without metastasis has now improved to 70%; however, in patients with metastasis or disease recurrence, survival is <20%.[4,7]

Metastatic involvement of the orbit is a rare condition; orbital metastasis can originate from any primary tumor in the body.[8] The most common solid tumors in adults with orbital metastasis are breast and lung tumors, followed by cancers of the genitourinary tract and gastrointestinal system.[9] Some studies showed that leukemia and neuroblastoma are the most frequent causes of orbital metastasis.[10] Ewing’s sarcoma and

Figure 1. Appearance of the patient at presentation revealed proptosis, severe lid swelling and chemosis of the left eye.

Figure 2. Orbital CT scan shows a heterogeneous intraconal mass lesion with small foci of hyperdensity. Axial view (a) Coronal view (b).

Figure 3. Orbital MRI showed an intraconal mass that was heterogeneous to hyperintense on T1 images (a) and heterogeneous to hyperintense on T2 images (b and c) with gadolinium enhancement at the rim of the lesion on T1 images (d). In some sections, the mass seems to be hyperintense on T1 images (a) and in some others, hypointense (d). The same finding can be seen on T2 images (b and c).

Figure 4. Histopathological studies revealed malignant spindle cells with osteoid formation and mineralization compatible with metastatic osteosarcoma.
Wilms tumor are less frequently the source of orbital metastasis.\(^9\)

The most common symptoms of metastatic orbital osteosarcoma are pain, diplopia and ocular dysmotility. Less common clinical manifestations include chemosis, epiphora and loss of vision.\(^3\) Misra et al\(^{11}\) reported the first case of osteosarcoma metastasizing to the orbit. The patient was an 8-year-old boy with osteosarcoma in his right tibia with involvement of the right eye 1 month after the diagnosis. Attili et al\(^{12}\) reported another case with right tibial osteosarcoma, painful protrusion of the right eyeball and metastatic orbital osteosarcoma.

It is interesting that in all previously reported cases of metastatic orbital osteosarcoma, the primary and metastatic sites were both on the right side,\(^{2,11,13}\) but in the present case, both of them originated from the left side. In previous reports, the primary site of osteosarcoma was the right tibia and in our case, it was the left scapula which may explain left orbit preference. As the current case is only the fifth subject with metastatic orbital osteosarcoma, this finding can be purely coincidental. However, it is notable that in all cases with metastatic orbital osteosarcoma, the lung was not involved by metastasis. It is believed that spread of tumor cells to the orbit may occur through Botson’s paravertebral venous plexus which has already been hypothesized in other studies.\(^{13}\)

Orbital MRI features were also challenging in our case. Most metastatic osteosarcomas show hyperintense signal on T1 scans, whereas signal intensity was heterogeneous to high in our case. We assume that various amounts of necrosis, hemorrhage and mineralization in osteoid tissue may have resulted in this particular presentation on T1-weighted images. Typical ring enhancement was seen on gadolinium enhanced images and a heterogeneous mass was revealed on T2 sections.

There are no specific guidelines for management of orbital metastases. Combination therapy using different treatment modalities including radiotherapy, chemotherapy and surgery may be effective for palliative measures, maintenance of vision and regression of metastatic tumor.

REFERENCES


Source of Support: Nil. Conflict of Interest: None declared.