

Ewing Sarcoma of Orbit with Intracranial Extension

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Ewing sarcoma causing unilateral proptosis in a child is unusual presentation. A young girl presented with painless proptosis of left eye with restricted eye movements. Her radiology revealed a large soft tissue mass causing destruction of lateral orbital wall and zygomatic arch measuring 2.0 × 3.0 cm. Total excision of intracranial and intra orbital part of the tumor brought about substantial relief. The clinical and radiological presentation and management of this entity are discussed.

Key words: Sphenoid bone, Ewing sarcoma, orbital proptosis, primitive neuroepithelial tumor (PNET), radiology, small round cell tumor, surgery.

Ewing's sarcoma is a malignant small round-cell tumor classically involving the long bones of the limbs, the ribs, or the pelvis.¹ Primary Ewing's sarcoma of the head and neck region is unusual and generally involves the mandible or Maxilla.¹ The mean age of occurrence is between the first and the second decades in 80% of cases.² The extra osseous Ewing sarcoma and PNETs share a unique and consistent genetic translocation t(11;22)(q24;q12).³ We report a young girl who presented with features of proptosis in left eye without visual deterioration. Her radiology revealed intraorbital tumor with intracranial extension involving the greater wing of sphenoid bone. Total excision of intraorbital and intracranial part of tumor brought about relief of proptosis.

CASE REPORT

A 13 year old girl was presented with features of painful, progressively increasing proptosis of left eye for the last 6 months in 2010. There was no associated loss of vision, headache and vomiting. There was no history of trauma. Family history was unremarkable. Examination revealed a left sided proptosis downwards and laterally measuring 23 mm on Hertel's exophthalmometer, with restriction of extra

ocular movements of all muscles. The dystopia was measured to be 5 mm laterally and 5 mm downwards (Fig. 1). Her visual acuity was 6/6 in both eyes. Her fundoscopy was normal. She underwent full systemic evaluation with only two abnormal results: Markedly raised Leucocyte count and MCV, MCHC, PCV slightly deranged.

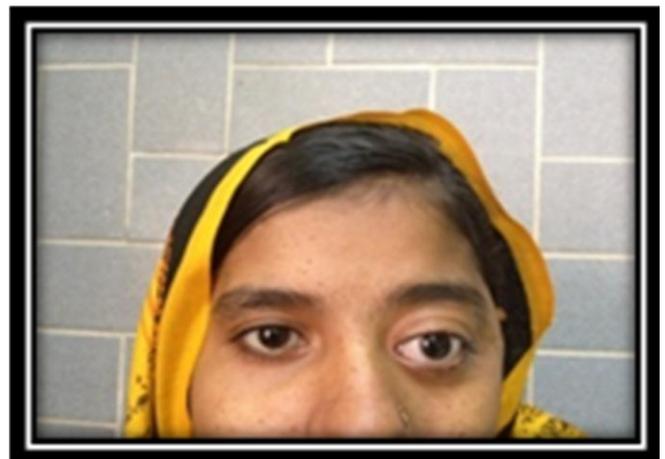


Fig. 1: Proptosis of Left Eye.

Her MRI Scan revealed abnormal high intensity signal within greater wing of sphenoid with

destructive permeative lesion. A heterogeneous soft tissue mass was seen involving zygoma and greater wing of sphenoid, measuring 3.0 x 3.0 cm (Fig. 2).



Fig. 2: MRI Axial view taken in 2010.

When she came to us in 2010 we proved Ewing sarcoma after an incisional biopsy. She was referred to oncology for consultation and was given 6 months course of VAC regime (vincristine, actinomycin, and cyclophosphamide). At a follow-up of 3 months her proptosis had substantially subsided and extraocular movements had recovered.

We repeated MRI of brain in 2011, which showed reduction in size of mass as comparison with previous one.

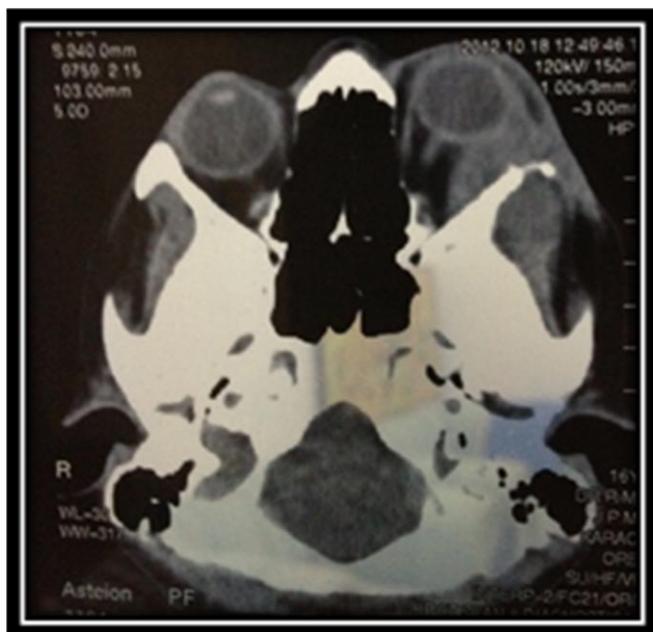


Fig. 3: CT scan axial view at the level of orbit.

In 2012, she again came to us with same complaints but with more aggressive in nature. Her MRI scan was repeated which showed cortical irregularity and thinning remodeling of greater wing of sphenoid associated with adjacent soft tissue swelling of lateral rectus and temporalis muscle (Fig. 3).

With the help of maxilo-facial department we performed panoramic orbitotomy on her. During surgery tumor was found to be firm, partly suckable and slightly vascularized. Complete intraorbital and intracranial mass was excised (Fig. 4), which was reddish - brown in color (Fig. 5). Part of greater wing of the sphenoid bone was also removed (Fig. 6).



Fig. 4: Tumor is being removed.



Fig. 5: Biopsy Specimen.



Fig. 6: Some of the bone also removed.

There was no damage done to vision or muscle. Vision remained 6/6 in both eyes with no cosmetic disfigurement (Fig. 7). On follow up 3 weeks after surgery proptosis decreased to 20 mm and Postoperatively her MRI was repeated which showed no evidence of mass (Fig. 8).



Fig. 7: Post-op picture after 2nd surgery.

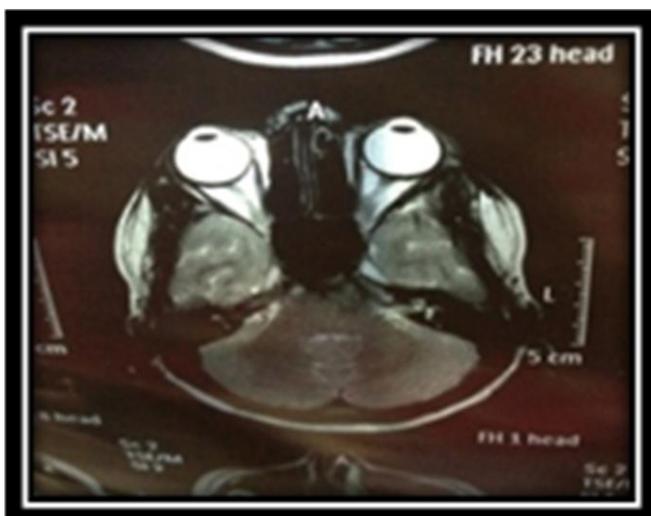


Fig. 8: Post-operative: MRI axial view shows no growth.

DISCUSSION

Ewing sarcoma is a malignant tumor that was first described as an endothelioma of the bone by James Ewing in 1921.⁴ Separate cases of involvement of either the cranial cavity or orbit have been reported, but a combination of the two has been aE rarity. Ewing

sarcoma is a highly malignant, small round cell tumor that primarily involves the pelvis and long bones. It accounts for 10% of all bony tumors and 4% of tumors in the head and neck region, typically involving the skull, mandible, and maxilla. However the Orbital involvement is rare.⁵ Metastases in the orbit from distant primary sites presenting as proptosis is rare, and unilateral, usually situated on the same side as the primary tumor. Primary orbital Ewing sarcoma/PNET are extremely rare with only 17 reported cases as per literature.⁶

It is composed of sheets of small cells with high nuclear to cytoplasmic ratio. The cytoplasm is scanty, eosinophilic, and usually contains glycogen, which is detected by periodic acid Schiff stain and is diastase degradable. The nuclei are round, with finely dispersed chromatin, and one or more tiny nucleoli.⁷ The histopathological examination revealed a characteristic round cell malignancy with a highly cellular tumor arranged in sheets with formation of nodules. Together with this Increased mitotic activity was identified. On immunohistochemistry, there was positivity for CD99 and neuron - specific Enolase (NSE). The lesion was negative for synaptophysin and leukocyte common antigen (LCA). Spread of this tumor into the orbits is most likely through blood. Metastases to orbits are extremely rare in Ewing's sarcoma.⁸

Computed tomographic scanning show mottled destruction of bone but typically no soft tissue enhancement with contrast. The characteristic periosteal "onion ring" reaction seen in long bones is not usually present in orbital cases.⁵

Although Ewing sarcoma was previously a tumor with high mortality, but with combined treatment of chemotherapy and surgery the prognosis has been improved greatly. In 2011, a case of ewing's sarcoma of orbit with intracranial extension has been reported. They gave treatment with combined regimen of surgery with chemo-radiotherapy. Their patient responded well with this therapy.⁹ Local treatment relying on surgical excision and radiotherapy alone has proven inadequate, with 5 - year survival rates of < 10%. The addition of chemotherapy has improved survival rates significantly to approximately 50%.¹⁰ Treatment of Ewing's sarcoma with a combination of surgery, chemotherapy and radiation therapy results in a 5 - year survival rate of approximately 65%. According to Esiashvili and colleagues, the 5-year survival of localized disease increased from 44 to 68% in the period after 1993, whereas 5 - year survival of

metastatic disease increased from 16 to 39%. The corresponding 10 – year survival increased from 39 to 63% for localized disease and from 16 to 32% for metastatic Ewing's sarcoma.¹¹

CONCLUSION

Ewing sarcoma presenting as proptosis with intracranial extension is rare manifestation in pediatric age group. Primary Ewing sarcoma of the orbit should be considered in the differential diagnosis of children or young adults with proptosis, diplopia, and periorbital swelling. Immunohistochemistry is essential to distinguish Ewing sarcoma from other small round cell tumors. However if diagnosed early and with appropriate management complete cure is likely.

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