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# A rare case of primary orbital Ewings sarcoma with intracranial extension

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**Keywords**
non-axial proptosis, small round blue cell tumours, skeletal metastases, extraosseous Ewing's sarcoma, orbital decompression surgery

## ABSTRACT

Primary Ewings sarcomas of orbit with intracranial extension are extremely rare with few cases reported to date. They form an important differential diagnosis in aggressive orbital tumours presenting in young males. We presented the case report of a young male presenting with right eye proptosis. The patient underwent complete tumour excision. Biopsy was suggestive of Ewings sarcoma. Metastatic workup showed extensive lesions in lungs and bones. Six months post-operative follow-up with radiation and chemotherapy showed disease remission. CD99 is a key IHC marker in differentiating from other similar tumours. Total tumour excision with adjuvant radiation and chemotherapy may be helpful in survivability in such patients but overall prognosis still remains poor.

## INTRODUCTION

Ewings sarcoma is a malignant tumour of bone with mean age of presentation of 10-14 years in male and 5- 9 years in female [1]. Ewings sarcoma is commonly seen in lower extremities in 35% , chest (21%), pelvis (17%), spine (10%), upper extremity (9%), head and neck (6%) and abdomen in 2% cases [2]. Rare case reports of Primary Ewings sarcoma of orbit are present [3]. Genetically Ewings sarcoma is due to cytogenetic translocation , t(11;22) (q24;q12) [4].

## CASE PRESENTATION

Here we present the case report of 29 year old male who presented with history of proptosis of right eye for last 1 year. There is also associated history of double vision of lateral and medial gaze. On examination his right eye was non axially proptosed medially and downward. There was restriction of movement in eye in lateral, medial and upward gaze. His visual acquity of Right eye was 6/12 and left eye was 6/6. However there was no cranial nerve palsy on examination. His vitals were stable and no abnormality detected on rest of his systemic examination. Opthalmology consultation was taken and fundus examination was normal.

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**Figure 1.** Severe downward proptosis right eye.

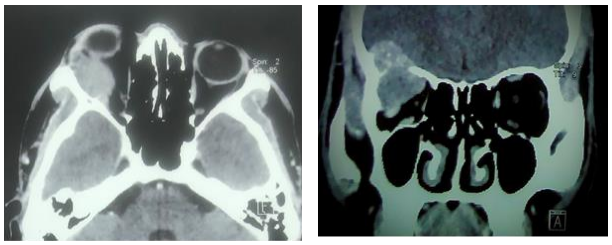


**Figure 2.** Contrast MRI showing enhancing Right orbital lesion with intracranial extension.

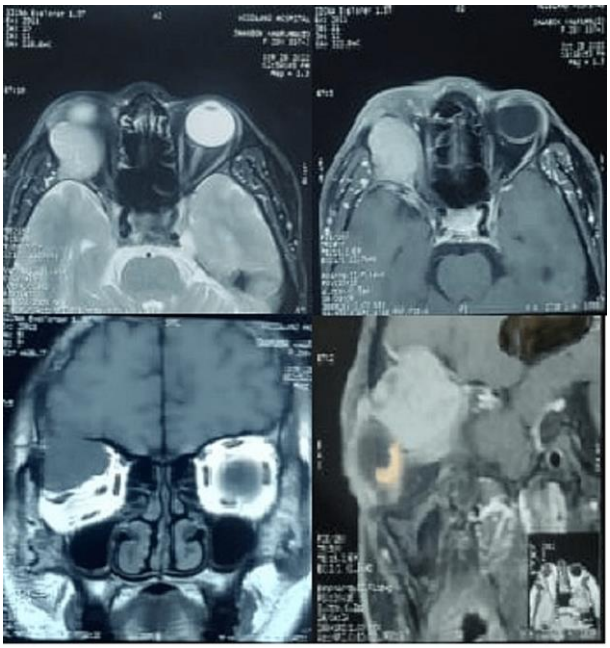

**Figure 3.** CT scan brain showing the Right frontal bone erosion and intracranial extension.

We requested urgent magnetic resonance imaging of his brain and orbit which was suggestive of T1 hyperintesnse and T2 isointense homogenous lesion in the superolateral extraconal compartment of right orbit measuring 3.4X 2.6X 3.47 in dimensions. There was extension of the lesion through roof of orbit to Anterior cranial fossa. Based on MR findings a provisional diagnosis of high grade lacrimal tumour was made.

**Figure 4.** Showing complete resolution of proptosis and opthalmoparesis postoperatively.

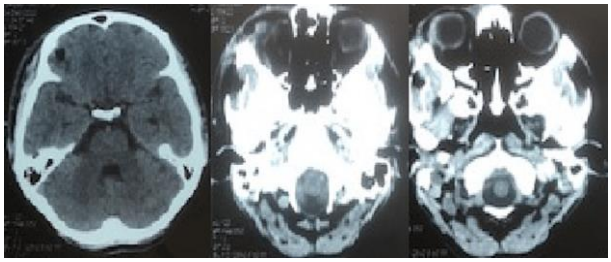

**Figure 5.** Post operative CT scan showing complete excision of tumour along with intracranial extension.

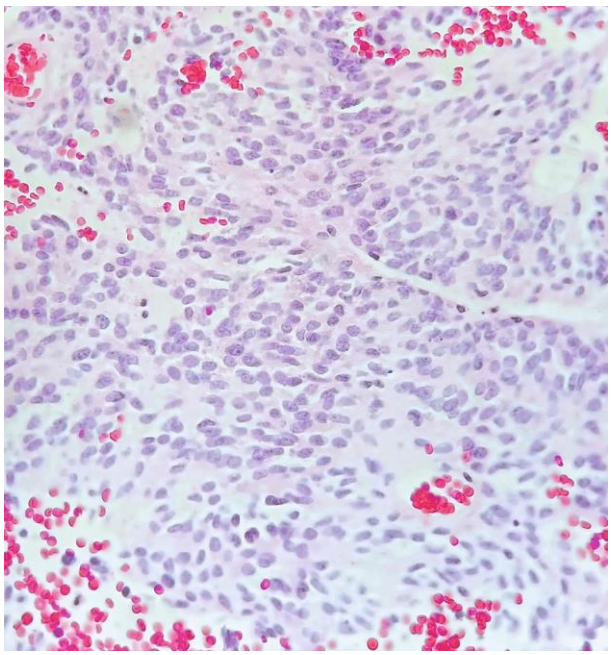

**Figure 6.** Histiopathology showing small round cells arranged in sheets (H&E stain 400X).

Rest of his blood investigations were unremarkable. He underwent Right lateral orbitotomy and tumour excision. Intraoperatively tumour was soft, vascular, pinkish in colour and suckable. Erosion and extracranial extension was seen in right orbital roof and gross total excision of tumour was done. Post operatively patient recovered well. His vision in right eye improved to 6/6 and opthalmoparesis resolved completely.

Tumour histiopathology report was suggestive poorly differentiated small round cell tumour. IHC showed strong positivity towards CD 99 and a final diagnosis of Ewing sarcoma was made.

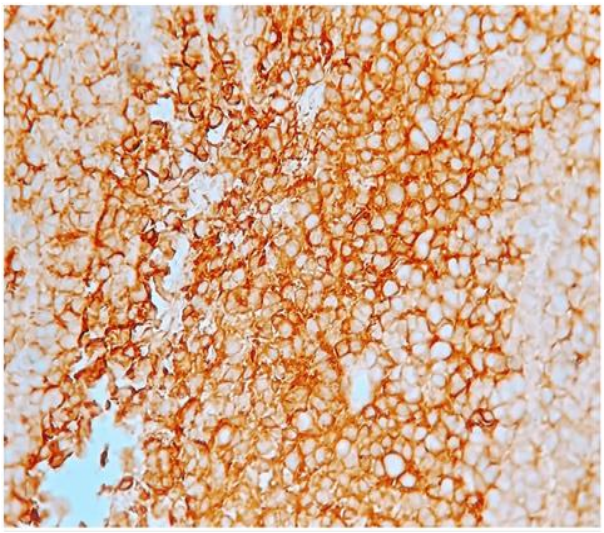

**Figure 7.** Tumour cells showing strong membranous positivity for CD99 (IHC 400X).

Patient underwent complete metatstatic workup and multiple lesions were detected in bilateral lung and lytic lesions in multiple ribs and pelvis. Patient underwent radiotherapy followed by chemotherapy with vincristine , adriamycin and cyclophosphamide alternating with etoposide and ifosamide . Patient has been on followup for last 6 months and is doing well.

## Discussion

Primary orbital Ewing with intracranial extension is extremely rare and only few case reports are present [5, 6]. Orbital Ewings sarcoma most commonly occurs in young male similar to our case [7]. In our case tumour was located in superolateral quadrant of orbit which confers with the most common location in other reported cases [8, 9]. Patient usually presents with proptosis and the median duration of presentation is 9 weeks [7], however our patient presented to us at an advanced stage of disease with distant metastasis. Metastasis from primary orbital Ewings is seen in 20-25% cases [1]. Our patient underwent surgical excision followed by radiotherapy and chemotherapy according to current treatment strategy for Ewings Sarcoma [1]. CD99 is the key IHC marker which is invariably positive in Ewings sarcoma and in our case helped to differentiate it from Neuroblastoma [10]. Orbital Ewings is considered to be less aggressive than Ewings at other location [10] and patient usually have survival more than 6 months in most of the cases. Similar findings were seen in our case where our patient is doing well on his 6 month follow up.

## Conclusions

Primary Ewings Sarcoma with intracranial extension is extremely rare condition with only few case reports available in literature. One should consider Primary Ewings sarcoma of orbit as differential diagnosis for orbital tumours in young male patients especially with intracranial extension and distant metastasis. Despite multimodality treatment involving surgical excision and adjuvant chemo-radiotherapy these tumours are rapidly progressive and invariably fatal.

## References


1. Carvajal R, Meyers P: Ewing's sarcoma and primitive neuroectodermal family of tumors . Hematol Oncol Clin North Am. 2005, 19(3):501-525. 10.1016/j.hoc.2005.03.004.
2. Le Deley MC, Paulussen M, Lewis I, Brennan B, Ranft A, Whelan J .: Cyclophosphamide compared with ifosfamide in consolidation treatment of standard-risk Ewing sarcoma: results of the randomized noninferiority Euro-EWING99-R1 trial.. J Clin Oncol . 2014, 32(23):2440-2448. 10.1200/JCO.2013.54.4833. Epub 2014 Jun 30
3. Woodruff G, Thorner P, Skarf B: Primary Ewing's sarcoma of the orbit presenting with visual loss . Br J Ophthalmol. 1988, 72:786-792. 10.1136/bjo.72.10.786.
4. Ewing J : Classics in oncology. Diffuse endothelioma of bone. James Ewing. Proceedings of the New York PathologicalSociety,1921.CACancerJClin.1972,Mar-Apr;22(2):95-98. 10.3322/canjclin.22.2.95.
5. Abbasi S, Cheema A: Ewing sarcoma of orbit with intracranial extension . Pak J Ophthalmol. 2015, 31:111-14.
6. Naqvi SH, Hameed S, Naqvi SH, Musani MA: Primary Ewing's sarcoma of the orbit with intracranial
7. extension abutting the temporal lobe: a rare case report. Int Ophthalmol. 2014, 34:1137-1140. 10.1007/s10792-014-9937-5
8. Kaliki S, Rathi SG , Palkonda VAR: Primary orbital Ewing sarcoma family of tumors: a study of 12 cases . Eye. 2018, 32:615-621. 10.1038/eye.2017.278 .
9. Harbert F, Tabor GL Jr : Ewing's tumor of the orbit, report of two cases . Am J Ophthalmol. 1950, Aug 33(8):1219-1225. 10.1016/0002-9394(50)90991-3.

10. Yamada S, Takahashi H: A dissected case of Ewing's sarcoma (reticulum cell sarcoma) of the orbit . Hirosaki Med J. 1957, 8:202-207.
11. Alfaar AS, Zamzam M, Abdalla B, Magdi R, El-Kinaai N: Childhood Ewing sarcoma of the orbit . J Pediatr Hematol Oncol. 2015, 37(6):433-437. 10.1097/MPH.0000000000000346.