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# A Rare Presentation of Trauma Induced Rhabdomyosarcoma

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#### Abstract

**Introduction:** Orbital rhabdomyosarcoma (RMS) is a highly malignant tumor that originates mainly from mesenchymal tissue. It is considered a rare and highly aggressive malignancy that should be considered in any orbital which rapidly progresses. Delay in diagnosis and inappropriate treatment negatively affects the outcome.

**Case Discussion:** 5yr/F presented with LE mass since 4 months associated with globe distortion. h/o trauma 2 years back followed by cataract extraction and secondary IOL implantation with trabeculectomy. 6 months later h/o fall leading to IOL explant, hyphema and vitreous hemorrhage. On examination cavity was filled with mass. CT Scan and MRI Orbit suggestive of Retinoblastoma. Histopathology report following enucleation suggestive of Rhabdomyosarcoma Grade III.

**Conclusion:** Although rare but it has been postulated that RMS may occur following multiple trauma and since it is one of the few life-threatening diseases that present first to an ophthalmologist; therefore prompt diagnosis and treatment is a life-saving issue.

#### Introduction

Rhabdomyosarcoma (RMS) is a highly malignant, most common soft-tissue sarcoma of the head and neck in childhood with 10% of all cases occurring in the orbit. Two- third of them occur in superonasal quadrant with 1st decade being the common age of presentation.

#### **Case Report**

- 5 yr/female child.
- c/o Left eye mass since 2 yrs, progressive in nature.
- associated with
  - Diminution of vision
  - Conjunctival congestion
  - Distortion of globe

 purulent discharge, pus and bleeding

#### **Past History**

- H/o Trauma to LE due to fall, which was followed by development of traumatic cataract 1 week later
- Multiple Surgical history
- 1. LE cataract extraction (phacoemulsification )
- 2. LE trabeculectomy with anterior vitrectomy after 2-3 months of cataract extraction
- 3. LE sec IOL implantation.
- H/O fall 6 month after IOL implantation followed by development of hyphaema

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and vitreous haemorrhage. IOL was explanted.

### **Clinical Features**

#### **General Examination**

Cervical lymphadenopathy ipsilateral side (left)- 2 palpable lymph nodes, firm, discrete, approximately 3 cm in widest diameter.

#### **Ocular Examination**

Left eye- Distorted irregular globe with multinodular mass visible temporally, with swollen, hyperemic and retracted lid due to mass effect.

perception of light was negative.

Rest details not appreciable.

RE- anterior and posterior segment -WNL

#### Investigations

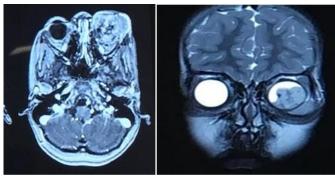
- Routine blood investigation- Hb= 9 g/dl, rest WNL.
- Blood sugar = 95.2 mg/dl
- Coagulation profile= WNL
- Imaging studies- Orbital CT scan
- Irregular hypodense area within left vitreous cavity –AC region with internal calcification area and a soft tissue extending outside left eyeball represent post operative calcification is more likely.
- Atrophy of left optic nerve

#### FNAC- Cervical Lymph Node



27<sup>th</sup> April 2017, FNAC left cervical lymph node shows malignant round cell tumor

MRI



Left sided lesion involving the anterior and posterior chambers of globe with extra-orbital extension. extension seen as proptosis on left side. The bilateral optic nerve head appears to be uninvolved

Features are suggestive of neoplastic etiology more likely to be retinoblastoma than rhabdomyosarcoma.

#### Treatment

**Chemotherapy-** 3 cycles of Etoposide + Cisplatin following which there was visible reduction in tumor size

• There were no palpable LN after chemotherapy



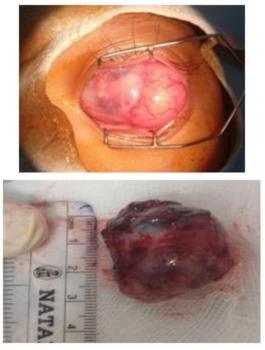
Pre - chemotherapy



Post – chemotherapy

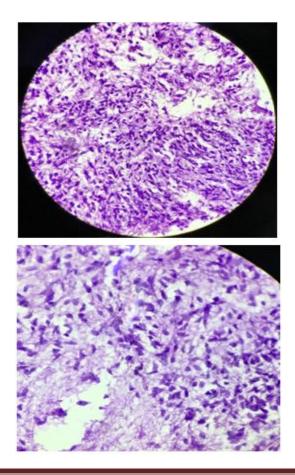
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**Surgical intervention:** Left eye was enucleated and sent for histopathology



### Histopathology

- Exophytic rhabdomyosarcoma grade III, with involvement of intraocular structure
- No involvement of optic nerve



### Discussion

Orbital Rhabdomyosarcoma usually presents as a space-occupying lesion in the orbit during the first decade and may mimic other neoplastic or inflammatory masses. The cause of rhabdomyosarcoma is unclear. Several genetic and environmental factors syndromes are associated with it. Association with the trauma is very rare.

Although imaging and clinical signs and symptoms are helpful in delineating the differential diagnosis, the decisive diagnosis is based on histopathologic confirmation. Fineneedle aspiration biopsy can provide insufficient or misleading information. Histopathology is the gold standard to make the final diagnosis.

## Conclusion

The presence of internal calcifications in MRI was suggestive of Retinoblastoma but histopathology report confirms diagnosis as Rhabdomyosarcoma. Although rare it has been postulated that rhabdomyosarcoma may occur following multiple trauma and since it is one of the few lifethreatening diseases that present first to an ophthalmologist; therefore prompt diagnosis and treatment is a life-saving issue

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