

UNILATERAL PROPTOSIS IN A CHILD- A DIAGNOSTIC DILEMMA

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PRESENTATION OF CASE

Proptosis is the abnormal forward protrusion of the eyeball. Proptosis though an uncommon presenting complain in childhood needs to be thoroughly investigated as many conditions may mimic this condition, keeping in mind a categorized differential to expedite early treatment. Management is a team approach with contribution of the paediatrician, haemato-oncologist, neurosurgeon and pathologist. We present one such case of acute proptosis in a child which clinically looked different from the histopathological diagnosis.

A two year old girl presented with acute onset proptosis since one week after a bout of cold and fever. There was no history of redness, pain or discharge. No significant antenatal, birth and post-natal history. No significant family history. There was no history of trauma or any other surgeries.

On examination, the child has right 30° esotropia. The right globe had eccentric proptosis of 12 mm, displaced downwards and medially with extraocular movements restricted in all directions. A globular mass was felt in the inferolateral aspect of the right eye which was soft, non-pulsatile, non-reducible and non-tender around 1x1 cm (Fig 1). Borders were ill-defined and fingers could be insinuated between the mass and orbital margins. No bruit was heard over the swelling. Anterior segment and fundus was normal in both the eyes. Systemic examination, blood investigations and blood culture were normal.



Figure 1. Right Eccentric Proptosis

CLINICAL DIAGNOSIS of Lymphangioma was made- B-scan revealed multilobulated hypoechoic collection around 3.5 cm x 3 cm in the retro orbital region.

CT scan showed no bony erosions or calcifications.

MRI reported a well encapsulated cystic lesion in the intraconal space of retro orbital region. Lesion is hyperintense on T2W image and isotense on T1W image and encasing the optic nerve, the globe was displaced anteriorly and pressed posteriorly. (Figure 2). Features suggestive of lymphangioma.



Figure 2. Hyperintense Lesion on T2W Image and Isotense on T1W Image

Common Causes of Childhood Proptosis¹

Congenital	Exorbitism (craniosynostosis, skull anomalies) Meningocele / Encephalocele Dermoid cyst
Traumatic	Orbital hematoma Traumatic haemorrhage in existing neoplasm
Inflammatory	Orbital cellulitis, abscess Pseudotumour Mucocele

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Neoplastic	Ophthalmic / Orbit	Haemangioma, Optic nerve glioma Rhabdomyosarcoma Orbital retinoblastoma Teratoma
	Non - ophthalmic	Granulocytic sarcoma Metastatic neuroblastoma Lymphoma, Lymphosarcoma Histiocytosis X
Table 1		

DISCUSSION OF MANAGEMENT

Patient was started on Tab. Prednisolone 5 mg BD under antibiotic cover. Anterior transcutaneous orbitotomy was done as the proptosis remained same despite medical line of management. Intra operatively a purplish cystic mass was seen with thin wall resembling a chocolate cyst. (Figure 3 and 4) The mass was excised and sent for histopathology examination.

Patient was followed up after 1 week and was uneventful, except for mild restriction of abduction of right eye.

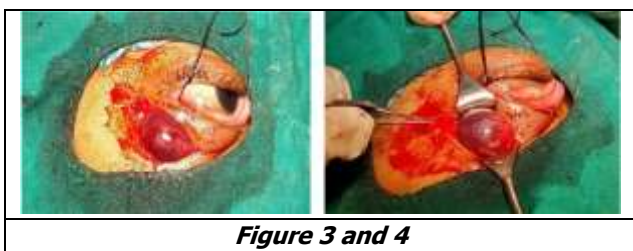


Figure 3 and 4

PATHOLOGICAL DISCUSSION

Histopathology report showed organized tissue comprising of thick walled vascular channels, admixed with proliferating congested capillaries, lobules of mature fat and fibrocollagenous tissue. The venous channel shows dilatation. There was absence of lymphoid tissue. (Figure 5 and 6).

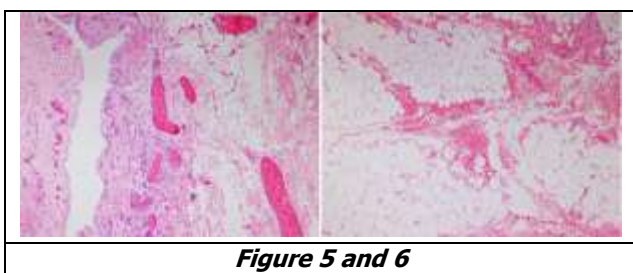


Figure 5 and 6

Histological Differential Diagnosis of Vascular Growths²

Hamartomas

- Capillary Haemangioma
- Cavernous Haemangioma
- Venous Haemangioma

Tumours

- Haemangiopericytoma
- Haemangioendothelioma

Discussion

Proptosis in a child can often present as a diagnostic dilemma. Many conditions may mimic a case of proptosis in a child. Younger the age, more acute is the onset and more rapid is the progression. Some of the common conditions are orbital cellulitis, dermoid cyst and haemangioma. However, primary malignancies like rhabdomyosarcoma as well as proptosis secondary to AML and neuroblastoma with grave outcomes should be kept in mind during evaluation. Diagnosis of such cases may require a combination of a good clinical examination and relevant investigations for an early diagnosis and appropriate management as the treatment for each cause is essentially not the same. Investigations that can be done include imaging studies (CT Scan, MRI, USG), pathology (aspiration cytology and biopsy) and haematological (hemogram, liver and kidney functions).¹ Few of the differential diagnosis in our case were orbital cellulitis, lymphangioma, metastatic neuroblastoma, rhabdomyosarcoma, lymphoproliferative diseases, capillary haemangioma and intra orbital retinoblastoma.

Vascular malformations in a child are unusual and difficult to manage. These anomalies are considered to result from errors of vascular morphogenesis. They can be categorized by their principal channel type: Capillary, venous, arterial, lymphatic or a combination.³ Vascular anomalies usually take the form of combined lesion and the classification has tended to be somewhat controversial. Those with venous connection have been differentiated as varices and those without as lymphangiomas. Comprising a spectrum of vascular anomalies that blur the distinction between lymphangioma and vascular anomaly, they used the term of a vascular hamartoma. A vascular hamartoma reproduces the whole structure of the vessel wall, and a true vascular neoplasm arises from the individual tissue of the wall of the blood vessel.⁴ Hogen and Zimmerman classified the orbital angioma as benign and malignant. Benign group includes capillary haemangioma, cavernous haemangioma, hemangiopericytoma and haemangioendothelioma. The malignant group includes malignant hemangiopericytoma, malignant haemangioendothelioma and Kaposi's sclerosing angiosarcoma.⁵

In Capillary haemangioma treatment consists of observation since many lesions involute, although few completely disappear. Treatment is indicated principally for amblyopia. If the lesion is large local radiotherapy or corticosteroids may be indicated. In a small, well defined lesion surgery may be attempted. Propranolol has shown to cause significant involution of tumours in the proliferative phase. The prognosis with capillary haemangioma is excellent for vision and for life.

With Lymphangioma observation is justified in most cases as surgery may be hazardous. If acute haemorrhage causes symptoms, the lymphangioma may be evacuated and partial resection or ligation attempted. Recurrences are common. Prognosis is variable. Amblyopia is common from globe compression and recurrent haemorrhage. Orbital varices generally do not require treatment. Surgical excision combined with specialized techniques such as embolization

and carbon dioxide laser surgery may be indicated for recurrent thrombosis, pain, severe proptosis and optic nerve compression.⁶

Thus, it is important to keep in mind a categorized differential so that relevant investigations may be ordered to facilitate early treatment and a better outcome.

FINAL DIAGNOSIS

Vascular Hamartoma.

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