

Orbital Lymphangioma in a Child: A Diagnostic Dilemma

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ABSTRACT

We report a case of significant unilateral left proptosis after a fall in a 10-year old child. Magnetic resonance imaging showed an extraconal hyperintense orbital mass extending into the maxillary sinus which was opacified. After drainage the proptosis resolved. The cause of the acute proptosis was haemorrhage within an orbital lymphangioma.

Keywords: Chocolate cysts, orbital lymphangioma, orbital tumour, proptosis

Linfangioma Orbitaria en un Niño: Un Diagnóstico Dilema

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RESUMEN

Se reporta el caso de una proptosis izquierda unilateral significativa en un niño de diez años de edad después de una caída. La tomografía por resonancia magnética mostró una masa orbitaria hiperintensa extracónica que se extendía hasta adentro del seno maxilar, opacificándolo. Después del drenaje, la proptosis se resolvió. La causa de la proptosis aguda fue la hemorragia dentro de un linfangioma orbitario.

Palabras claves: Quistes de chocolate, linfangioma orbitario, tumor orbitario, proptosis

West Indian Med J 2012; 61 (7): 764

CASE REPORT

A 10-year old boy presented with a three-day history of proptosis of the left eye. Five days earlier, whilst at school, he had fallen and hit his head on the concrete pavement. He noted immediate swelling of the left eyelid. He had no associated loss of vision or eye pain. There was no history of loss of consciousness or bleeding at the time of the fall. Two days later, the mother noted that the left eye became proptosed and blood shot. He complained of double vision and eye pain on extraocular movements. His visual acuity remained unchanged and he had no photophobia, eye discharge or fever. There was no history of weight loss.

His best corrected visual acuity was 20/15 and 20/25 in his right and left eyes, respectively. The right eye was normal; significant findings were confined to the left orbit.

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He had a significant non-axial left proptosis and traumatic subconjunctival haemorrhage with posterior limits (Fig. 1). There was significant limitation of elevation, moderate limitation of adduction and minimal limitation of depression in his left eye with associated diplopia.



Fig. 1: Non-axial proptosis of the left eye with significant limitation of elevation.

The intraocular pressures were normal at 10 and 15 mmHg in the right and left eyes, respectively. His fundal examination was normal. No pulsation or bruit was present on auscultation of the orbit. His colour vision was normal and no relative afferent pupillary defect was present. No fractures were present on his orbital X-rays.

Magnetic resonance imaging (MRI) of the brain and orbits revealed an extraconal, non-enhancing mass (Fig. 2). The lateral rectus muscle appeared to be thickened and the

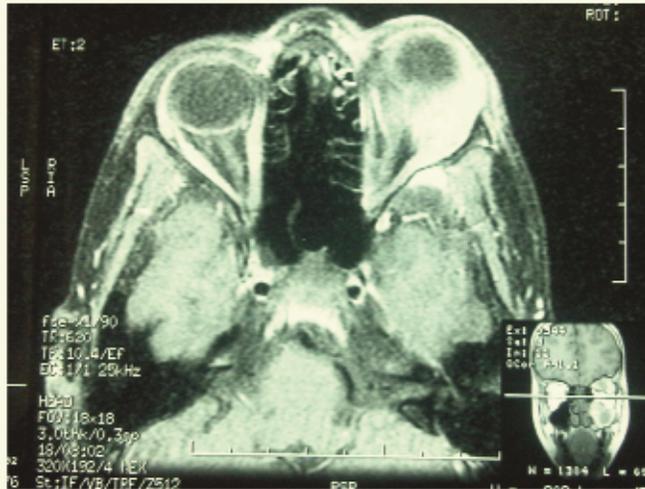


Fig. 2: T1 weighted magnetic resonance imaging shows right proptosis with a poorly defined hyperintense mass.

superior oblique and superior rectus muscles were displaced inferiorly. The left maxillary sinus was completely opacified by a heterogenous mass (Fig. 3). No bony destruction was present.

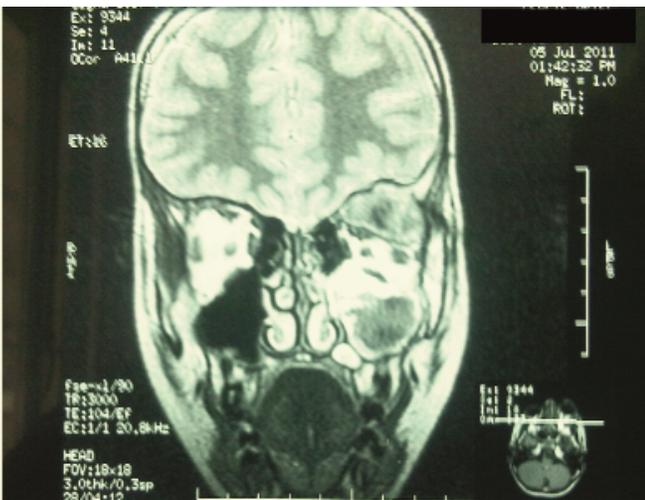


Fig. 3: Magnetic resonance imaging – T1 shows hyperintense circumscribed mass compressing the orbital contents. Note opacification of left maxillary sinus.

Differential diagnosis of the orbital mass included lymphangioma, capillary haemangioma and rhabdomyosarcoma. He was referred to the otorhinolaryngologist and underwent a biopsy of the superotemporal orbital tissue and its extension in the maxillary sinus. Histopathological analysis of the sample revealed spindle cell proliferation in a loose, oedematous myxoid stroma associated with granulation tissue type vessels and a mixed acute and chronic inflammatory cell infiltrate.

A large collection of blood was removed and this resulted in a significant improvement of his proptosis and extraocular movements postoperatively. One-week postoper-

atively, the vision in his left eye was 20/15, diplopia had resolved and colour vision remained normal.

DISCUSSION

Orbital lymphangiomas are vascular malformations of the orbit which are classified based on haemodynamic parameters as Type 1–3. Type 1 lesions have minimal connection to the vascular system whereas Type 2 lesions are connected to the venous system and may be distensible or non-distensible. Type 3 lesions have arterial flow *eg* arteriovenous malformations (1, 2).

Lymphangiomas are abortive vascular systems within normal structures and are usually seen in young children, with a predilection for females under the age of ten years. Approximately 20% of these lesions involve the orbit and ocular adnexa. Jones estimates their incidence to be about 3.4 in 100 000 (3). Lymphangiomas are unencapsulated tumours composed of vascular channels lined with endothelium within a collagenous stromal network (4).

Orbital lymphangiomas may result in acute onset of proptosis secondary to spontaneous intralesional haemorrhage or to lymphoid proliferation as in an upper respiratory tract infection. This may be accompanied by restriction in ocular motility, dystopia, ptosis and compressive optic neuropathy (3, 5).

Orbital lymphangioma may present in several clinical scenarios. Eighty-five per cent will present with proptosis, 73% with ptosis and 46% with limited extraocular movements in a lesser number, with retinal folds (6). Lymphangiomas may be visible on high resolution B scan ultrasound, with diffuse ones having an irregular contour (due to lack of encapsulation) and the cystic ones with a regular contour. Both will have acoustic hollowing present (7). Magnetic resonance imaging can assist with the diagnosis as lymphangiomas have a propensity to bleed into cystic spaces (chocolate cysts). The paramagnetic qualities of the MRI allow it to pick up subacute haemorrhage, making the MRI the modality of choice in investigating lymphangiomas (8, 9). Radiological analysis is only specific in 77% (6) which can lead to a dilemma in diagnosis. It is also important in planning surgery according to the anatomical extent (intra or extraconal or diffuse).

The index case presented with a history of proptosis after minor trauma to the globe. This proptosis improved remarkably after orbital decompression. An MRI scan of the orbit was useful in ascertaining the diagnosis in this case. It may reveal a multilobular structure with septae and haemorrhagic cysts. These haemorrhagic cysts show hyperintense signal on both MRI-T1 and – T2-weighted images whereas lymphatic cysts are hypo-intense on T1-weighted images and hyper intense on T2-weighted images (9).

Orbital lymphangiomas may be managed conservatively provided there is no optic neuropathy or potential for development of exposure keratopathy. In the index case, he had significant proptosis and an early left optic neuropathy

reducing his visual acuity to 20/25. Surgical options include orbital decompression, surgical debulking with carbon dioxide laser and intralesional sclerotherapy (10, 11). Results with systemic steroids have been shown to be a useful adjuvant to surgery (12), however, this has not been corroborated in another study (13). A positive response to steroid treatment may lead to a misdiagnosis of orbital inflammatory disease (orbital pseudotumour), in cases where the MRI is not classical. Poor final visual acuity is associated with multiple surgeries, multiple recurrences and rebleeds (5).

Rapidly progressive proptosis in a child should raise the suspicion of acute orbital cellulitis or rhabdomyosarcoma, the former particularly in the event of fever and post-trauma. Trauma may cause haemorrhage into a previously asymptomatic orbital lymphangioma resulting in the pathological chocolate cysts, proptosis and restriction of eye movements as in the present case. In this situation, radiological assessment can assist in determination of the aetiology of the lesion and surgical debulking/biopsy should be considered, especially in the event of reducing visual acuity from an optic neuropathy or exposure keratopathy.

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