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Congenital Orbital Lymphangioma in a 20-Years Old Girl
A Case Report and Review of Literature

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Abstract; We report a case of a 20-year-old girl who presented to the out-patients' department with congenital, progressive unilateral proptosis and reduced vision. Ultrasound, computed tomography (CT) scan and magnetic resonance imaging (MRI) were performed. Diagnosis of orbital lymphangioma was made on imaging. Authors highlight the crucial role of imaging in diagnosis and to plan therapeutic approach. This case is reported because of its extreme rarity and unusual presentation.

Key words: *Orbital, Lymphangioma, Proptosis, Ptosis.*

Introduction

Orbital lymphangioma is an uncommon, benign cystic lesion generally manifesting in childhood. It usually presents with a slowly progressive proptosis, displacement of the globe, ptosis and restriction of eye movements. Occasionally, focal lesions may remain asymptomatic. Spontaneous intraorbital hemorrhage may cause acute proptosis, compressive optic neuropathy and loss of vision. A 20-year-old girl presented with congenital, progressive unilateral proptosis. We report this case because of its rarity and also review the relevant literature.

Case report

A 20-year-old girl presented to the out-patients' department with unilateral proptosis since birth. This was progressive in nature. There was also an ipsilateral facial deformity with fullness in the supraorbital space (Figure 1). There was no association with pain. There was no history of sudden increase of proptosis. In addition, there was a reduced vision 6/36 in the affected eye. The cornea was clear and so was the lens. Choroidal folds were seen on fundus examination with normal disc and macula. There was no history of redness of the eye. No watery discharge was seen. Ultrasound examination showed multiple cystic intraconal spaces (Figure 2) with very low but turbulent flow. CT scan showed multiple hypodense nonenhancing intraconal lobulated lesions extending into the preseptal space (Figure 3). MRI showed an unencapsulated homogenous lobulated intraconal cystic masses with no extension into the orbital apex or superior orbital fissure (Figure 4). Enlargement of the orbital space however, was seen. In view of these findings, the diagnosis of orbital lymphangioma was considered. Patient underwent excision of the mass as her vision was threatened.

Discussion

Hemangioma and venous lymphatic malformation are the two most common orbital vascular lesions seen in the pediatric patient. Orbital lymphangiomas are benign hamartomatous tumors and may have a locally aggressive nature. They may enlarge slowly producing mild proptosis. However, intrinsic hemorrhage can occur within these lesions, thereby acutely increasing the size of the mass and hence its 'mass effect' leading to intractable, painful progressive sudden proptosis. Differentiation from orbital hemangiomas is important. Unlike hemangiomas, orbital lymphangiomas are not encapsulated, do not respect anatomic boundaries, tend to penetrate into normal structures and can bleed profusely during surgery. For these reasons, an early diagnosis is important. Orbital lymphangiomas are known to be associated with noncontiguous, intracranial venous angioma [1,2] and with vascular abnormalities of the retina and iris [3]. Hemangiomas should be considered in the differential diagnosis as lymphangiomas may rarely simulate cavernous hemangiomas [4]. Ultrasound is the first line of investigation and should always be performed in all such patients. Ultrasonography of hemangiomas show high amplitude closely packed echoes from vessel walls adjacent to blood filled spaces. Lymphangiomas have a similar ultrasound pattern, but with very wide separation of echoes due to larger fluid lakes. Although ultrasound has a high sensitivity and simplicity to be the first choice diagnostic test [5], it suffers from lack of objectivity and extra-orbital delineation of the disease. CT scan plays an important role in diagnosis as well as depiction of extent of the disease due to its multiplanar reformation capability and high spatial resolution. Graeb et al have shown that CT findings correlate well with the surgical and histological findings. On CT scan, orbital lymphangiomas are poorly defined lesions that cross anatomic boundaries such as the conal fascia and orbital septum. Some degree of enhancement is the rule. Areas of hemorrhage cause cyst-like masses with rim enhancement. Hemmer et al emphasized the role of thin-slice CT scanning with multiplanar reformations in preoperative assessment and surgical planning of the disease. In last decade, the role of MR imaging has been emphasized in the literature as it has the capability to precisely delineate and characterize these lesions [6]. It is recommended to use surface coils for higher spatial resolution as this can differentiate between the typical vascular tumors [7]. Moreover, MRI has unique ability to characterize hemorrhage with hemodynamic isolation. MRI has been found to be particularly sensitive to the presence of cysts within the tumor and is also able to predict the contents of the cysts. Tumor feeding vessels can also be detected on MRI by the flow-void phenomenon unique to MRI without the need for intravenous contrast. Song GX in 1991 compared ultrasound, CT scan and MRI in diagnosis of orbital disorders and concluded that MRI was superior to the other imaging modalities in contrast resolution and spatial localization. However, the technique of MRI is particularly

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important for this to be true. In our patient, as there was no history of sudden increase in proptosis, no hemorrhage was detected on MRI. However, CT scan is important in detecting the vascular enhancing component within the tumor as it has a propensity for hemorrhage. Therefore, CT scan enables the surgeon to resect this area to prevent postoperative hemorrhage. The diagnosis of orbital lymphangioma can be made on imaging and biopsy is usually not required. Systemic corticosteroids have been used as an adjuvant treatment to surgery [8], although their role is controversial. The diffuse form of orbital lymphangioma is well known for its difficult surgical treatment and frequent recurrences [9]. Small, repeated excisions may be required. Hsuan et al in 2004 [10] suggested a surgical debulking with a carbon dioxide laser through a lateral orbitotomy combined with a 3-wall orbital decompression. They concluded that bony orbital decompression may be a useful alternative treatment in patients with severe proptosis, secondary to orbital lymphangioma.

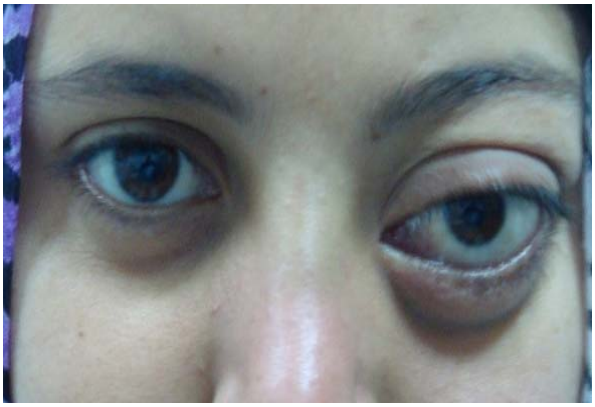

Figure 1 Patient with left proptosis.

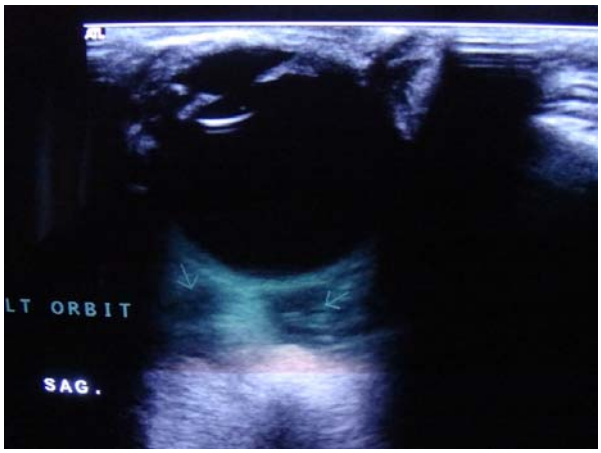

Figure 2: B-mode ultrasonography showed intraconal cystic lesions (Arrow).

We conclude that high resolution CT is of great value in the diagnosis and preoperative treatment planning of orbital lymphangioma. MRI is suitable in those patients who have a rapid increase in proptosis associated with pain.

Acknowledgement

We would like to express our heartfelt gratitude to the patient for providing us her consent to report this case in the scientific journal.

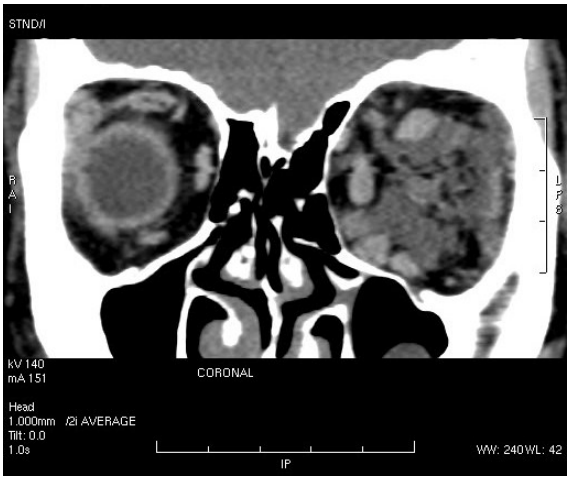

Figure 3: Coronal post-contrast CT scan thin section shows the retrobulbar non-enhancing lesions with preseptal extension.

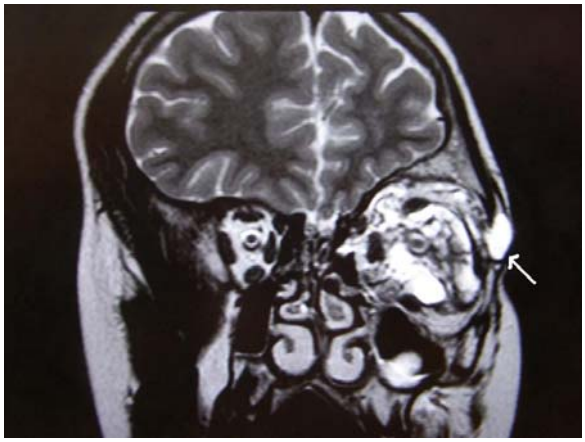

Figure 4: T2-coronal MRI section shows the intraconal cystic spaces with extraconal extension (white arrows).

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