Journal of Advances in Medicine and Medical Research

33(15): 21-26, 2021; Article no.JAMMR.70111
ISSN: 2456-8899
(Past name: British Journal of Medicine and Medical Research, Past ISSN: 2231-0614, NLM ID: 101570965)

Lymphangioma Presenting as Sudden Proptosis in a Toddler

Rajagopal Arvinth[1,2], Visvaraja Subrayan[1], Mimiwati Zahari[1], Prepagaren Narayanan[3] and Sagili Chandrasekhara Reddy[4*]

[1]Department of Ophthalmology, Faculty of Medicine, University of Malaya, Kuala Lumpur, Malaysia.
[2]Department of Ophthalmology, Hospital Ampang, Selangor, Malaysia.
[3]Department of Otorhinolaryngology, Faculty of Medicine, University of Malaya, Kuala Lumpur, Malaysia.
[4]Department of Ophthalmology, Faculty of Medicine and Defence Health, National Defence University of Malaysia, Kuala Lumpur, Malaysia.

Authors' contributions

This work was carried out in collaboration among all authors. Author AR took care of the patient in the ward and followed up the patient's progress, and wrote the first draft of the manuscript. Author VS was the consultant who managed the patient and gave valuable inputs in case write up. Author MZ supervised the preparation of the case write up and corrected the initial draft. Author PN performed endoscopic orbital decompression operation for this patient. Author SCR corrected the paper, added the update references and contributed in writing the introduction and discussion of the paper. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/JAMMR/2021/v33i1530982
Editor(s):
(1) Dr. Evangelos Marinos, University of Athens, Greece.
Reviewers:
(1) Bogdan Ioan Stefanescu, University "Dunarea de Jos" Galati, Romania.
(2) Mosammat Suchana Nazrin, Shahajalal University of Science & Technilogy, Bangladesh.
Complete Peer review History: http://www.sdiarticle4.com/review-history/70111

Case Report

Received 20 April 2021
Accepted 25 June 2021
Published 03 July 2021

ABSTRACT

Orbital lymphangioma commonly occurs in the first decade of life. It is usually congenital, slow growing and can lead to orbital haemorrhage, ptosis, orbital cellulitis, proptosis and compressive optic neuropathy. A 14-months-old male child was brought to the eye clinic with sudden protrusion of the left eye for the past two days. On examination of left eye, a non-axial, non-pulsatile proptosis with ptosis and inferior eyelid ecchymosis were noted. Anterior segment and fundus were normal. Pupil was normal and reacted briskly to light. Intraocular pressure measured with Tonopen was 16 mm Hg. Right eye: anterior segment, fundus, and intraocular pressure were normal. Magnetic resonance imaging of orbits and brain showed left sided multicystic intraconal mass with area of

**Corresponding author: E-mail: profscreddy@gmail.com;*

haemorrhage. The diagnosis of left orbital lymphangioma was made. The initial treatment with Propranolol syrup did not reduce the proptosis. Left nasal endoscopic aspiration of blood and orbital decompression was done by the otolaryngologist. The child was followed up for one year and the proptosis was found to be reduced markedly on the left side. Although various modalities of treatment are available for this condition, unfortunately there are no definitive curative treatments currently. In view of its rarity, this case is reported.

Keywords: Proptosis; orbital lymphangioma; orbital decompression; sclerotherapy.

1. INTRODUCTION

Orbital lymphangioma, a rare benign venous and lymphatic malformation, occurs most commonly in the first decade of life. It is congenital and slow-growing, consisting 0.3 to 1.5% of all orbital tumours [1]. It can present with spontaneous orbital haemorrhage [1], ptosis [2], orbital cellulitis [3] and proptosis [4]. In some of these patients proptosis may develop either slowly as the mass invades the orbit or suddenly during hemorrhage in the lesion [5].

In childhood, the diagnosis is often made when proptosis occurs after bleeding as a result of minor trauma or upper respiratory infection, and may even occur spontaneously. The lymphangioma itself or the associated bleeding can restrict ocular motility and cause compressive optic neuropathy because of its mass [6]. Lymphangiomas are unencapsulatetd tumours composed of vascular channels lined with endothelium within a collagenous stromal network [7].

Several methods have been used to treat orbital lymphangioma, including systemic corticosteroids, intralesional injection of a sclerosing agent (sclerotherapy), and partial resection of the major cyst, needle aspiration in cases of spontaneous haemorrhage, local radiotherapy, and endoscopic orbital decompression in cases of spontaneous haemorrhage; but currently, there are no definitive curative treatments [5-8].

The literature search (PubMed, Scopus, Science Direct and Google scholar) did not show any published report on orbital lymphangioma from Malaysia. Therefore, we report this rare case of a 14 month old boy with bleeding orbital lymphangioma manifesting as acute proptosis.

2. CASE REPORT

A 14 month old boy was brought to the eye clinic with history of sudden onset of protrusion of the left eye for the past two days, associated with upper respiratory tract infection. Parents told that the child has sight prominence of the left eye compared to the right eye since birth. They consulted local ophthalmologist who referred the patient to the teaching university hospital for further management.

On examination, child demonstrated preference to see with the right eye. Left eye: There was a non-axial, non-pulsatile proptosis of the left eye with ptosis and inferior ecchymosis (Fig.1) Anterior segment and fundus were normal. Pupil was normal and reacted briskly to light. Intraocular pressure measured with Tonopen was 16 mm Hg. Right eye: anterior segment, fundus, and intraocular pressure were normal. Best corrected visual acuity (using preferential looking tests) was 6/60 with cyclo-refraction of +6.00 D in the left eye and 6/12 with a cyclo-refraction of +1.25D in the right eye.

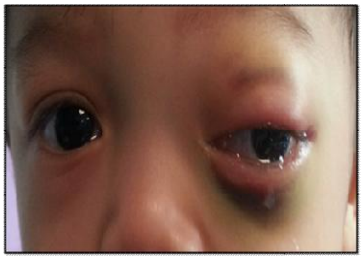

Fig. 1. Showing proptosis of left eye and ecchymosis below the lower eyelid

The child was admitted in the eye ward. Informed consent was taken from the father for investigations and operation, if required. The common causes of proptosis in pediatric age group such as inflammations (orbital cellulitis), vascular malformations (capillary hemangioma), developmental (dermoid cyst) and neoplasms (retinoblastoma, rhabdomyosarcoma, leukemia and lymphoma) were excluded by history taking, clinical examination and doing appropriate tests (Full blood counts, nasal swab for culture, and Magnetic resonance imaging (MRI) orbits and brain). There was no evidence of

inflammation/infection in the blood investigations. Nasal swab culture was sterile. The MRI of the orbits showed left sided multicystic intraconal mass with area of haemorrhage; there was no intracranial extension (Figs. 2,3).

The child was then given a course of syrup Propranolol 15mg/kg in a single dose for a month. Upon follow up review one month later, there was no reduction in the proptosis. An otorhinolaryngology consultation was taken, and a left nasal endoscopy revealed a bleeding cystic mass (Fig.4).

Under general anaesthesia, aspiration of the blood from the cyst and orbital decompression were performed by removing part of the medial and inferior wall of the orbit. Histopathology of the removed mass showed blood clots and thus histological diagnosis was not confirmed. Post-operatively, syrup Cloxacillin 15mg/kg six hourly was given for one week along with nasal package. He was discharged from the hospital after review by the otorhinolaryngologist. Ciprofloxacin eye drops were given three times daily in the left eye and the frequency was reduced weekly. Review of the child one month later showed resolution of ecchymosis and mild reduction of proptosis (Fig. 5).

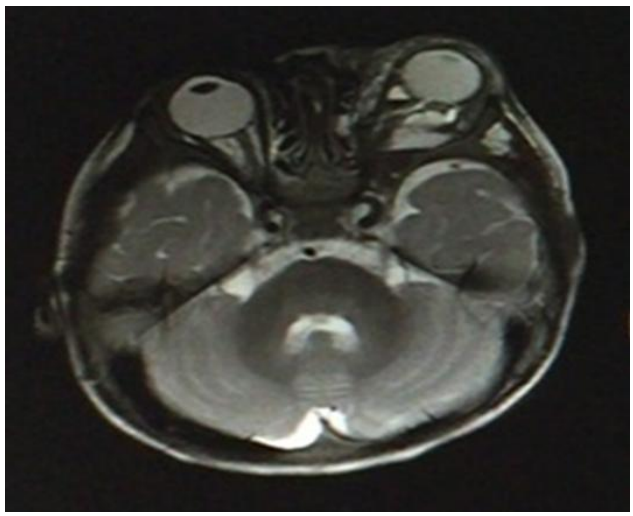

Fig. 2. Showing the tumour in the left orbit and proptosis of left globe on T2 MRI

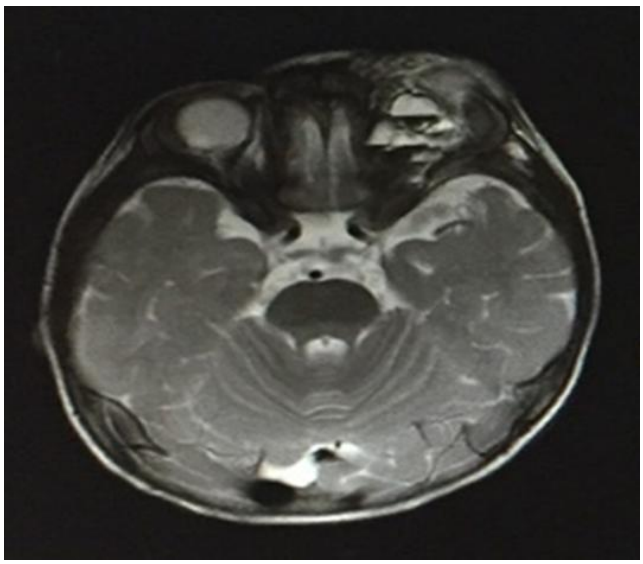

Fig. 3. Showing hyper dense area of haemorrhage in the left orbit on T2 MRI

The child was followed up every two months for one year. On the last follow up, the left eye proptosis was found to be reduced markedly.

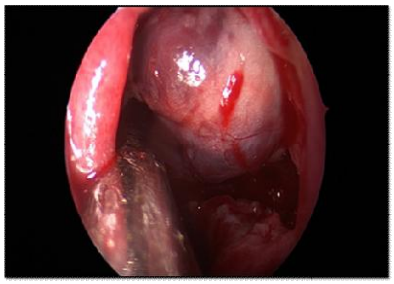

Fig. 4. Showing endoscopic view of orbital lymphangioma with bleeding chocolate cyst and blood clot

Fig. 5. Showing resolution of ecchymosis below the lower eyelid and mild reduction of proptosis of left eye

3. DISCUSSION

Orbital lymphangioma can be classified into four types based on location [8]: the superficial type is characterized by subcutaneous or conjunctival lesions; the deep type shows orbital invasion; the combined type has superficial and deep components; and the complex type involves other head and neck structures.

In our case, the lymphangioma was deep type in view of haemorrhage in the orbit and proptosis of left eye; and there was no conjunctival lesion.

Orbital lymphangiomas are not very commonly detected in children less than 6 months of age. In a study conducted by Tunc et.al [9], out of 26

patients only five of them were diagnosed before 6 months of age. in their study, 85% of patients had proptosis; 73% had ptosis; and 46% had reduced ocular motility. Woo et al [4] reported that two out of 12 patients were aged 1 year; and all patients had proptosis and limited ocular motility at presentation. Our patient was a 14 months old boy who was brought to the eye clinic with acute proptosis of two days duration.

The progression of orbital lymphangiomas to cause proptosis which usually occurs much later; second decade of life, as shown in case reports by Salihu et.al [1] and Mishra et.al [2]. However, in our patient, the progression was seen at such an early age of 14 months. The orbital lymphangioma being space-occupying lesions of the orbit, may cause compressive optic neuropathy, amblyopia, secondary glaucoma and exposure keratopathy [9] over a period of time if it remains untreated.

Ozeki et.al [10] reported that propranolol may have some benefit in the shrinking of intractable lymphangiomas. Our patient was first managed conservatively with syrup propranolol. However, there was no reduction in the proptosis. Therefore, a nasal endosopic orbital decompression the orbit was done by the otorhinolaryngologist, which is a safe, minimally invasive procedure.

Hsuan et al [11] performed surgical debulking with a carbon dioxide laser through a lateral orbitotomy combined with a 3-wall orbital decompression in a patient with 11 mm of proptosis and exposure keratopathy, secondary to an orbital lymphangioma. They suggested that orbital decompression reduces the proptosis more than the debulking surgery itself.

Gandhi et al [12] recently published their success with sildenafil in two young boys with aggressive lesions who did not respond to drainage and sclerosing procedures. The use of intraoperative fibrin glue has been described as an aid to surgical excision [13]. The fibrin glue is injected into the cystic components of lymphatic malformations in order to provide a more solid structure to aid in dissection and improve hemostasis. Other techniques involve the combination of fibrin glue and sclerosing agents to help in debulking these lesions.

A potentially promising treatment involving the injection of OK-432 (which is prepared from a low-virulence strain of group A Streptococcus pyogenes of human origin treated with penicillin G) into the lesion showed shrinkage of the orbital lymphangioma without causing functional

disturbance and scar in the facial skin. However, it requires more time to assess the efficacy and safety of this treatment as it is still in its early stages [14,15].

Radiographic features of orbital lymphangiomas are ill-defined, poorly marginated, non-encapsulated trans-spatial multiloculated cystic and solid orbital lesions that commonly involve the whole orbital spaces including preseptal, postseptal, intraconal, and extraconal compartments [16]. In computed tomography (CT) imaging, the lesion is complex and contains cystic and solid areas. The cystic areas elicit fluid density and represent the lymphatic component of the lesion while the solid areas appear hyperdense to brain parenchyma on non-contrast CT and represent the venous component of the lesion. Intralesional phleboliths are occasionally seen and appear as small dense foci within the solid venous part of the lesion.

CT may demonstrate associated bony remodeling secondary to the compressive effect of the lesion on adjacent bones.

MRI is superior to CT in the evaluation of these lesions, perfectly assessing their extension and depicting various components. MRI signal is variable, depending on the fluid contents and the age of internal blood, however, it is commonly presented as follows:

T1: iso- to hyperintense to brain parenchyma
T2: hyperintense to brain parenchyma with occasional multiple fluid-fluid levels
T1 C+ (Gd): marginal and septal enhancement of the cystic spaces with a variable enhancement of the solid components.

MRI can be of great help for the surgeon to plan the treatment accordingly, because they have a tendency to bleed profusely during a total surgical resection of the mass. This is also useful in the assessment of recurrence of these tumours.

Early childhood orbital lymphangiomas are extremely challenging to the managing team to decide on the appropriate time for surgery as careful consideration needs to be given on the benefits that would be obtained following the procedure. In the management of pediatric lymphangiomas, many specialists (ophthalmologist, radiologist, pediatrician, otorhinolayngologist) should become the team members so that the diagnosis can be made early and appropriate treatment can be given in-time.

4. CONCLUSION

Oral corticosteroids can be used in the treatment of lymphangioma in patients with uncompromised vision. In patients who have raised intraocular pressure or threatened vision loss due to acute haemorrhage, aspiration of blood and intralesional injection of a sclerosing agent is the preferred treatment. In patients with organized hematoma, partial resection may be effective. In patients with severe proptosis causing compressive optic neuropathy, orbital decompression will be helpful.

CONSENT

The child was admitted in the eye ward. Informed consent was taken from the father for investigations and operation, if required.

ETHICAL APPROVAL

It is not applicable. Written informed consent was obtained from the parent for the publication of the case report and accompanying photos of the child.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Salihu N, Sylaj A. Orbital lymphangioma. International Journal of Case Reports and Images. 2014;5(1):28-31.
2. Mishra A, Abuhajar R, Alsawidi K, AlAoud M, Ehtuish EF. Congenital Orbital Lymphangioma in a 20 years old girl. A Case Report and Review of Literature. Libyan J Med. 2009;4(4):162-3.
3. Brooks SE, Pillai JJ, Bains H. Orbital Cellulitis due to Occult Orbital Lymphangioma. South Med J. 2001; 94(5):532-5.
4. Woo YJ, Kim CY, Sgrignoli B, YoonJS. Orbital Lymphangioma: Characteristics and Treatment Outcomes of 12 Cases. Korean J Ophthalmol. 2017;31(3):194-201.
5. Russin JJ, Rangel-Castilla LM., Kalani YS, Spetzler RF. Surgical Management, outcomes and recurrence rate of orbital lymphangiomas. J Clin Neurosci. 2015; 22(5):877-2.
6. Lee KH, Han SH, Yoon JS. Successful treatment of orbital lymphangioma with intralesional bleomycin and application of

continuous negative pressure. Korean J Ophthalmol 2015;29(1):70-2.
7. Yanoff M, Sassani J. Ocular pathology. 6 th edition Philadelphia: Mosby Elsevier; 2009: 546-7.
8. Saha K, Leatherbarrow B. Orbital lymphangiomas: a review of management strategies. Curr Opin Ophthalmol 2012;23(5):433-8.
9. Tunc M, Sadri E, Char DH. Orbital Lymphangioma: An Analysis of 26 patients. Br J Ophthalmol 1999; 83(1):76-80.
10. Ozeki M, Kanda K, Kawamoto N, Onishi H, Fujino A, Hirayama M et.al. Propranolol as an alternative treatment option for paediatric lymphatic malformation. Tohoku J Exp Med 2013; 229(1):61-6.
11. Hsuan J , Malholtra R , Davis G , Selva D. Orbital decompression for gross proptosis associated with orbital lymphangioma. Ophthal Plast Reconstr Surg. 2004; 20(6):463-5.
12. Gandhi NG, Lin LK, O'Hara M. Sildenafil for pediatric orbital lymphangioma. JAMA Ophthalmol. 2013;131(9):1228-30.
13. Boulos PR, Harissi-Dagher M, Kavalec C, Hardy I, Codere F. Intralesional injection of Tisseel fibrin glue forresection of lymphangiomas and other thin-walled orbital cysts. Ophthal Plast Reconstr Surg. 2005;21(3):171-6.
14. Suzuki Y, Obana A, Gohto Y, et al. Management of orbital lymphangioma using intralesional injection of OK-432. Br J Ophthalmol 2000;84(6):614-7.
15. Yoon JS, Choi JB, Kim SJ, Lee SY. Intralesional injection of OK-432 for vision-threatening orbital lymphangioma. Graefes Arch Clin Exp Ophthalmol 2007;245(7): 1031-5.
16. Bell DJ, Mohamed Saber M et al. Orbital lymphangioma. Availavle:https://radiopaedia.org/articles/orbital-lymphangioma Accessed on June 9, 2021

Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle4.com/review-history/70111