Case Report

Adult Onset Orbital Lymphangioma Presenting with Globe Luxation: A Rare Presentation of A Rare Disease

Deepsekhar Das, Swechya Neupane, Sahil Agrawal, Mousumi Banerjee
Mandeep Singh Bajaj, Seema Kashyap

Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India

Abstract

Lymphangiomas or lymphatic vascular malformations typically present in adolescent age groups. Adult onset lymphangiomas are extremely rare occurrences. The condition usually presents with proptosis or extraocular motility disorder. The authors here report a 50-year-old female who presented with a painless progressive proptosis of the left eye for the last 10 years. She gave history of episodes, where the globe spontaneously came out of the socket, which she used to then push back. Radiological imaging of left orbit revealed a large retrobulbar lesion occupying the whole of the left orbit with luxation of the left globe. There were no obvious bony erosions or intracranial extension. She underwent an excisional biopsy which came out to be positive for adult onset lymphatic vascular malformation. Although proptosis is a usual presenting feature of an orbital lymphatic malformation, globe luxation as a presenting complaint has never been documented.

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Introduction

Orbital lymphangioma is a rare, diffusely infiltrating benign non-encapsulated vascular malformation, usually diagnosed in the first or second decade of life.[1] Adult onset lymphangioma or late onset lymphangioma is the one which presents after 30 years of age.[2] Here we report a case of adult onset lymphangioma in a 50 year old female.

Case Report

A 50-year-old female presented with painless progressive proptosis of the left eye for the past 10 years. (Figure 1) There was no history of trauma, weight loss, anorexia, or any significant systemic illnesses. There was history of episodes of the eyeball coming out of the orbit spontaneously causing pain. The patient used to push the eyeball back inside the socket.

On general examination, she was alert, conscious and well-oriented to time and place and systemically stable.

On local examination, there was gross abaxial proptosis of the left globe with a visible mass in the supero-temporal aspect causing globe dystopia. The left upper eyelid was lifted by an underlying mass. There was exposure keratopathy of the cornea and marked extraocular muscle movements limitation was noted.

On palpation, the globe almost luxated (Figure 2), we could feel a 5.5 cm x 5.5 cm well defined solid, non-tender, non-pulsatile mass extending from the superior orbital rim to the lateral canthus, free from the overlying eyelid with fixity to the underlying structures with no increase in size on Valsalva maneuver.

On examination, BCVA in her right eye was 20/20. However, she denied any perception of light in her left eye. Her right eye had brisk direct pupillary response whereas there was fixed, 5 mm dilated pupil noted in the left eye with no appreciable fundal glow.

A Contrast Enhanced Computed Tomography (CECT) brain and orbit revealed a 6 cm x 5 cm well defined orbital mass with inferomedial globe dystopia with no intracranial

Figure 1 *: Clinical picture of the patient on presentation showing severe proptosis with globe dystopia*

Figure 2 *: Clinical picture showing luxated left globe with a mass located superotemporal to the globe along with exposure keratopathy*

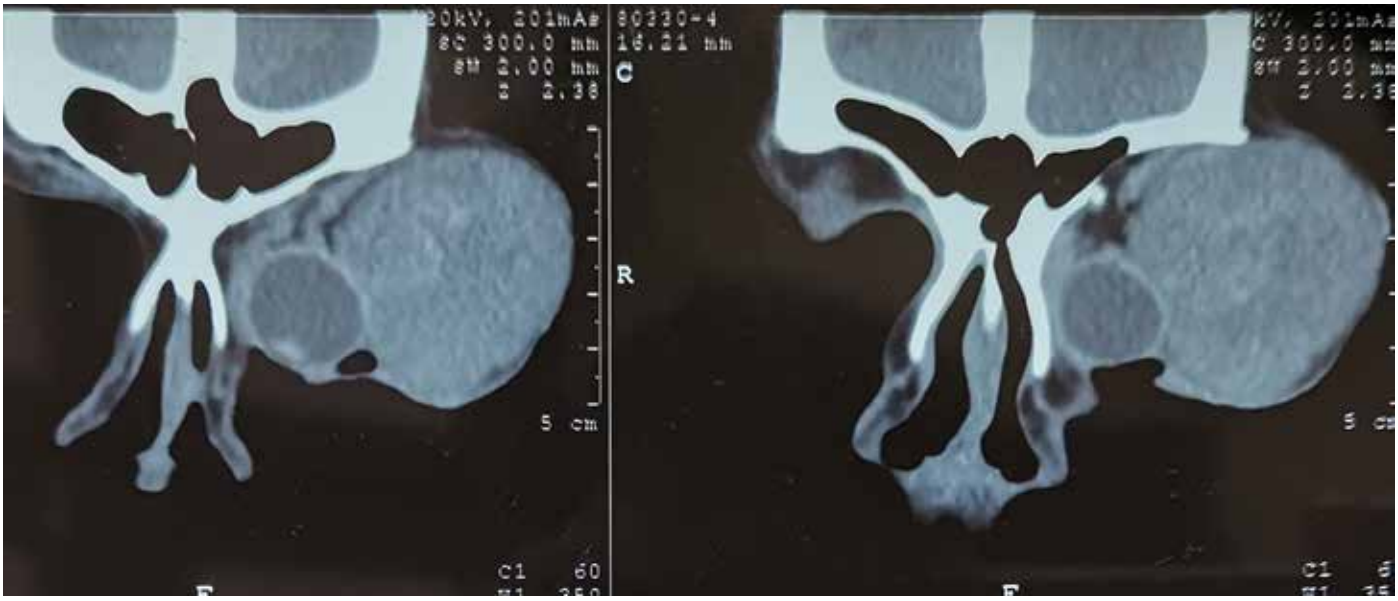

Figure 3 *: CECT images showing a well-defined mass in the left orbit with globe dystopia.*

extension or bony changes (Figure 3). Ultrasonography of the left eye revealed an anechoic vitreous cavity without and obvious retinal pathology or optic nerve head cupping. Left orbital ultrasound showed a well-defined lesion with few cystic spaces behind the globe.

Left anterior orbitotomy with complete removal of mass was planned. Under general anaesthesia, using a modified Stallard Wright incision, a plane was created, and the lesion was identified as a well-defined, non-encapsulated, soft mass. The mass was then removed and its base was cauterised making sure not to damage the optic nerve. In post-operative phase, the patient was treated with oral antibiotics and steroids post-operatively. Histopathology of the specimen revealed numerous thin walled vascular channels lined by flattened endothelial cells filled with lymph along with lymphoid aggregates in the stroma (Figure 4).

At 1 week follow up, the globe dystopia and proptosis had completely gone away (Figure 5). There was no recurrence noted on 6 months follow up.

Discussion

Orbital and adnexal lymphangiomas comprise 20 % of all lymphangiomas and only 2% of them account for orbital biopsies.1 One of the most important features of orbital lymphangioma is swelling that increases gradually in size. Sudden increase in size of the swelling can be attributed to intraorbital haemorrhage or respiratory infections. Various signs include ocular proptosis, blepharoptosis and cellulitis. Acute proptosis, compressive optic neuropathy and loss of vision are attributed to spontaneous intraorbital haemorrhage.1 There is a lot of controversy regarding pathogenesis of orbital lymphangiomas. According to Harris et al., lymphangioma is considered isolated from normal orbital vasculature whereas Wright et al. considered them to be venous anomalies.[3] The congenital form occurs because of improper connection of lymphatic channel to the main lymphatic draining duct. Adult onset lymphatic malformations, or late onset lymphatic malformations which are characterised by presentation after 30 years of age are extremely rare with very limited number of cases to be reported in literature. Acquired lymphangiomas occur due to impaired lymphatic drainage as a consequence of surgery, trauma, malignancy, or radiation therapy.

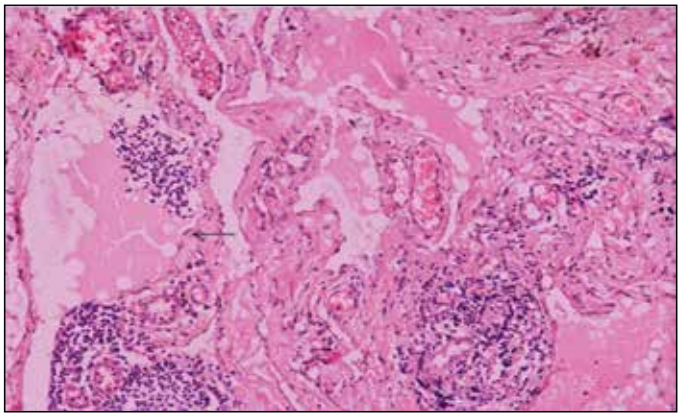

Figure 4 *: Histopathology of excised lesion showing numerous thin walled vascular channels lined by flattened endothelial cells (arrow) filled with lymph along with lymphoid aggregates in the stroma.*

Figure 5 *: Clinical picture of the patient on follow up with no proptosis.*

Lymphangiomas show benign features on histopathology in contrast to their locally aggressive nature with friable and non-encapsulated lesions on gross appearance, thus making recurrences common even after resection of the tumor. On histology, lesions have thin walled stroma, a single layer of flattened epithelium along with occasional breaks in continuity and lymphoid infiltration. There may be presence of lymphoid aggregates in subendothelial spaces and haemorrhagic material in cystic spaces. Lesions often show large, irregular vascular spaces that are lined by a single layer of endothelial cells within fibroblastic stroma.[4]

Diagnostic imaging like Magnetic Resonance Imaging (MRI) plays an important role in anatomical demonstration of cystic nature and haemorrhages present in lymphangiomas. On T2 weighted images, venous component enhances whereas lymphatic component shows fine enhancement of septations.5 Pathognomonic finding includes cyst with multiple fluid-filled levels. However, the appearance may be absent in cases of adult onset lymphangiomas. Treatment options for orbital lymphangiomas are conservative management, partial resection of cyst, needle aspiration, anterior orbitotomy with total removal and intralesional injection of sclerosing agents like alcohol, bleomycin and tetracycline. In presence of orbital haemorrhage leading to compressive optic neuropathy, orbital decompression is the only option to restore vision. Apart from compressive optic neuropathy, other complications of orbital lymphangiomas include astigmatism, corneal exposure, hyperopia secondary to pressure on posterior globe, strabismus and glaucoma.[5] Based on clinical findings and histopathological examination, diagnosis of adult onset orbital lymphangioma was made in this case. Adult onset orbital lymphangioma is an exceedingly rare entity that has very limited literature. The usual management varies from surgical excision, debulking to intralesional bleomycin application.[6]

Globe luxation is a situation when the eyeball comes in front of the orbital rim and appears caught in between the eyelid aperture. Usual causes of globe luxation are trauma, strenuous conditions like chronic obstructive pulmonary disease or spontaneous luxation in cases of shallow orbital cavity.[7] After performing a thorough literature search, we believe that this is the first time a case of adult onset lymphangioma has presented with globe luxation.

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Address for correspondence

Sahil Agrawal
Oculoplasty and Orbital Tumor Services
Dr Rajendra prasad centre
for ophthalmic sciences,
All India Institute of Medical Sciences
New Delhi India
E-mail-agrawalsahil03.acad@gmail.com

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