CASE REPORT

An acute presentation of orbital lymphangioma in a five-year-old child in Pakistan

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Abstract

Lymphangiomas are multi-cystic malformations involving the lymphatic and vascular systems. Most commonly occurring in the head and neck region in the paediatric age group, lymphangiomas generally are benign in nature.

Given the tumour's mass, the lymphangioma itself or the accompanying haemorrhage may impair ocular movement and result in compressive optic neuropathy.

Furthermore, lymphangiomas have the potential to unevenly infiltrate nearby critical structures including the optic nerve. This trait poses numerous surgical difficulties and makes the therapy of ocular lymphangiomas particularly challenging, especially with the associated haemorrhage.

This case report presents an acute case of orbital lymphangioma in a five-year-old child who was treated with monthly sessions of intralesional foam sclerotherapy. Complete resolution of the lesion was observed at the end of the sclerotherapy sessions.

Keywords: Orbital Lymphangioma, Child.

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Introduction

Lymphangioma is a benign tumour of the lymphatic system characterised by abnormal endothelial lined channels.[1,2] It can be congenital due to improper conduits of lymphatic channels in the main lymphatic duct or acquired later in life as a result of surgery, trauma, or malignancy.[3] Orbital lymphangioma is a lymphatic and vascular channel pathology occurring in early childhood. The tumour is normally not evident until late in the first decade of life.[4] Patients typically present with pain, diplopia, visual loss and demonstrate axial or non-axial proptosis, restricted eye movement and/or globe displacement on further examination.[2,5] Current treatment modalities for orbital lymphangioma include partial resection, needle aspiration or sclerotherapy via sclerosing agents.[4]

This report details the treatment of an acute orbital lymphangioma in a five-year-old child who underwent monthly intralesional foam sclerotherapy sessions. Consent was taken from the patient's guardians for recording of data and reporting of findings.

Case Report

A five-year-old boy presented on October 5, 2021, to the Ophthalmology outpatient department at the Lahore General Hospital, Lahore, with sudden onset of painful proptosis of 34mm (on Hertel) and a temporal dystopia of 16 mm in the left eye. Visual acuity at the time of presentation was counting fingers at one metre, while extraocular movements were restricted in all gazes. Anterior segment examination demonstrated conjunctival chemosis, corneal oedema, and ongoing exposure keratopathy. Increased resistance to retrodisplacement of the globe was appreciated on compression (retropulsion) test. No ocular bruit on auscultation, no previous history of head trauma, pre-existing orbital disease or any upper respiratory tract infection was documented.

The B-mode ultrasound scan (B-scan) of the left eye revealed a retro-orbital, intra-conal, hyperechoic, and haemorrhagic lesion. T1- and T2- weighted magnetic resonance imaging scans (MRI scans) of the orbit were done to establish a diagnosis which showed a well-defined and lobulated intraconal mass with the optic nerve encasement but no involvement, as shown in Figure 1. Two sessions of sclerotherapy were conducted six weeks apart. Before the initiation of sclerotherapy, the patient underwent a thorough paediatric evaluation, including medical history, physical examination, and screening for conditions that might contraindicate the use of Bleomycin. Respiratory and renal functions were specifically assessed to avoid potential Bleomycin toxicity. The evaluation confirmed that the patient was fit for treatment. Bleomycin dosage was calculated according to body surface, according to paediatric safety guidelines[6].

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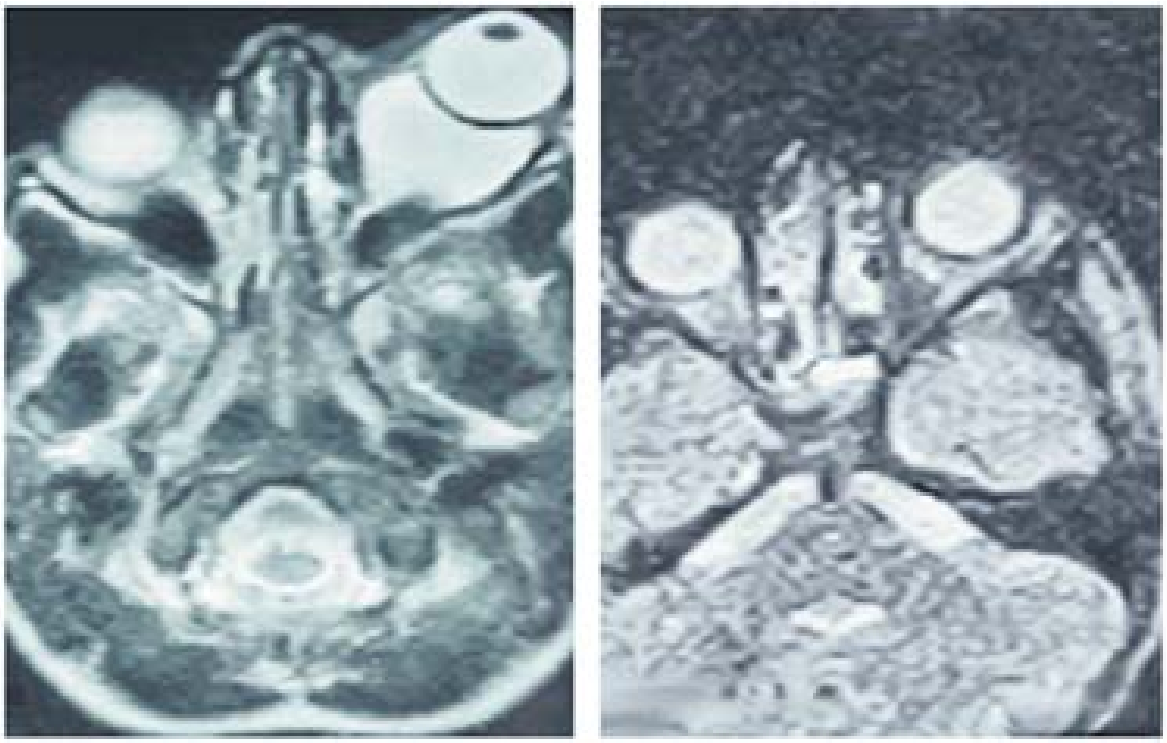

Figure-1: Pre- and Post-treatment T2- weighted MRI scans of the orbit.

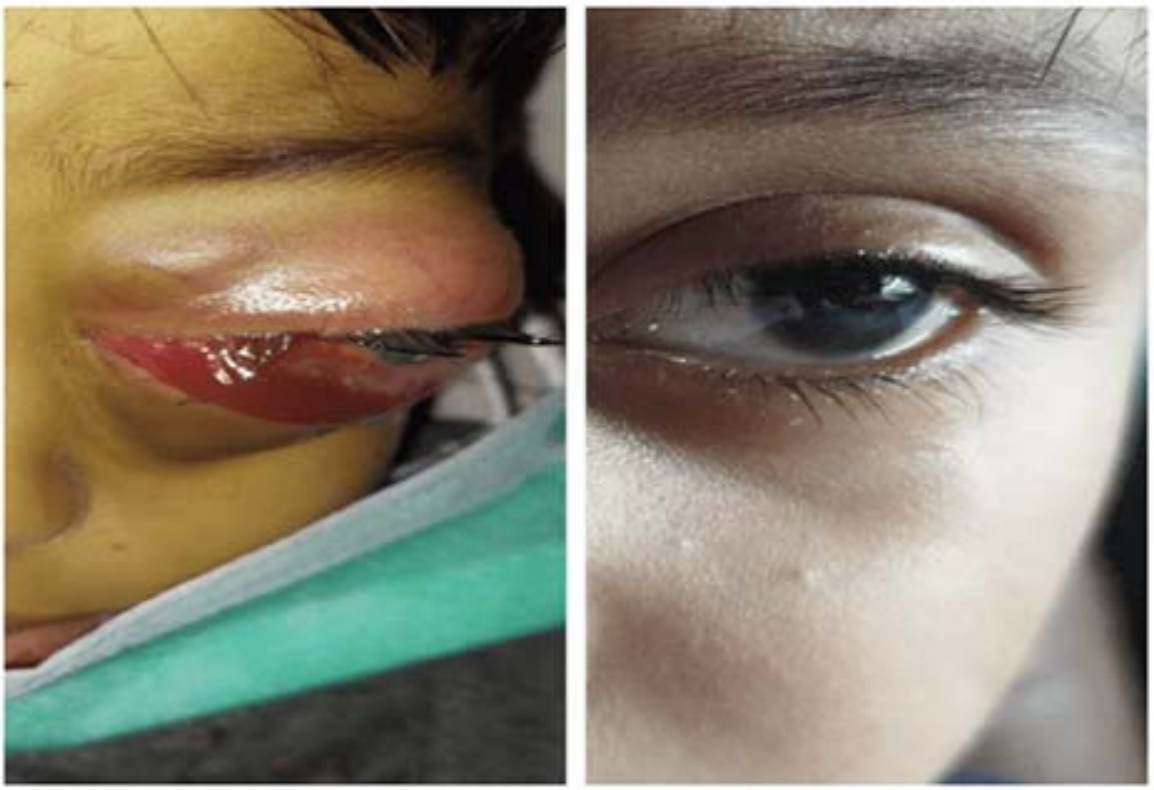

Figure-2: Pre- and post-treatment gross transformation.

The ophthalmic surgeon then proceeded with aspiration of the haemorrhagic cyst under aseptic measures. Histopathological analysis confirmed the diagnosis of lymphangioma. The biopsy revealed large cystic lymphatic channels, lined by a single layer of endothelial cells, consistent with the descriptions in ophthalmic pathology literature.[7] These channels were interspersed with small, thin-walled blood vessels. The histopathological examination did not reveal any evidence of malignancy, supporting the benign nature of the lesion. Monthly sessions of intra-lesional sclerotherapy were planned using Bleomycin foam as a sclerosing agent under Digital Subtraction Angiography (DSA) guidance. The foam was prepared using two 5ml syringes, each with a three-way stopcock valve and then carefully one part of Bleomycin sulphate 3mg/ml, one part of 20% human albumin, and four parts of sterile air were mixed. This resulted in the mixture of Bleomycin foam. DSA-guided foam sclerotherapy using the negative subtraction technique was performed with 12ml Bleomycin foam in the first session. The patient was discharged on a short course of oral Prednisolone (Deltacortil) 1mg/kg for seven days.

On the thirty-day follow-up, the proptosis had resolved completely, and the dystopia had significantly reduced from 16mm to 2mm with a slight residual lesion persisting laterally. The second session of DSA-guided foam sclerotherapy with negative subtraction technique was performed with 6ml Bleomycin foam. Pre- and post-treatment contrast images of DSA were also taken to monitor the progress of the treatment. On the ninety-day follow up, the patient showed no physical disfigurement as shown in Figure 2. An MRI scan was conducted which showed minimal residual lesions with no contrast enhancement and complete vision had been restored.

Discussion

Depending on the site of the lesion, orbital lymphangiomas can be categorised into four types: the superficial type involves the conjunctiva; deep type is characterised by involvement of the orbit; combined type has features of both superficial and deep variants; and the complex type involves the head and neck region.[2] This patient's tumour was classified as deep. Common presentations include sudden onset of painful proptosis, decrease in visual acuity, conjunctival chemosis, and cosmetic defects.[5] Patients also commonly give a history of an upper respiratory tract infection, a minor head trauma or even physical exertion which can exacerbate a gradually progressing proptosis.[6] However, this is in contrast with the patient's situation who was afebrile on presentation and gave no history of recent trauma to the head. Soft and hard palate haemorrhage and intracranial arteriovenous malformation were also not present even though they usually tend to co-exist with this disease.[3]

The imaging evaluation of suspected orbital tumours, including orbital lymphangiomas can be achieved by ultrasonography (USG), magnetic resonance imaging (MRI), computed tomography (CT), with MRI being the recommended modality of choice.[8] In this case, T1- and T2- weighted MRI scans of the orbit as a pre-treatment workup and post-treatment monitoring were used for assessment of tumour progression and recovery. The mass showed isointense T1 signals with post contrast enhancement and hyperintense signals on T2-weighted imaging. Layering blood products, indicating intralesional haemorrhage, were also appreciated. B-scan ultrasonography revealed a typical round, regular, and well-demarcated hyperechoic haemorrhagic lesion in the intraconal space. High resolution B-scan is useful in differentiating orbital lymphangioma from other orbital tumours, especially when no clinical lid or conjunctival involvement is observed.[9]

Surgical resection or needle aspiration are also workable approaches but carry possibilities of recurrence and complications.[4] In contrast, sclerotherapy offers a minimally invasive and highly targetted treatment option, particularly effective for cases with haemorrhagic components, like the one under discussion. Aspiration of the haemorrhagic component was completed before administering intralesional injections of Bleomycin with application of continuous negative pressure. A literature search revealed recurrence of haemorrhage in patients who received injection of a sclerosant without application of continuous negative pressure.[2] Studies have demonstrated that an increase in contact time between the sclerosing solution and the vessel wall has prolonged therapeutic effects in treatment, as a uniform distribution of the agent results within the vasculature, thus requiring a minimal treating dose and allowing for more consistent results with fewer complications as compared to liquid sclerotherapy or traditional surgery.[10]

Conclusion

Orbital lymphangiomas are uncommon and can be challenging to diagnose considering the various orbital pathologies they resemble. Early diagnosis and intervention are crucial. Intralesional foam sclerotherapy proved effective in reducing the tumour size with complete vision restoration. This case highlights the efficacy of sclerotherapy as a safe first line treatment in managing orbital lymphangioma.

Consent form: Informed consent was taken from the

patient's guardians prior to the submission of the manuscript for publication.

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Authors Contribution:
JR: Patient care and follow-up, literature review and research, principal investigator.
AM: Literature Liasson and research, manuscript editing.
SA: Manuscript writing.
KKA: Critical review and approval of final manuscript.