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Case Report

Elective sclerotherapy of an orbital lymphatic malformation: A case report and review of the literature

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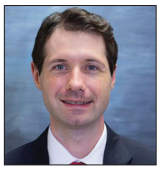

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ABSTRACT

Orbital lymphatic malformations (LMs) are a unique subtype of vascular anomaly that can swell and lead to blindness if left untreated in the acute setting. Detrimental acute and chronic effects of the malformation on vision suggest that treatment should be considered pre-emptively rather than in the acute setting. We describe the effectiveness and safety of treating these LMs electively using sclerotherapy, a minimally invasive treatment method.

Keywords: Sclerotherapy, Ultrasound, Interventional radiology

INTRODUCTION

Lymphatic malformations (LMs) are abnormally developed lymphatic tissues present from birth characterized by fluid-filled pockets of lymph. LMs can swell and cause pain or damage to adjacent structures, especially in closed spaces such as the orbit. Swelling from orbital LMs is often an emergency that requires immediate drainage, surgical removal, or sclerotherapy. We report a case of elective orbital LM treatment.

CASE REPORT

An 8-year-old girl was referred to our clinic with 6 days of painless right eye swelling and proptosis. She was born with a small bump over her right eyebrow but did not have any prior episodes of orbital swelling or globe displacement. No similar lesions were present elsewhere on her body, and the family denied any recent history of infection, trauma, contact lens use, or topical medication administration over the eye.

Physical examination revealed a proptotic orbit displaced inferolaterally with partially reducible swelling and a brown skin stain along with the medial eyebrow. The cornea, sclera, iris, pupillary size, extraocular movements, and vision were normal.

Initial head CT scan revealed a multiloculated right orbital lesion with fluid-fluid levels [Figure 1]. Subsequent MRI of the orbits confirmed the presence of multiple complex non-enhancing cysts consistent with a mixed macrocystic and microcystic lymphatic malformation [Figure 1].

Malformation treatment was scheduled as an outpatient procedure given her urgent, but not emergent symptoms. However, when she presented for treatment, the malformation had

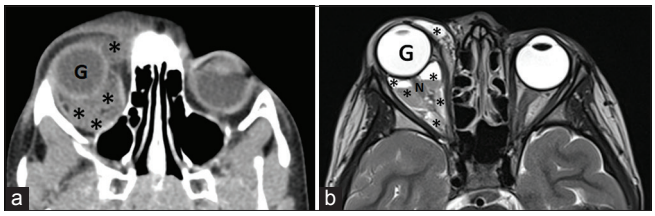

Figure 1: 6 year-old girl presenting with painless right orbital swelling. (a) Axial contrast-enhanced CT demonstrates a lobulated right orbital mass (*) displacing the globe (G) anteriorly with pre-septal and postseptal involvement. (b) Fluid sensitive MR shows multiple orbital cysts with fluid-fluid levels (*) that surround the optic nerve (N).

significantly decreased in size. Ultrasound (US) demonstrated a persistent posterior component surrounding the optic nerve, and treatment was elected given the potential for the malformation to enlarge in the future [Figure 2]. Given the visibility of the malformation on the US, the dominant LM cyst was accessed with a 21-gauge micropuncture needle (Cook, Bloomington, IN, USA) using a medial paraseptal approach [Figure 2]. Cyst fluid was aspirated, and a gentle digital subtraction angiogram was performed using Optiray 350 contrast (Guerbert, Villepinte, France) to exclude any venous or arterial communications with the cyst. The contrast was aspirated and approximately 1 mL of doxycycline hyclate (Fresenius Kabi, Bad Homburg, Germany) mixed in a 10 mg/mL concentration with Optiray 350 contrast was injected. Pre- and post-cone-beam CT of the orbit verified near-complete coverage of the malformation [Figure 3]. The patient was observed overnight and discharged the following day. She did not require any anti-inflammatory medications to reduce swelling. No complications occurred post-procedure and US 6 weeks later demonstrated a decrease in the malformation size. There were no episodes of recurrence at 7-month follow-up.

DISCUSSION

Lymphatic malformations comprise 1–8% of all orbital masses and are characterized as macrocystic, microcystic, mixed, and venolymphatic subtypes.[1,2] Macrocystic malformations are characterized by cysts >1–2 cm in diameter, while microcystic malformations have smaller cysts that may not be visible on the US. The venolymphatic subtype is frequently reported in orbital LMs given the common congenital origin of venous and lymphatic malformations from developing veins.[3] Microcystic and venolymphatic malformations are important to identify as they often require more treatment sessions and are less responsive to sclerotherapy.[4]

This child had a mixed, predominantly macrocystic, and LM with both pre-septal and retrobulbar components. Macrocystic LMs are multicystic lesions on US and MRI without internal flow or enhancement that usually has

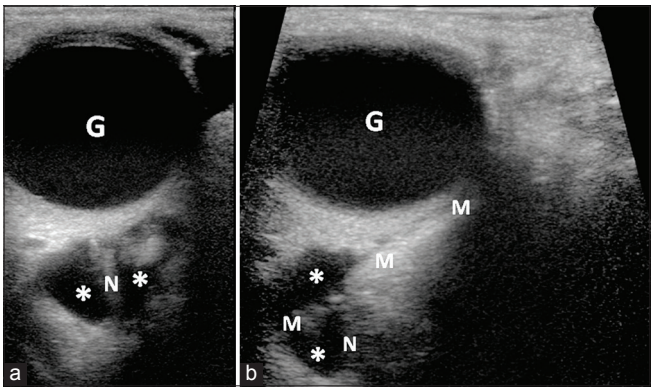

Figure 2: 6 year-old girl presenting with painless right orbital swelling. (a) Grayscale ultrasound of the orbit performed the day of sclerotherapy demonstrates a significant decrease in the size of LM macrocysts (*) with the dominant cyst surrounding the optic nerve (N). (b) Ultrasound-guided micropuncture needle (M) access of the LM cyst (*) using a medial approach (G = globe).ls (*) that surround the optic nerve (N).

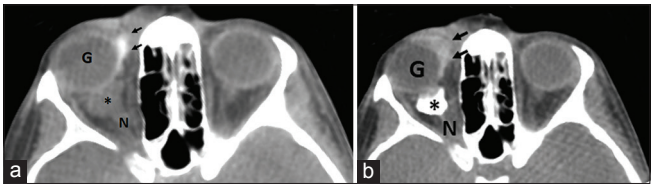

Figure 3: 6 year-old girl presenting with painless right orbital swelling. (a) Cone-beam CT demonstrates the small LM cyst (*) with sclerosant already injected (arrow) into the LM tissue medial to the globe (G). (b) Conebeam CT following completion of sclerotherapy shows adequate deposition of sclerosant (*) in the residual LM cyst (N = optic nerve).

fluid-fluid levels in the acute phase due to layering of hemorrhagic debris.

Interventional radiology treatment of orbital LMs involves the placement of a needle or drain into the largest cystic components, drainage of the cyst, digital subtraction angiography to document any cyst communication with other lymphatics or vessels, and injection of a sclerosing medication. The needle may be guided into the malformation using the US, fluoroscopy, and/or cone-beam CT with MR fusion capabilities.[5] Sclerosants used include doxycycline, bleomycin, OK-432, bevacizumab, sodium tetradecyl sulfate (STS), and ethanol.[2,6,7] Bleomycin and OK-432 have traditionally been preferred over other agents to reduce inflammation-related complications, although more potent medications such as STS may have similar visual acuity improvement (>75%) and complication rates when used in low concentrations (<2%) and volumes (<2 mL) at 1–2 year follow-up.[8]

Orbital LMs are treated on an urgent basis when acute swelling causes compartment syndrome and optic nerve

tension that can lead to blindness.[9] The treatment of the macrocystic components may be difficult in the non-acute setting when they are not distended. However, earlier drainage and sclerotherapy of macrocysts are now supported given the risks of recurrent swelling, vision loss, and chronic fibrosis.[4]

CONCLUSION

Our case illustrates the technical feasibility of semi-elective treatment for orbital LMs, which should be considered in the future to avoid long-term vision loss. Sclerotherapy should increasingly be considered a first-line treatment for orbital LMs.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

Dr. Joseph Reis is in the Editorial Board of the journal.

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