Case Report

Managing Recurrent Orbital Lymphangioma in a Pubertal Female with Negative Pressure Aspiration and Intralesional Bleomycin Injection: A Case Report

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Abstract

Introduction: Management of orbital lymphangioma is challenging. Complete surgical excision is often impossible due to its infiltrative nature. Sclerosing agents have been used in its management with variable outcomes. We report a case of recurrent orbital lymphangioma managed with intralesional bleomycin.
Case: A 14-year-old female presented with proptosis of the right eye for two weeks. She had a similar history at five years of age for which she underwent surgical excision. We performed negative pressure aspiration using a 20-gauge angiocatheter, injected bleomycin, and left the cannula in situ for repeat aspiration to maintain cyst collapse.
Observation: The lymphangioma regressed, and there was no recurrence at six months of follow-up.
Conclusion: This report highlights the use of negative pressure aspiration and intralesional bleomycin injection by minimal intervention using angiocatheter in the successful management of orbital lymphangioma.

Key words: Bleomycin, Case report, Lymphangioma, Orbit.

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Introduction

Lymphatic malformation (LM), previously termed as lymphangioma, are rare, benign congenital lesions of the lymphatic system which frequently occurs in the head and neck region, usually in children. They are composed of endothelial-lined lymphatic cysts, which may vary in size from a few millimetres to several centimetres (MacIntosh et al, 2014). Although they are found in all parts of the body, about 20% of LM involve the orbit and ocular adnexa. It accounts for less than 2% of orbital biopsies (Tunç et al, 1999).

Management of orbital LM is challenging. Complete surgical excision is often impossible and is incomplete because of its infiltrative

nature and chances of injury to surrounding vital structures. There is a high possibility of recurrence with incomplete excision (Lam and Yuen, 2019). Hence, nonsurgical methods such as intralesional sclerosing agents like Sodium tetradecyl sulphate, Ethanol, Bleomycin, OK-432 (Picibinil), Doxycycline, have been tried as an alternative or adjunct therapy in the management of orbital LMs (Gooding and Meyer, 2014).

Here, we report a case of orbital lymphangioma that had recurred after surgical excision but successfully treated with negative pressure aspiration and Intralesional Bleomycin Injection (IBI) using angiocatheter with minimal intervention. To our knowledge, this is the first case report of this form of management in orbital lymphangioma in Nepal.

This case report was prepared following the CARE Guidelines (Riley et al, 2017).

Case Description

A 14-year-old female presented with painless, forward bulging of her right eye (RE) for two weeks. It was sudden in onset and gradually progressive (Figure 1a). It was not associated with diminution of vision, diplopia, redness or discharge. There was no history of trauma.

On ocular examination, visual acuity was 6/6 in both eyes. Globe in the RE was pushed forward and laterally. Tenderness, pulsation, thrill, and periorbital changes were absent. There was no change in proptosis with a change in position or Valsalva. Hertel's exophthalmometry with a 100 mm base was 23 mm and 14 mm in the right and left eye, respectively. A mass (2 cm x 1 cm) could be palpated in infero-medial orbit, which was non-tender, soft, and non-pulsatile. Extraocular movement in RE was restricted medially. There was no lagophthalmos. The rest of the ocular examination was within normal limits. The examination of the left eye was normal. B-Scan ultrasound showed a cystic mass in the infero-medial and posterior orbit (Figure 2a). CT-Scan of the orbit was suggestive of lymphangioma or haemangioma (Figure 2b).

The same patient had presented to us nine years back, at the age of five years with a similar history. She underwent transconjunctival anterior orbitotomy with partial excision of the cyst. Histopathological examination of the excised mass confirmed the diagnosis of lymphangioma (Figure 1b). After surgery, she was asymptomatic until the current presentation. With this, the diagnosis of recurrent lymphangioma of the orbit was made.

A 20 G angiocatheter attached to a 10 ml disposable syringe was used to aspirate the cyst via the infero-medial eyelid approach where the mass was palpable under local anaesthesia with strict aseptic precautions, without attempting surgical excision this time. About 8.5 ml of hemorrhagic fluid was aspirated, and 8.5ml of injection Bleomycin (1 IU per ml of saline) was injected via the same cannula. Bleomycin was kept for 5 minutes and then aspirated out (Figure 3a). The angiocatheter was left in situ and secured with 4-0 Silk suture to maintain the collapsed state of the cyst and facilitate further aspirations on the next day (Figure 3b). Aspiration was done the next day from the same catheter, and it was removed after no fluid could be aspirated, on the third day.

At one month of follow-up, proptosis was completely resolved with minimal ultrasonographic detected cystic mass in the posterior orbit. At six months follow-up, cystic mass completely disappeared (Figure 4).

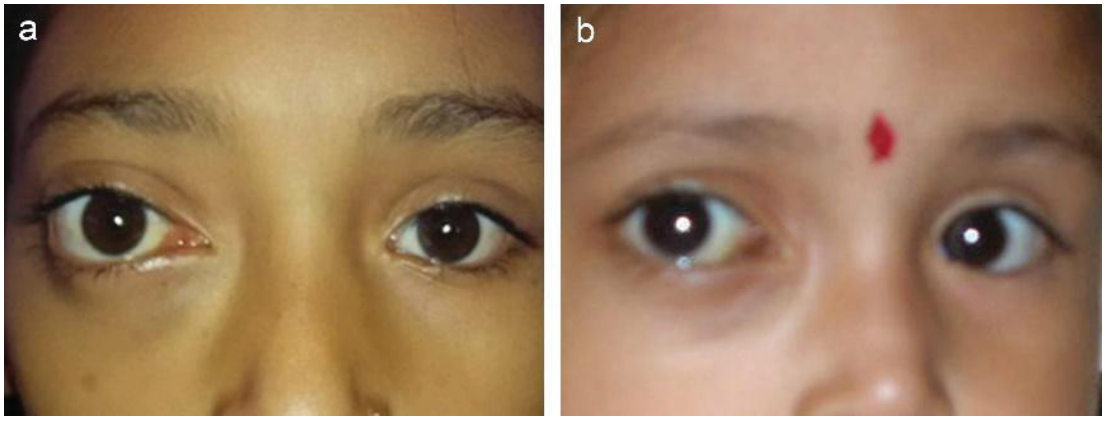

Figure 1: (a) Clinical picture (b) Clinical picture at 5 years of age

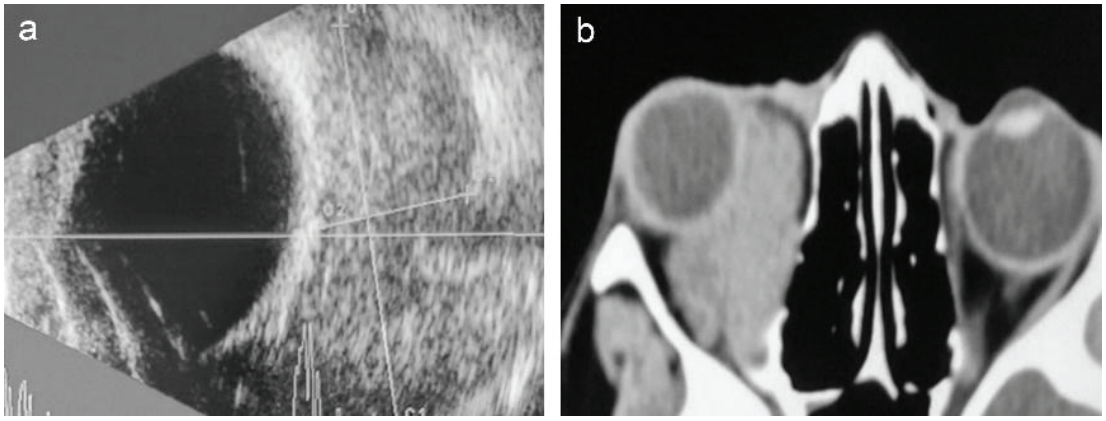

Figure 2: (a) USG-B Scan (b) CT-Scan

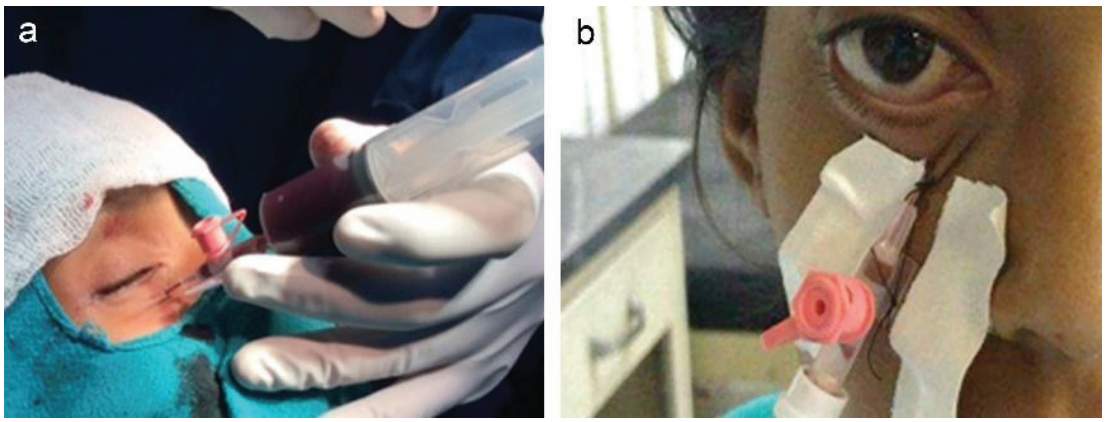

Figure 3: (a) Aspiration of hemorrhagic fluid by Angiocatheter (b) Angiocatheter left in situ

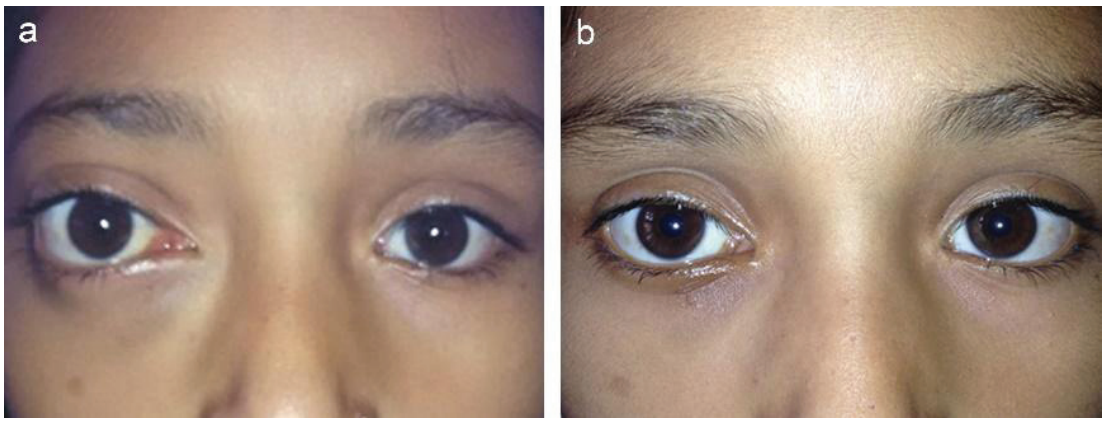

Figure 4: (a) Pre-operative (b) Post-operative Picture

Discussion

There are no published randomized control trials on the treatment of orbital lymphangioma. Thus, the effectiveness of medical or surgical treatment on orbital lymphangioma cannot be compared (Patel et al, 2019). Sclerotherapy has been popular in the past three decades in the management of orbital lymphangioma. These are tissue irritants that cause vascular thrombosis and endothelial damage leading to endofibrosis and vascular obliteration. Compared to surgery, they are less invasive and have faster recovery time (Lam and Yuen, 2019).

Bleomycin is an antitumor, an antiviral and antibacterial glycopeptide isolated first from *Streptomyces verticillus* in 1966 (Gooding and Meyer, 2014). It is currently used as a sclerosing agent. The use of bleomycin in the management of orbital LMs has been reported in the literature but has not yet been reported from Nepal.

Gooding and Meyer (2014) reported four cases of orbital lymphangioma not responding to intralesional steroid, bevacizumab, and external beam radiotherapy but responded positively to IBI. In all cases, no attempt to aspirated fluid was made, and no negative pressure was applied. Repeat injection was required in most cases. In a retrospective series of 22 cases by Paramasivam et al (2014), 16 were treated with IBIs, and 57% had >80% volume reduction. Raichura et al (2017) conducted a prospective study in 13 patients with IBIs, with most cases having a 60% reduction in diameter of the lesion at final follow-up with 2-3 injections of bleomycin. Sen et al (2019) also reported a similar case, which was managed by aspiration of cyst and injection of intralesional bleomycin.

Woo et al (2017) reported outcomes of 12 cases of orbital lymphangioma, five of which were treated with aspiration of blood, intralesional injection of a sclerosing agent, and continuous negative pressure application after incision. Lee et al (2015) in Korea successfully managed a case of LM with IBI after surgical incision, and a drain was placed in the incision site to provide negative pressure to induce cyst shrinkage and control bleed. There was no recurrence at 6 months of follow-up.

Our patient is a case of recurrent orbital lymphangioma. She underwent cyst negative pressure aspiration with IBI and repeat aspiration to maintain cyst collapse with angiocatheter placed in situ. We report no recurrence at the 6 months follow-up though a longer follow-up is required to see the treatment efficacy regarding the recurrence.

Conclusion

IBIs can be a useful option in the management of recurrent orbital lymphangiomas. Using angiocatheter to aspirate lymphangioma, inject sclerosant, and repeat aspiration to ensure cyst shrinkage, can be used safely and effectively with minimal intervention.

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