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Bleomycin Sclerotherapy In A Case Of Recurrent Lymphaticovenous Malformation

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Abstract:
Orbital lymphaticovenous malformation (LVM) is a lymphatic and venous system disorder that presents in childhood . There are multiple approaches to management of this lesion. Sclerotherapy is used to treat and shrink the lesion prior to or as an alternative to surgery. It minimises the amount of surgical management or the need for surgery. Several sclerosants are now commonly used to treat these lesions- Bleomycin, sodium teteradecyl sulfate, ethanol, doxyciline.
Orbital LVM , unencapsulated vascular malformation of the lymphatic system (composed of irregular vascular channels lined with a single layer of attenuated endothelial cells), primarily seen within the first decade of life[1]. Traditional management through surgery can be challenging. This can result in partial resection, with risk of recurrence, scarring or injury to important adjacent structures[1,2]. Sclerotherapy has been reported as an alternative to surgery [3].

Key-words:*Lymphaticovenousmalformation , bleomycin , sclerotherapy*

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I. Introduction:

Orbital lymphangioma have a prevalence of 1.1 to 5.3 cases per 10,000 live births with an equal incidence between males and females [9]. It presents in young age. Surgery is not curative as they are intertwined within the normal tissues within the orbit.This case presented to us in adolescence after undergoing 2-3 surgeries for the same in his childhood. Sclerotherapy may minimize the amount of surgical management or prevent need for surgery[3].

II. Case History:

14 year old male came to ophthalmology OPD with chief complains of redness , pain in left eye associated with lid swelling and no vision since childhood. He was a previously operated case of left frontal orbitotomy for left orbital lymphangioma with cavernous hemangioma in 2005 and subtotal excision was done for the same, got operated for left orbit hemangioma in 2007 and 2009 as well. Patient was operated for recurrent in february 2019. Examination of the affected eye includes proptosis, edema and hypertrophy of upper lid, subconjunctival hemorrhage and chemosis associated with corneal opacity. Extraocular movements were restricted in all directions of gaze. The other eye was normal with visual acuity of 20/20 on alphabet snellen's chart.

MRI Orbit + brain, plain + contrast shows- (Figure 1 & 2: T2 weighted axial and STIR coronal orbital MRI images before the procedure respectively)

Ill defined lobulated STIR/T2 hyperintense lesion which is hypointense on T1 weighted images in intraconal and extraconal compartment of left orbit of 7.8x6x4 cm in superoinferior, anteroposterior and transverse dimensions suggestive of residual / recurrent lymphangioma. Other changes s/o optic nerve atrophy. Small T2 hyperintense lesion with peripheral hypointense right gangliocapsular region suggestive of cavernoma.

Image guided bleomycin sclerotherapy was performed in radiology suite of the hospital . Dynamic ultrasound was used for image guidance. Bleomycin was reconstituted 0.5 IU /kg body weight , given with 2% lignocaine transcutaneously , under short sedation. Multiple injections given in the lesion , prior to which the intralesional contents were aspirated , a few were dry , few had blood.

Repeat MRI Brain + orbit (Plain + contrast) done 40 days after the procedure (Figure 3 & 4: T2 weighted axial and STIR coronal orbital MRI images after the procedure respectively) suggestive of ill defined lobulated lesion STIR/T2 hyperintense and T1 isointense with mild heterogeneous post contrast enhancement in the intraconal and extraconal compartment of left orbit which measures approximately 7 X 4.8 X 4 cm in superoinferior , anteroposterior and transverse dimension. Small T2 hyperintense lesion with peripheral

hypointense signal noted in right gangliocapsular region measuring suggestive of cavernoma.

Clinical examination of the left eye shows decrease in lid edema , subconjunctival hemorrhage and chemosis with significant reduction in proptosis.(Figure 5: clinical picture before and after the procedure)

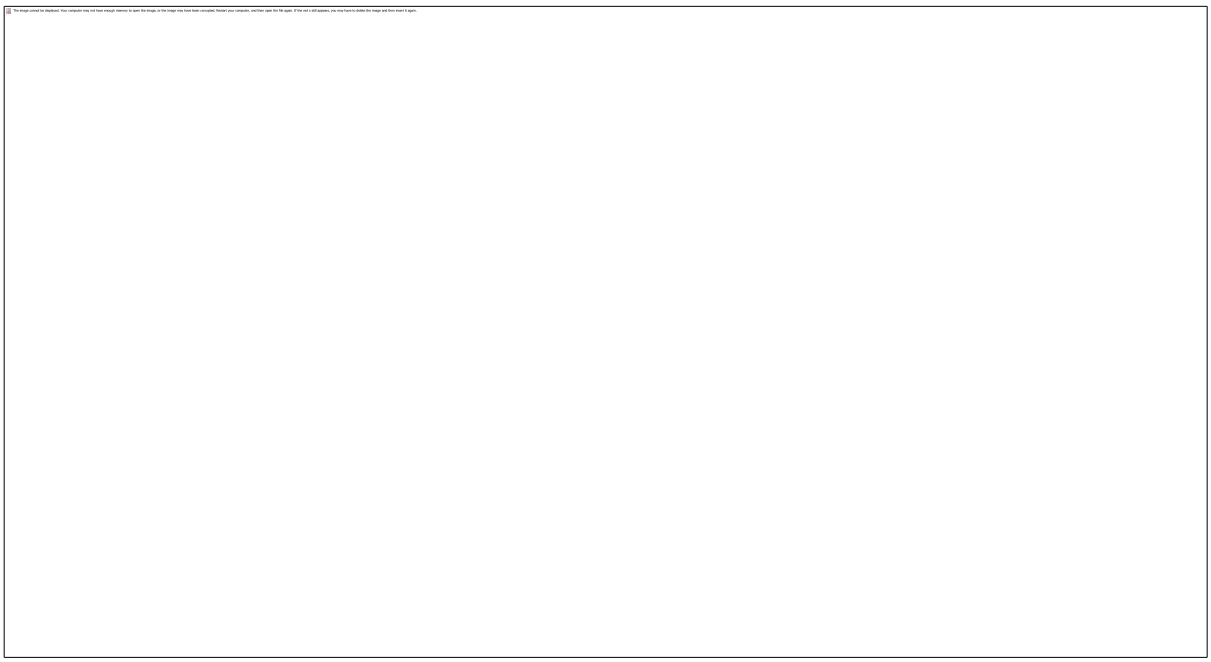

Figure 1 &2 :T2 weighted axial and STIR coronal orbital MRI images before the procedure

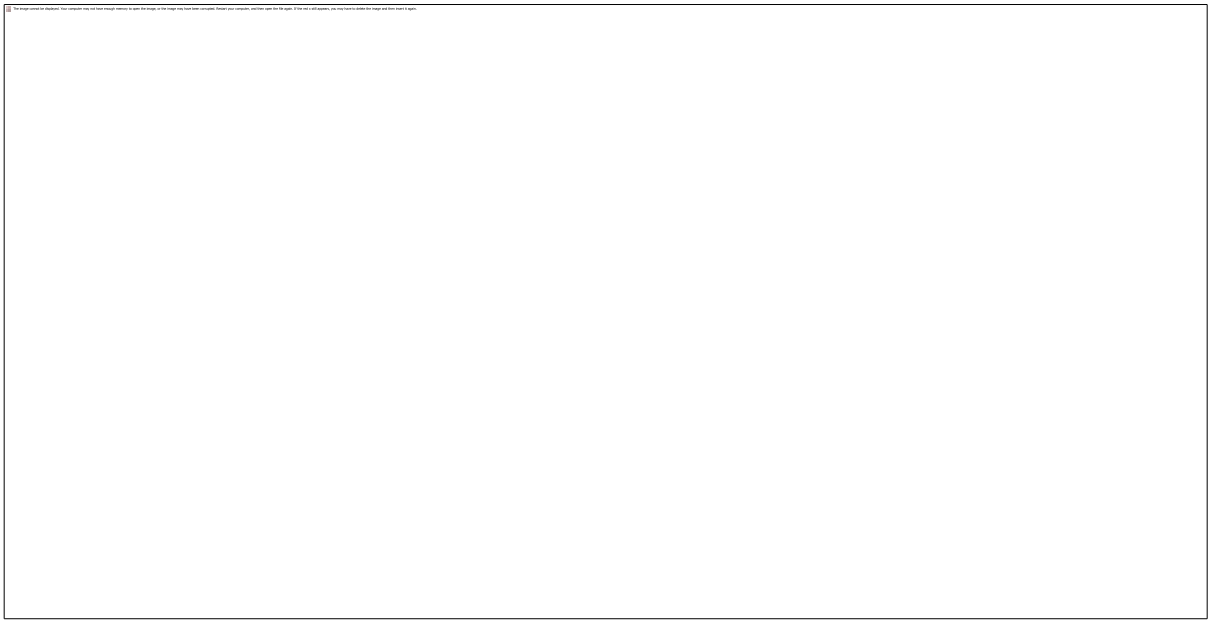

Figure 3 &4 :T2 weighted axial and STIR coronal orbital MRI images after the procedure

Figure 5 : clinical picture before and after the procedure

III. Discussion:

Approximately 25% of lymphangioma cases involve the orbit[2]. The reported signs on presentation include progressive proptosis (axial or non-axial), ptosis, ophthalmoplegia, and strabismus[2,3]. Thesymptomsincludepain, diplopia, and visual loss[4]. Intra-orbital haemorrhage is considered to be a common cause of exacerbation of proptosis. Such haemorrhage is spontaneous, or after local/systemic infection, or local trauma[2,5,6].

Orbital lymphangiomas are currently classified as Type 1 (low flow and no vascular system connection) orbital vascular malformations by the International Orbital Society. Any lesions that are found to have any vascular system connection fall under Type 2[7] .

Surgical excision is challenging, since orbital lymphangiomas are unencapsulated. Thus, in some cases only partial excision is performed to debulk the lesion and reduce it's mass effect[1,7]. The risk of intra-operative haemorrhage is significant, but can be reduced using diathermy, or carbon dioxide laser[5,7]. Boulos et al. have reported injecting fibrin glue into cystic components of lesions during excision to stabilise the structures being removed. Sclerosants have been used for treating lymphatic malformations for several years. Agents that have been previously reported as being used for orbital lymphangioma include OK-432, sodium tetradecylsulfate , 5% sodium morrhuate, and bleomycin A5[10,12,13].

Percutaneous drainage and ablation of cystic components has also been advocated for orbital lymphangiomas, using a combination of intra-cyst injections of either sequential sodium tetradecylsulfate and ethanol for macrocysts (>1 cm), or doxycycline for microcysts (<1 cm)[15] .

Management of orbital lymphangioma continues to require a multi-disciplinary approach, and early involvement of other specialities when needed can prevent delays in arranging imaging or treatment. It is not unusual nowadays for treatment to comprise of both surgical and non-surgical aspects in order to remove/shrink as much of the lesion as possible while minimising visual and functional loss. It is important to remain in frequent communication with the patient and/or their parents, as well as colleagues, to ensure a consistent message and plan of action. What is clear is that sclerotherapy is now an established management option for these lesions, with good results (and minimal complications) in appropriately chosen patients[8].

Sclerotherapy is an umbrella term that characterizes the multiple types of agents that are injected (usually under ultrasound guidance) into the cystic spaces of the lesion, leading to scar formation and reduction in the size of the cyst and lesion. Few agents used in orbital lymphangiomas include OK-432 (Picinibil), sodium tetradecyl sulfate, doxycycline, ethanol, pingyangmycin, and bleomycin.

It is important to note that sclerotherapy has been reported to be effective in treating macrocystic lymphatic malformations (size >2cm), with less efficacy in microcystic malformations[18]. A 2016 prospective interventional study looked at 29 patients with macrocystic orbital lymphangiomas who underwent sclerotherapy injection with sodium tetradecyl sulfate. At an average follow-up time of 21.8 months, all patients showed a reduction in maximal lesion diameter of 50% or greater, with improved visual acuity in 78.2% and complete radiological resolution in 51.7%[19]. Similarly, a 2017 prospective interventional studied looked at 13 patients with orbital lymphangiomas who were underwent sclerotherapy injection with bleomycin. With an average follow up time of 19.69 months, 92% of patients showed a >60% reduction in the maximal lesion diameter[20] .

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