Image-Guided Percutaneous Sclerotherapy for Orbital Low-Flow Malformation: Our Experience

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Abstract

For a safe sclerotherapy session to be completed in the orbital low-flow malformation (namely lymphovenous malformation or venolymphatic malformation), accurate identification of the target lesion for the drug injection is crucial. Regarding the dependability and viability of the injection approach, the authors have discussed their experiences with image-guided percutaneous sclerotherapy on a few patients.

Keywords

- sclerotherapy
- orbital
- low-flow malformation
- image-guided
- LVM
- fluoroscopy

Introduction

Image-guided percutaneous sclerotherapy in low-flow orbital malformations is the preferred treatment option. Imaging is required for confirmation of the diagnosis of low-flow vascular malformation (namely, LVM, lymphovenous malformation, or VLM, venolymphatic malformation), for seeing its extent, and for planning the treatment. Image-guided intervention helps us achieve accurate needle placement, which is critical for good results. Yet there is no established protocol for low-flow orbital malformation especially if "atypical" or "poorly defined." Herein, we propose using multimodality imaging for diagnosis and treatment in cases of low-flow orbital malformation, with discussion focused on the choice of imaging and its feasibility. All the cases were prospectively studied between December 2019 and December 2022 in compliance with the Declaration of Helsinki (►**Table 1**).

Case Description

Case 1, ►**Fig. 1**: A 8-year-old male child patient presented with right-sided sudden painful proptosis preceded by a small bluish swelling in the lower eyelid in the past 1 week. Frozen globe in presence of "no PL (perception of light)" was present, which could be alarming signs of delayed referral. B-scan ultrasonography (USG) and computed tomography (CT) scan revealed two large cysts (36 and 22, mm) in the right deep orbit indicating chocolate cyst formation in the macrocystic LVM causing orbital compartment syndrome. Urgent right orbital cyst aspiration with bleomycin injection (15 U bleomycin vial mixed with 4.5 mL normal saline, 1 mL lignocaine, and 1 mL iohexol in a typical ratio of drug: lignocaine: contrast, 3–5: 1: 1; drug concentration 2.3 U/ mL; total injectable volume per sitting, 4 mL = 9.2 U; dose, 0.5–1 U/ kg, maximum 10 U or 4 mL for orbit per sitting) guided by Doppler-USG and fluoroscopy,

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Table 1 The cases of sclerotherapy in brief

Sl. no.	Age (year), Sex	Low-flow malformation				Preinjection imaging workup	Sclerotherapy (bleomycin)					Adverse effect apart from transient inflammation
		Nature[1]	Location[1]	Complaint	Duration		Indication	Image guidance	Confirmed needle tip inside the target lesion (not blind)	Complete filling of the lesion with the drug was confirmed by postinjection CT	Sittings	Clinical outcome	
1	8, male	Macrocystic LVM	Orbit, deep	Sudden painful proptosis	1 week	USG B-scan, CT	Orbital compartment syndrome	Doppler-USG, fluoroscopy	Yes	Yes	2	Marked resolution	No
2	50, female	Macrocystic LVM	Orbit, deep	Sudden painful proptosis	1 day	USG B-scan, MRI	Orbital compartment syndrome	USG B-scan	Yes	No (CT was not done)	1	Complete resolution	No
3	16, female	Microcystic LVM	Orbit, complex	Eyelid lump	More than 10 years	Doppler-USG, CT, MRI	Cosmetic	Doppler-USG, CT	Yes	Yes	1	No change	No
4	21, male	VLM	Orbit, combined	Eyelid lump	8 years	Doppler-USG, CT, MRI, MRA	Cosmetic	Doppler-USG, CT	Yes	Yes	1	No change	No

Abbreviations: LVM, lymphovenous malformation; CT, computed tomography; MRI, magnetic resonance imaging; USG, ultrasonography; VLM, venolymphatic malformation.

under general anesthesia (GA) was performed in two sittings separated by an interval of 48 hours with the aspirated blood volume, 7 and 3 mL, respectively. The vision could not be salvaged despite obliteration of the cyst, and was recorded as "no perception of light." The fluoroscopy record ruled out the possibility of sclerotherapy-induced vascular occlusion (►**Video 1**), and therefore, the vision loss was attributed to compressive optic neuropathy as a result of the compartment syndrome. No recurrence was noted over 2 years follow-up.

Video 1

Fluoroscopy confirming intracystic injection of the drug. No intravascular contrast was seen. Online content including video sequences viewable at: https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0044-1779689.

Case 2, ►**Fig. 2**: A 50-year-old female patient presented with left-sided sudden painful severe proptosis (might be labeled as globe luxation), without any systemic abnormality. Imaging revealed a clear, large retrobulbar cyst (45 mm) with fluid–fluid level likely chocolate cyst formation in a macrocystic LVM. Urgent "blind" but B-scan-assisted cyst aspiration with bleomycin injection (4 mL, 9.2 U similar to case 1) was performed without GA. Postinjection CT could not be performed as the patient refused. On follow-up, there was no reported complication or recurrence.

Case 3, ►**Fig. 3**: A 16-year-old female patient presented with left sub-brow lump with mechanical ptosis, more toward medial aspect since early childhood. According to the parents, debulking of the lump had been attempted by a general surgeon at the age of 6, though no surgical detail was available. Since then, it has showed no change in size or color. Best-corrected visual acuity (BCVA) was 20/20 in both eyes with orthotropia. Hertel's readings showed left mild proptosis (right, 14; left, 16; base, 100 mm), with full extraocular movements, without any resistance to retropulsion or any sign of high flow like pulsation or bruit. On slit-lamp examination, ill-defined, dilated lymphatic, and venous channels along with a chocolate-colored cyst over the medial conjunctiva were noted. Neither the lump nor the conjunctival lesion showed any change on Valsalva or posturing. She was somewhat unaware of the conjunctival lesion. Arterial flow was ruled out by Doppler-USG and CT imaging. A working diagnosis of left orbital complex, low flow, microcystic LVM lesion was made. Bleomycin sclerotherapy under Doppler-USG and CT guidance was performed without GA. Over a year follow-up, no visible change in the lesion or adverse effect was seen.

Case 4, ►**Figs. 4**–**5**: A 21-year-old male patient presented with right lower lid lump, noticed for the last 8 years. The lump as well as globe was showing mild changes with posture and Valsalva, suggestive of minimally distensible, preseptal and some postseptal venous components. On slit lamp, the adjacent fornix and bulbar conjunctiva showed dilated and tortuous pinkish–purple dysplastic veins. On MRI, the lesion

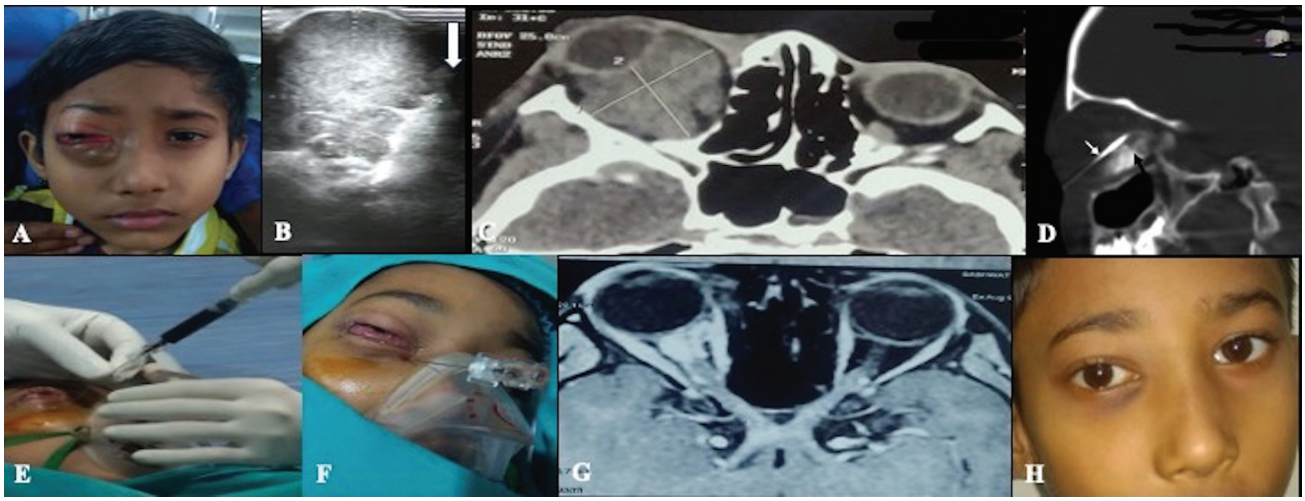

Fig. 1 (**A**) Right eye proptosis with periorbital swellings in a 8 years old child; (**B**) on ultrasonography, multiloculated cysts with homogeneous low level internal echoes were seen in the inferior orbit, in an anterior to posterior (*arrow's direction*) view; (**C**) noncontrast computed tomography (CT) scan of the orbit showed multiple cysts in the intraconal compartment tenting on the right globe causing proptosis; (**D**) intraprocedural CT scan sagittal view showing exact localization for the needle placement (*white arrow*) while aspirating the cyst (*black arrow*); (**E**) dark venous blood on aspiration; (**F**) immediately after the aspiration with the needle in situ, decrease in proptosis and periorbital swelling was seen; (**G**) axial T1 postcontrast magnetic resonance imaging at a month follow-up showed residual small enhancing lesions close to the right optic nerve, which was left untreated; (**H**) Stable, residual dystopia of the right globe at 1-year follow-up.

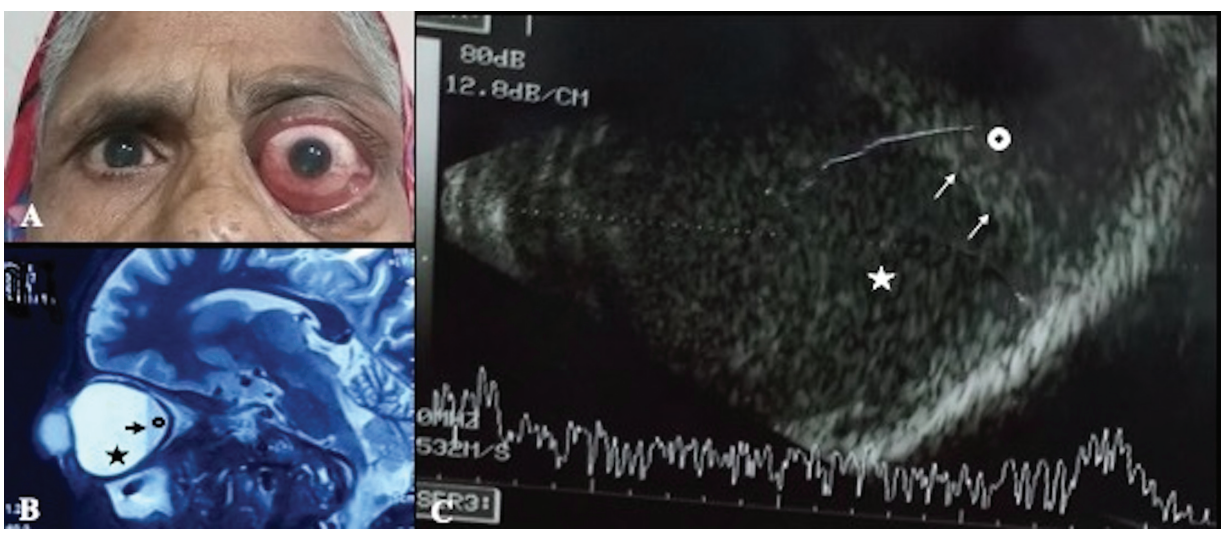

Fig. 2 (**A**) Left eye proptosis; (**B**) T2 sagittal magnetic resonance imaging showed a large, hyperintense cyst with fluid (*star*)–fluid (*doughnut*) level (*arrow*) in the retrobulbar, intraconal space of the left orbit; (**C**) ophthalmic ultrasonography showed a highly reflective membrane (*arrows*) likely due to fluids (*star*; *doughnut*) interface (as seen in ►**Fig. 1B**).

(24 mm in maximum dimension) displayed T1 hypointense signal intensity, T2 hyperintense signal intensity with internal septation, and fluid–fluid level like lymphatic macrocyst. No evidence of flow void within the lesion to suggest arterial flow. Magnetic resonance angiography (MRA) showed no aberrant arterial or venous communication of the lesion. Apart from a bright area seen on contrast enhancement localized to temporal part of the bulbar component, rest of the lesion displayed no enhancement, suggestive of combined VLM. BCVA was 20/20 and extraocular movement was full in both eyes. Rest clinical examination including retropulsion and auscultation was unproductive. After 1.2 mL of dark blood aspiration from the cystic component, bleomycin sclerotherapy under Doppler-USG guidance was performed without GA. Postinjection CT images were obtained. No clinical change or adverse effect has been observed till the last follow-up at 6 weeks.

Discussion

Preprocedure detailed clinical workup and imaging, namely Doppler-USG, multiphasic dynamic MRI, if needed MRA, magnetic resonance venography (MRV), or CT, are crucial for classifying and prognosticating the lesion for planning sclerotherapy as well as tracing aberrant vascular communications.[1,2] Case 1 was a pediatric emergency procedure performed in a digital subtraction angiography (DSA) laboratory under fluoroscopy, while case 2 was an adult emergency procedure performed in an ophthalmology-OT. Case 3 and 4 were adult

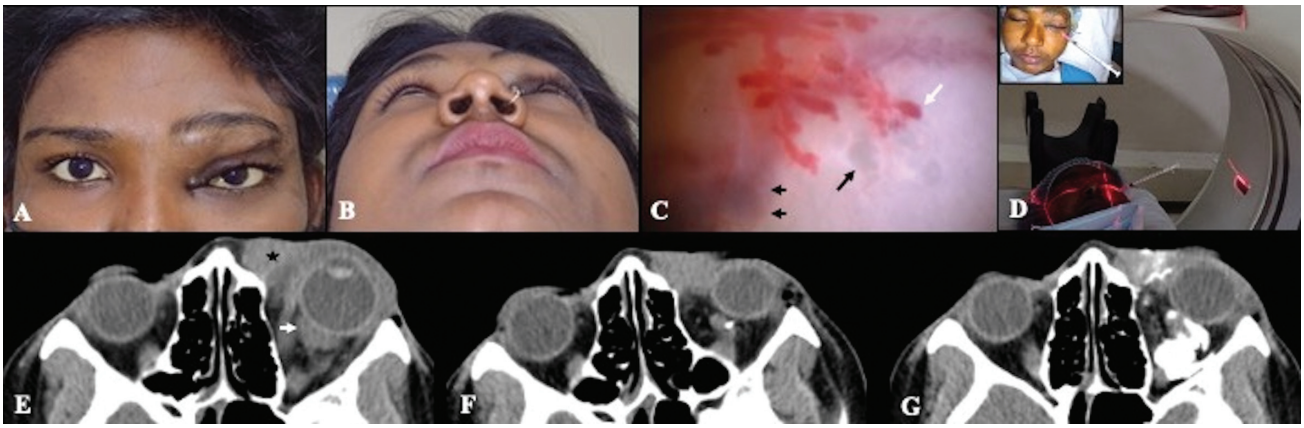

Fig. 3 (**A**) Left sub-brow swelling, orthotropic; (**B**) left eye mild proptosis in the worm's eye view; (**C**) slit-lamp photograph showing dilated lymphatics (*black arrow*) in the bulbar conjunctiva, partly filled with venous blood (*white arrow*), and a possible underlying chocolate cyst medially (*arrowheads*), representing dysplastic venolymphatic channels; (**D**) computed tomography (CT)-guided needle placement prior to aspiration (inset, 1cc syringe with 24 G needle in situ); noncontrast CT orbit, axial view showing, (**E**) pre- (*star*) and postseptal, diffuse cystic lesion. A globular retrobulbar component (*arrow*) molding along the sclera below the optic nerve was also seen; (**F**) confirmed needle-tip position (*white dot*) targeted the retrobulbar component before the drug injection; (**G**) complete filling the retrobulbar lesion immediately after the drug injection with very little extravasation. Note the irregular preseptal enhancement caused by the multiple intralesional punctures for the sub-brow lesion.

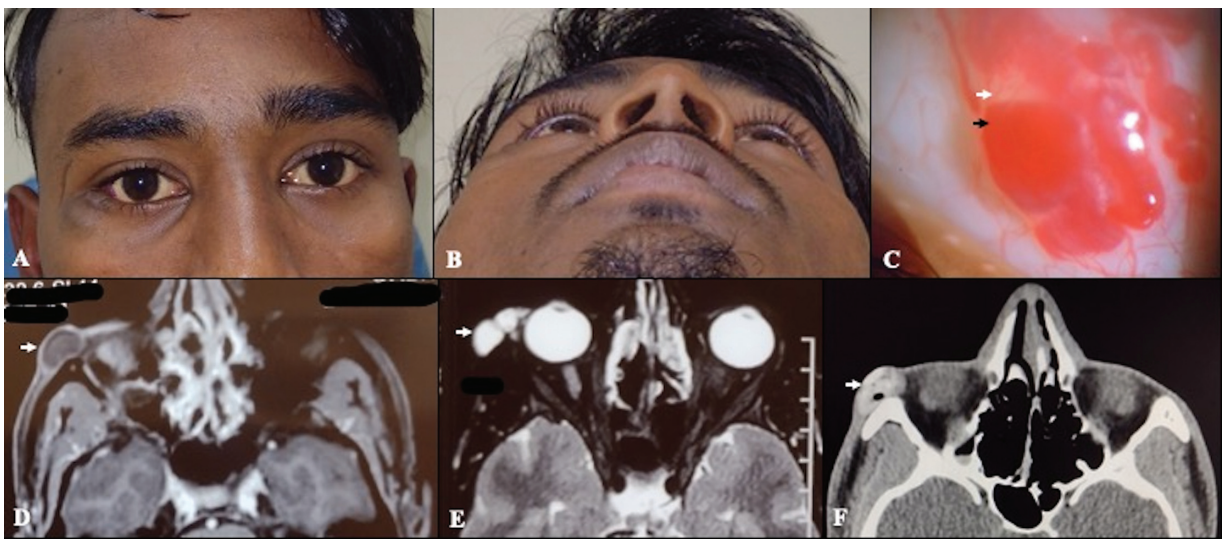

Fig. 4 ((**A**) Right lower lid swelling; (**B**) on worm's eye view with chin up, slight reduction in the swelling with no or slight enophthalmos was noticed; (**C**) stagnated blood (*black arrow*) level separated from the clear supernatant lymph (*white arrow*) in the abnormal, distended conjunctival vessels; axial magnetic resonance imaging (MRI) showing well-defined preseptal cystic lesion (*arrow*), (**D**) nonenhancing and appearing hypointense on postcontrast fat saturated T1-weighted MRI, and (**E**) hyperintense with internal septa on T2-weighted MRI; (**F**) computed tomography orbit, axial postprocedure showed complete contrast filling of the lesion (*arrow*) with an air bubble (black shadow) close to the entry side.

elective procedures performed in a general CT room primarily under CT and USG guidance, respectively. Indications for the treatment vary from progressive compressive symptoms in nondistensible venous or LVMs (case 1 and 2) to cosmesis in distensible venous or venolymphatic (case 3 and 4).[1] Though a DSA-laboratory well-equipped for fluoroscopy guided procedures is best suited for orbital or periorbital sclerotherapy, it might not be always feasible due to logistic reasons.[3] Alternatively, intraprocedural on-site CT could be used; though cumbersome and less safe, it is sometimes manageable. We believe intraprocedural preinjection CT or real-time fluoroscopy is needed only when the lesion is deep-seated, as in the first two cases, or in cases where an "atypical" or "poorly defined" low-flow lesion is present, while postinjection CT (optional) should be done in all lesions to confirm complete filling of all the targeted lesions. In all the four cases, real-time USG with doppler was used, which we think is the most dependable modality to localize the lesion, to choose the most accessible least vascular plane, to guide the needle into the lesion before aspiration and to track the drug injection. Easy availability, lack of dependency on DSA-laboratory or CT-laboratory, portability, no radiation, and cost-effectiveness are the qualities due to which Doppler-USG is preferred over fluoroscopy or CT. As the rare but possible risk of retinal vascular occlusion could not be ignored, we were not so much confident about safety of a "blind" injection without the Doppler-USG except in selected cases of large, pure lymphatic

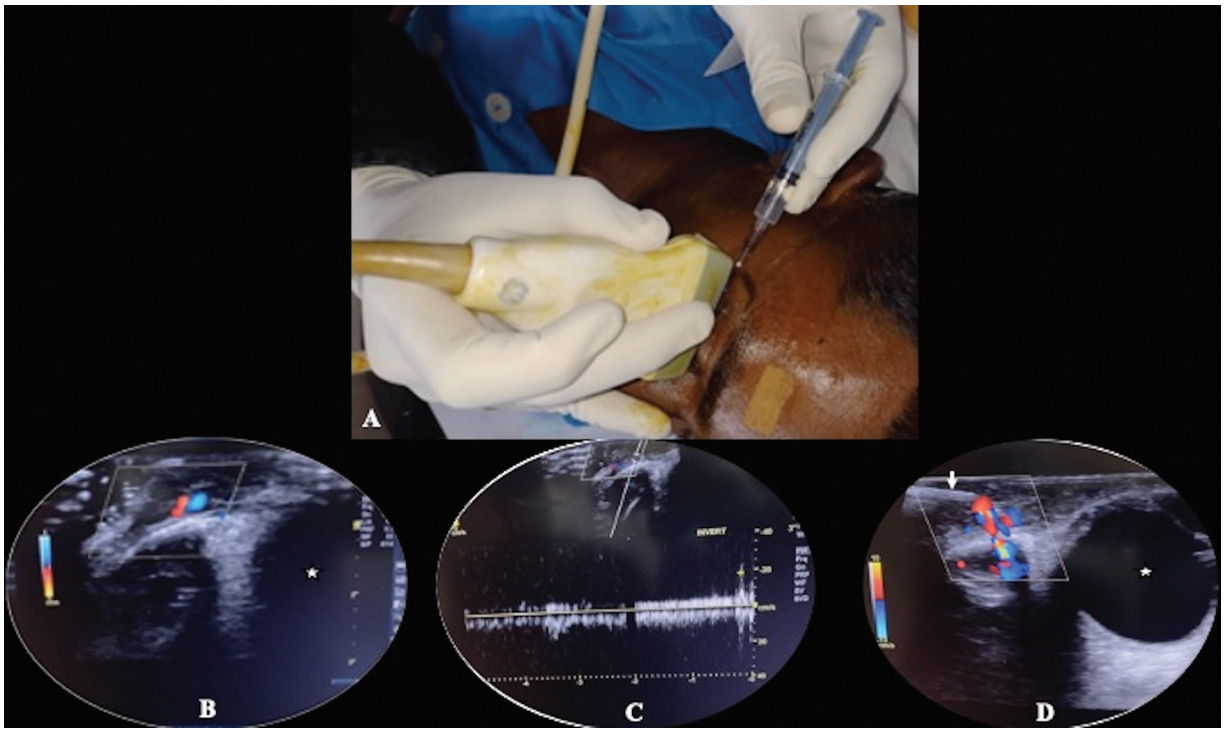

Fig. 5 Intraoperative color Doppler ultrasonography showing (**A**) position of the probe and the needle during injection; (**B**) vascularity within the lesion separate from the eyeball (*star*), (**C**) with venous flow pattern, (**D**) intralesional drug injection (*arrow*, needle tip inside the cyst) at a safe distance from the eyeball (*star*).

cyst not encasing optic nerve like case 2, in a method described by Chen et al.[3,4]

Irrespective of the imaging modality used, the response to the therapy, however, depends upon many factors like the predominant type of dysplastic vessels, depth and extent of involvement, size of cysts, its duration, their localization, accessibility for the needle aspiration, drug efficacy, repeat procedure, previous surgery, technical expertise of the treating physician, and the intervention radiologist.[4] Lesions that are less lymphatic or more venous, complex, or compound, microcystic (< 1 cm), long-standing or postpubertal, lesion abutting the optic nerve or orbital apex, previously operated, and have no response to previous sclerotherapy are poor candidates for sclerotherapy, as seen here.[5] In cases of no response, we might review or repeat the imaging to accurately categorize the lesion, which could be an important error as highlighted in the meta-analysis by De Maria et al.[4] Accordingly, we might plan a different or more potent intralesional agent, such as bevacizumab, doxycycline, ethanolamine oleate, OK-432 (Picibanil), or sodium tetra decyl sulfate in case 3, and percutaneous embolization with n-butyl-cyanoacrylate in case 4, followed by surgical debulking.[5,6]

In venolymphatic cases without prior sclerotherapy attempt, where no communication with orbital veins was obvious, one might plan percutaneous embolization. Additionally, before embolization, one might rule out potentially dangerous tributary by direct percutaneous intralesional injection of contrast with lignocaine and taking serial CT. Otherwise, endovascular embolization is the safer option left for cases where such communications exist.[2,7]

Conclusion

Sclerotherapy modality selection is based on lesion location (deep or superficial), patient age, physician preferences, accessibility, and urgency. As such, results may differ, and since there are only a few cases, we are unable to rely solely on one imaging modality in this situation. Therefore, we think that multimodal imaging with Doppler-USG provides a better localization of the target lesion and the needle tip for effective drug delivery.

Patient Consent

An informed consent was taken from the parents of the patient for publishing their data.

Ethical Approval

This study complies with the Declaration of Helsinki and CARE guidelines.

Authors' Contributions

G.L., Sh.K., Su.K., N.N., and N.G. prepared and edited and reviewed the manuscript. G.L. helped in data collection and literature search. G.L., Sh.K., Su.K., N.N., and N.G. helped in approval of the manuscript .

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Conflict of Interest

None declared.

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