Orbital infarction syndrome after multiple percutaneous sclerotherapy sessions for facial low-flow vascular malformation: A case report and literature review

Weng Sut Sio, Shwu-Huey Lee, I-Chia Liang

Vision loss following sclerotherapy for facial vascular malformations (VMs) is a rare but detrimental complication. Here, we report a case of an 11-year-old boy with acute onset blepharoptosis, ophthalmoplegia, and blindness in his right eye after the 14th sclerotherapy session (percutaneous intralesional injection of sodium tetradecyl sulfate) for a right facial low-flow VM without orbital involvement. Computed tomography angiography revealed no contrast enhancement in the right ophthalmic artery, superior ophthalmic vein, or extraocular muscles. He presented with the hallmarks of orbital infarction syndrome: Clear signs of anterior and posterior segment ischemia and disrupted arterial flow to the extraocular muscles. His blepharoptosis and eye movement improved 4 months later; however, he remained blind, and phthisis bulbi developed eventually. Thus, sclerotherapy for facial VM—even without orbital involvement—may result in severe ocular and orbital complications.

Key words: Low-flow vascular malformation, orbital infarction syndrome, sclerotherapy, sodium tetradecyl sulfate

Vascular malformations (VMs) are congenital structural abnormalities of the vascular system. Percutaneous sclerotherapy is a simple and effective therapeutic option for low-flow VMs (LFVMs).[1] Ocular complications have not been noted after percutaneous sclerotherapy for head and neck LFVMs without orbital involvement.[2] Here, we describe a patient with facial LFVM without orbital involvement who developed vision loss, blepharoptosis, ophthalmoplegia, and eventually phthisis bulbi after the 14th sclerotherapy session.

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Case Report

An 11-year-old boy was diagnosed with the right-sided facial LFVM when he was 5 years old, with an initial presentation of progressive swelling. He had received 13 sclerotherapy sessions over the previous 4 years without any adverse events. The LFVM gradually decreased in size after sclerotherapy [Fig. 1]. Because the swelling had almost dissipated, the patient was scheduled for the final (14th) sclerotherapy session. On day 1 of the session, 3% sodium tetradecyl sulfate (STS; 2 mL) was injected intralesionally under ultrasound guidance and intravenous general anesthesia. When he became alert, the patient complained of severe right-sided periocular pain and an inability to open the right eye. Further, physical examination showed abnormal pupillary light reflex in the right eye. The ophthalmology department was consulted because of his ocular symptoms.

His best-corrected visual acuities were no light perception and 20/20 in the right and left eyes, respectively. Ecchymosis patches were present on his right upper eyelid and frontal area. The patient had complete blepharoptosis with total ophthalmoplegia of the right eye [Fig. 2a]. The intraocular pressure was 11 and 10 mmHg in the right and left eyes, respectively. Slit-lamp examination of the right eye revealed pale and cyanotic conjunctiva, a clear cornea, a deep anterior chamber, and a clear lens with a fixed and dilated pupil. Posterior segment examination of the right eye showed disk edema with hyperemia, venous dilatation, arteriolar narrowing, and a relatively pale retina with no cherry red spot. In both examinations, the left eye was normal.

Orbital infarction syndrome (OIS) was suspected, and oral aspirin (200 mg), intravenous heparin (1650 U), and methylprednisolone (20 mg) were administered. However, the right eye remained blind. On the next day, slit-lamp examination revealed signs of anterior segment ischemia with marked hypotony, corneal edema, and an anterior chamber with inflammatory reaction in the right eye. Computed tomography angiography showed no contrast enhancement in the right ophthalmic artery (OA), superior ophthalmic vein (SOV), or extraocular muscles [Fig. 3]. The right eye had a mildly engorged SOV and mildly swelled extraocular muscles. Fluorescein angiography could not be performed because of severe corneal edema. During the 1st week, the right eye showed aggravated ischemia with conjunctival chemosis, a total corneal epithelial defect, and a collapsed anterior chamber. On day 7, the corneal epithelium began regenerating, but the ischemia of the periorbital region remained severe. To improve periorbital

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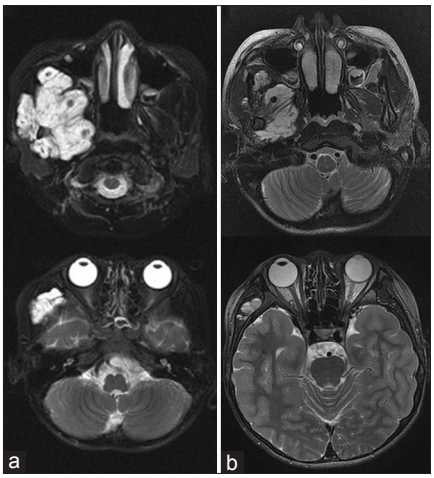

Figure 1: Magnetic resonance images of the right facial vascular malformation before (a) and after (b) sclerotherapy. Note that the lesion did not involve the orbit

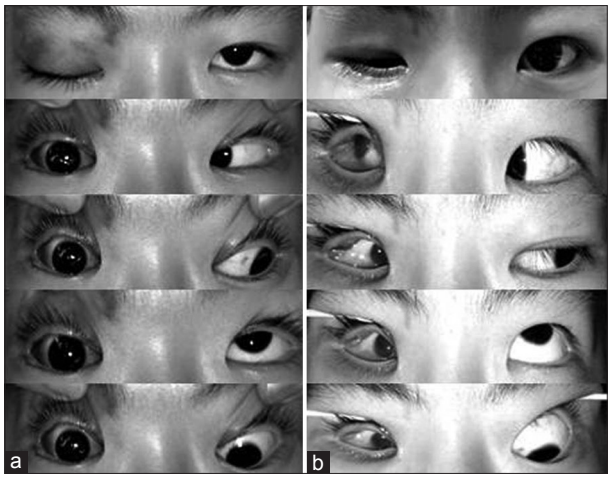

Figure 2: (a) Immediately after sclerotherapy, ecchymosis patches were noted on the right upper eyelid along with complete blepharoptosis and total ophthalmoplegia. (b) Four months after sclerotherapy, the right eye movement and blepharoptosis improved; however, enophthalmos and phthisis bulbi had developed

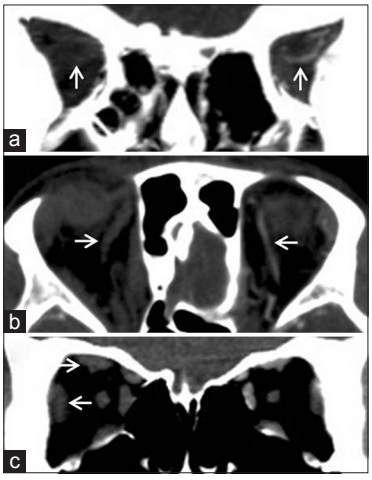

Figure 3: Computed tomography angiography 1 day after sclerotherapy showed no contrast enhancement in the right ophthalmic artery (a), superior ophthalmic vein (b), or extraocular muscles (c). Moreover, mild swelling of the extraocular muscles was observed in the right eye

circulation, hyperbaric oxygen (HBO) therapy was added on day 7. Even after ten courses of HBO therapy, the result was nonsignificant. In the 3rd week, temporary tarsorrhaphy was performed on the right eye because of persistent exposure keratoconjunctivitis. In the 4th week, when the tarsorrhaphy was removed, the conjunctival chemosis had subsided, with the cornea and anterior chamber becoming clearer and deeper, respectively. The fundus still could not be examined because a dense cataract had developed. One month later, the right eye remained blind, and the blepharoptosis and ophthalmoplegia had not improved. Examination of the right eye revealed near total hyphema. B-scan echography showed total retinal detachment. Four months later, ocular movement and the blepharoptosis had partially improved, but phthisis bulbi had developed [Fig. 2b].

Discussion

STS is one of the most commonly used detergent sclerosing agents for facial LFVMs. It acts on endothelial lipid molecules, causing surface damage and collagen exposure and leading to an inflammatory response that results in the fibrosis, scarring, and shrinkage of the VM with minimal thrombus formation.[3,4] Endothelial damage to the vein occurs 15 min after sclerosant injection, and adhesion between the thrombus and wall occurs 2–3 h after sclerotherapy.[5]

Sclerotherapy is considered a safe and effective treatment for certain VM types. Because VMs tend to recanalize and recur, multiple sclerotherapy sessions (1–13 sessions) are always required.[2] Complications of sclerotherapy include skin necrosis, transient pain, motor or sensory nerve injuries, deep vein thrombosis, pulmonary embolus, and cardiopulmonary collapse, with 0–61.4% complication rates.[2,6] Ocular complications of sclerotherapy are rare in head and neck VMs. If the VM extends into the orbit, the risk of developing ocular complications is higher during sclerotherapy. Siniluoto *et al*. and Sachin *et al*. each reported a case of unilateral vision loss after sclerotherapy for facial VMs with orbital involvement.[1,7]

In 1993, Borruat *et al*. first defined OIS as the ischemia of all intraorbital and intraocular structures.[8] The hallmarks of OIS include acute blindness, orbital pain, total ophthalmoplegia, and anterior and posterior segment ischemia. Although most orbital and ocular blood supplies originate from the OA, occlusion of the OA alone does not induce global orbital ischemia because of numerous anastomoses present between the branches of the OA and the external carotid artery (ECA). Therefore, the possible mechanisms of OIS include the occlusion of either an anomalous OA lacking anastomoses alone or both the OA and its anastomoses.[8] Treatments for OIS include anticoagulant, systemic steroid, and HBO therapies. However, the prognosis of OIS is poor. Blindness and retinal and optic nerve damage are often permanent. Nevertheless, blepharoptosis, ophthalmoplegia, and anterior segment ischemia may improve over time.[8]

In our case, the possible causes of OIS were as follows: (1) Retrograde flow of sclerosant into the OA during intralesional injections, leading to OA endothelium damage,

thrombosis, and occlusion; (2) an allergic and inflammatory reaction to the sclerosant, possibly causing orbital tissue swelling and a further compression of the OA; and (3) previous sclerotherapy sessions may have compromised most of the anastomoses between the branches of the OA and the ECA. If the VM extends into the orbit, the risk of ocular complications is higher during sclerotherapy. However, our case had no orbital involvement. Based on our research, this is the first reported case of OIS after treatment with STS for facial VM without orbital involvement. In addition, OIS prognosis in our case (phthisis bulbi) was the worst among all the reported cases.

In general, patients undergo an average of two sclerotherapy sessions;[2] however, our patient had received 14 sclerotherapy sessions. The multiple sclerotherapy sessions may have compromised most of the anastomoses between the OA and ECA, leading to poor circulation around the orbit and further resulting in OIS and phthisis bulbi. Thus, any physician administering sclerosant for facial VM with or without orbital involvement should know the possible ocular and orbital complications, particularly in patients undergoing multiple treatments.

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Conflicts of interest

There are no conflicts of interest.

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