American Journal of Ophthalmology Case Reports 15 (2019) 100457

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: www.elsevier.com/locate/ajoc

Case report

Ocular infarction following ethanol sclerotherapy of an arteriovenous malformation

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ARTICLE INFO

Keywords:
Ocular infarction
Arteriovenous malformation
Ethanol sclerotherapy
Cherry red spot
Ethanol embolization

ABSTRACT

Purpose: To illustrate a case of ocular infarction following percutaneous ethanol sclerotherapy of an orbital arteriovenous malformation.

Observations: The patient is a 31-year-old female who developed ocular infarction of the left eye with light perception vision, proptosis, ophthalmoplegia, and a cherry red spot following sclerotherapy of an orbital arteriovenous malformation. Fluorescein angiography demonstrated decreased arteriolar filling with vascular leakage, indocyanine green angiography showed decreased choroidal perfusion, and optical coherence tomography revealed full thickness retinal edema. Magnetic resonance angiography and venography were negative for venous sinus thrombosis or intracranial vascular compromise.

Conclusions and Importance: Ocular infarction is a rare and devastating disorder that may result in permanent vision loss. Ethanol sclerotherapy has been reported to be effective in treating arteriovenous malformations. To the best of our knowledge, this is the first report in the literature of ocular infarction following percutaneous ethanol sclerotherapy to highlight this disease with multimodal imaging.

1. Introduction

Arteriovenous malformations (AVMs) are abnormal feeding blood vessels which form a nidus that drains directly into the venous system without any intervening capillaries. AVMs are rare, with AVMs involving the orbit or the periorbital region being even less frequent.[1] AVMs can present anywhere in the orbit or the periorbital region with complex anatomical considerations in both their clinical presentation and treatment. Symptomatic orbital AVMs can present as a periocular mass, periocular edema, pulsation or bruit, proptosis, episcleral congestion, elevated intraocular pressure (IOP), pain, decreased vision, reduced extraocular motility, and diplopia.[1] Management of orbital and periorbital AVMs often requires multidisciplinary approach including embolization, surgical resection, and preoperative embolization with surgical resection.[2,3] Long-term results showed AVM recurrence rates of 81% after surgical resection (with or without embolization), and 98% with embolization alone.[4]

Ethanol sclerotherapy is among the treatment modalities used to address vascular malformations. Ethanol is unique in that it permanently destroys endothelial cells, and thus the nidus.[5] Traditionally, the use of sclerosants has been reserved for low-flow malformations.[6] However, high-flow AVMs with a percutaneously accessible nidus treated with ethanol sclerotherapy have been reported to have a ≥50% reduction in size in about 50% of patient with lower permanent complication rates compared to surgical excision with embolization.[7] *Pekkola* et al. reported cures in 11 out of 19 patients treated with ethanol sclerotherapy with low recurrence and complication rates, but did comment on periorbital AVMs having more complications (e.g. temporary Horner's syndrome, and severe epistaxis).[8]

Ocular infarction (OI) is infarction of intraocular structures, including the retina and choroid. Recognizing the etiologies and clinical symptoms of OI is important, but the prognosis is notably poor in many cases. The goal of this report is to illustrate a case of OI after percutaneous ethanol sclerotherapy of an orbital AVM.

1.1. Case report

A 31-year-old female with a history of left medial orbital and nasal AVM presented to an outside practice for AVM sclerotherapy. The AVM was noted to be primarily supplied by the bilateral ophthalmic arteries and left internal maxillary artery on preoperative arteriogram. A total of 14 mL of ethanol was reportedly used to thrombose the left orbit AVM compartments percutaneously. The patient was noted to have intact vasculature on immediate postoperative arteriogram. However,

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https://doi.org/10.1016/j.ajoc.2019.100457
Received 7 April 2018; Received in revised form 21 July 2018; Accepted 22 April 2019
Available online 25 April 2019

significant postoperative swelling developed resulting in the patient being unable to open her left eye. Once she could open her eye on postoperative day 4, she reported significant vision loss, and presented to an outside hospital. Due to orbital edema, proptosis, decreased vision, and an intraocular pressure of 30 mmHg, the patient underwent lateral canthotomy and cantholysis to relieve suspected orbital compartment syndrome. Patient was then transferred to us for further management on postoperative day 5.

On arrival, exam of the left eye showed visual acuity of light perception, positive left afferent pupillary defect, IOP of 21 mmHg, visual field deficit to count fingers in all quadrants, and motility deficit of −2 in abduction, adduction, infraduction and −4 in supraduction. There was upper and lower lid edema, periorbital ecchymosis, proptosis, and chemosis. Patient's cornea, anterior chamber, iris and lens were unremarkable. On dilated fundus exam a pale retina with a cherry red spot and no cilioretinal artery were apparent. The right eye had an unremarkable exam. Cranial nerve exam for V, VII, VIII, IX, X, XI, and XII were unremarkable. Patient was admitted to the hospital for further work-up to rule-out cavernous sinus thrombosis, orbital apex syndrome, carotid-cavernous fistula, and other vascular compromise.

Urgent CT angiogram (CTA) revealed possible decreased contrast filling of the left superior ophthalmic vein and the left cavernous sinus compared to the right. This was suspicious for decreased flow or thrombosis. Further evaluation with MR angiography and venography (MRA/MRV) revealed a symmetric and unremarkable cavernous sinus, and widely patent major dural venous sinuses with no filling defect to suggest thrombosis. Patient was subsequently discharged and followed closely as an outpatient.

On day 6 after AVM repair, the patient was seen in clinic for further evaluation. Left eye clinical exam was unchanged except for a mild decrease in orbital swelling. Fundus photos demonstrated an indistinct optic disc, arteriolar attenuation and segmental filling, diffuse patchy retinal whitening, retinal edema, and a cherry red spot (Fig. 1A). Optical coherence tomography (OCT) macula revealed diffuse retinal thickening with intraretinal cystic spaces (Fig. 3A). Early phase fluorescein angiography (FA) revealed poor retinal vascular filling with a leading arterial edge of dye (Fig. 2A), while indocyanine green angiography (ICGA) showed decreased choroidal perfusion (Fig. 2A). Late phase FA revealed vasculitis and leakage of dye (Fig. 2B). Repeat evaluation 20 days later was remarkable for hand motion vision, improved extraocular motility, decreased retinal thickening and areas of peripapillary atrophy (Fig. 1B). OCT macula revealed decreased retinal thickening consistent with retinal atrophy, and areas of choroidal thinning (from 242 μm thickness on post injection day 6, to 81 μm thickness at post injection day 25) corresponding to the areas of decreased perfusion observed on ICGA (Fig. 3B).

2. Discussion

OI has previously been reported in the treatment of orbital and temporal lobe AVMs with polyvinyl alcohol, and in the treatment of prominent facial veins with sodium tetradecyl sulphate,.[9–11] However, OI due to ethanol sclerotherapy has not been reported before, this case highlights the disease with multimodal imagining including fundus photos, OCT, and FA. On the other hand, orbital infarction defined as infarction of both orbital and intraocular structures, has been reported to occur due to sclerotherapy with sodium tetradecyl sulphate injected percutaneously into a facial low-flow venous malformation without orbital involvement, CTA in that case showed absent contrast enhancement of the ophthalmic artery and vein, suggesting thrombosis as an explanation for the cause of orbital infarction[12]. In our case, MRA/MRV on post injection day 5 showed patent orbital vasculature.

Retrograde flow of ethanol into the ophthalmic artery is suspected to be the cause of OI in our patient. This is supported by evidence of intraocular ischemia on exam and multimodal imaging, in addition to the retrieved arteriogram before and after sclerotherapy showing a sclerotic ophthalmic artery (Fig. 4 A & B). Inadvertent injection of ethanol directly into the orbit or the globe may produce a similar result as ethanol has been shown to be cytotoxic and induce vasospasm.[13] However, this is less likely as the dilated fundus exam was negative for any puncture wounds, the procedure was done under direct interventional radiology guidance, and the vasculitis seen on the fluorescein angiography is more consistent with retrograde flow of ethanol rather than vasospasm (Fig. 2B). Unfortunately, due to the patient's significant eyelid edema postoperatively, the timing of the patient's vision loss remains uncertain.

This case demonstrates the complex nature of orbital AVMs, and while ethanol sclerotherapy can be an effective treatment option, it may be associated with serious complications.[8,14] Patients should be made aware of irreversible vision loss and injury to the orbital structures as part of obtaining an informed consent. Furthermore, physicians managing AVMs of the orbital region should consider a multidisciplinary approach including a facial/orbital surgeon, interventional radiologist, and an ophthalmologist.

3. Conclusions

To the best of our knowledge this is the first reported case of OI after percutaneous ethanol embolization of an orbital AVM. In addition, this is the first report in the literature to highlight clinical retinal and

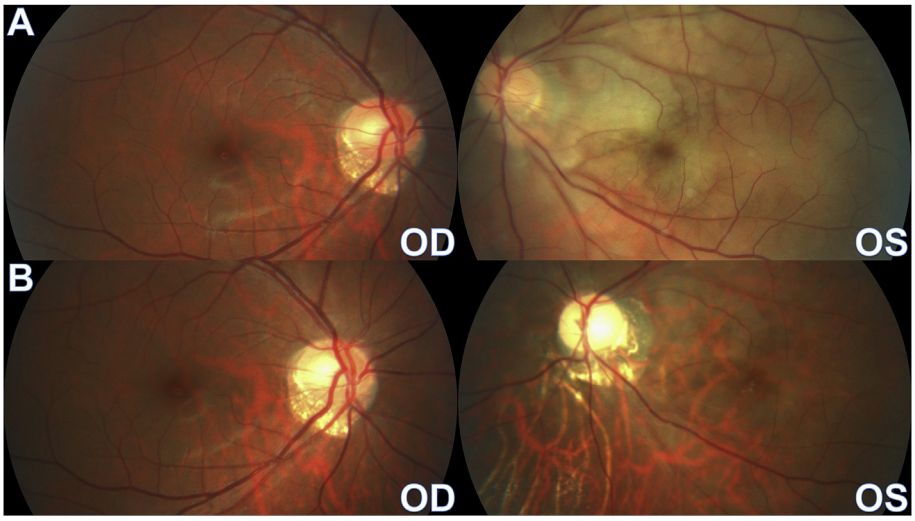

Fig. 1. Color fundus photos at six days (A) and twenty-five days post sclerotherapy (B). A) Fundus photos demonstrate indistinct optic nerve margins, arteriolar attenuation, diffuse patchy retinal whitening, retinal edema, and a cherry red spot (OS) vs. normal fundus (OD) at six days post sclerotherapy. B) Fundus photos demonstrate disc pallor, peripapillary atrophy, marked attenuation and obliteration of arterioles, residual macular edema, tessellated fundus appearance, and inferior coarse hyper pigmentation (OS) vs. normal fundus (OD) at twenty-five days post sclerotherapy. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

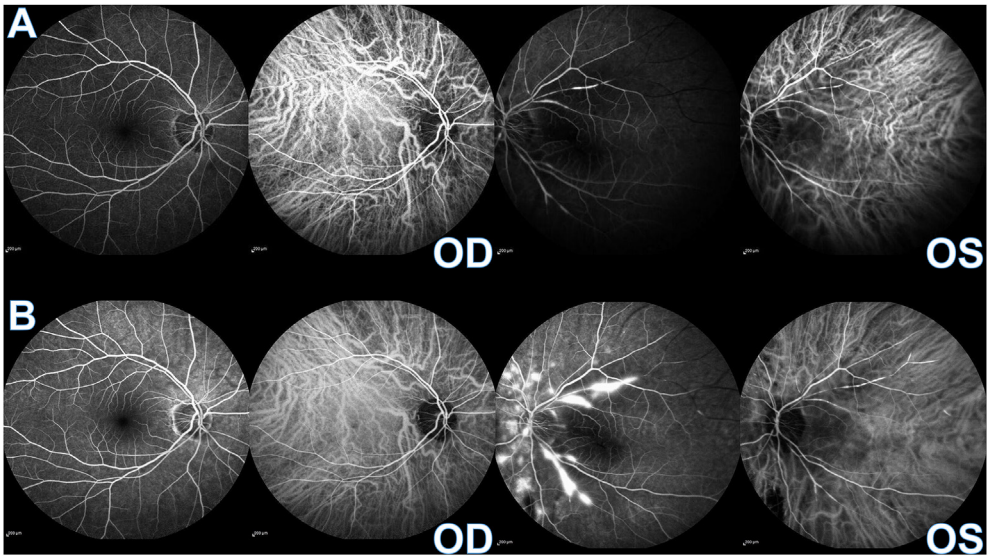

Fig. 2. Early phase fluorescein/indocyanine green angiography (A) and late phase fluorescein/indocyanine green angiography six days post sclerotherapy (B). A) Fluorescein angiography (FA) shows retinal hypofluorescence, segmental arteriolar hyperfluorescence, and delayed arteriolar fluorescein perfusion at 48 s, and indocyanine green angiography (ICGA) shows decreased choroidal perfusion (OS) vs. unremarkable angiography at 1 min and 16 s (OD). B) FA shows persistent arteriolar hypofluorescence and vasculitis at 5 min and 17 s, and ICGA shows persistent decreased choroidal perfusion (OS) vs unremarkable angiography at 4 min 55 s (OD).

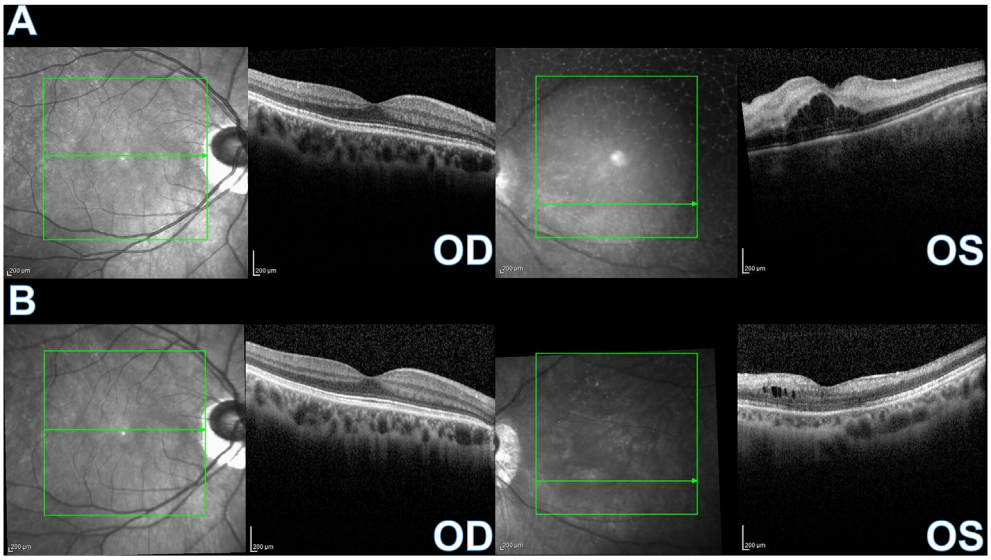

Fig. 3. Optical coherence tomography six days post sclerotherapy (A) and twenty-five days post sclerotherapy (B). A) Optical coherence tomography (OCT) demonstrating full-thickness retinal thickening, hyperreflectivity, disruption of foveal contour, outer retinal hyporeflectivity (OS), and choroidal thickness of (242 μm) vs. preserved foveal contour with distinguished retinal layers (OD) and choroidal thickness of (232 μm) at six days post sclerotherapy. B) OCT demonstrating persistent retinal thickening, irregularity, intraretinal cystic spaces, and thinning of the choroid (choroidal thickness of (81 μm) (OS) vs. unremarkable retinal scan (OD) with choroidal thickness of (213 μm) at twenty-five days post sclerotherapy.

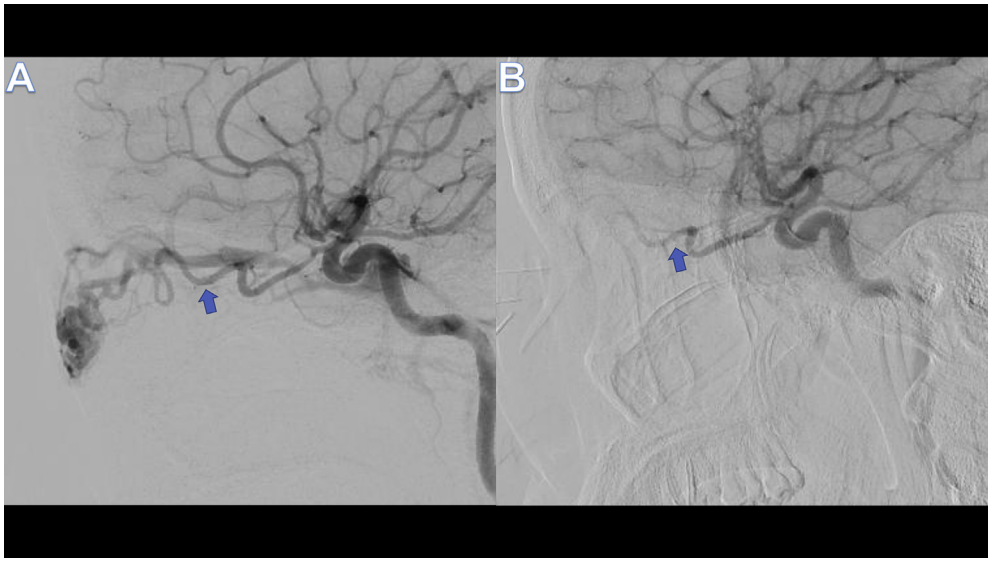

Fig. 4. Angiography of the left arteriovenous malformation pre-embolization & post-embolization. A) Pre-embolization arteriography of the left internal carotid artery demonstrates a lateral view of the orbital arteriovenous malformation supplied by the left ophthalmic artery (arrow). B) Post-embolization arteriography of the left internal carotid artery demonstrates decreased thickness of the left ophthalmic artery (arrow) without evidence of the arteriovenous malformation.

choroidal findings on multimodal imaging.

As novel therapies and innovative techniques are introduced in the management of orbital AVMs, the awareness of the potential complications, such as OI, becomes even more important. As such, close collaboration with an ophthalmologist or a professional experienced in the presentation and treatment of ocular pathology is essential.

Patient consent

Consent to publish was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

Funding

No funding or grant support.

Conflicts of interest

The following authors have nothing to disclosure: TL, YK, RA.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Acknowledgements

None.

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