Case Report

Intraorbital arteriovenous fistula with thrombosed varix: Diagnosis and treatment without catheter angiography in a developing country

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Abstract

Background: Pure intraorbital arteriovenous fistula (AVF) that do not connect to the cavernous sinus are quite rare. Presence of thrombosed varix in association with a spontaneous onset pure orbital AVF is even rarer.

Case Description: We report an interesting case of a 50-year-old female with pure intraorbital AVF of spontaneous onset with thrombosed varicose superior ophthalmic vein (SOV). She presented with 18 months history of right eye proptosis, upper lid swelling, and conjunctival chemosis with recent onset of retro-orbital pain and decreased vision. Computed tomography (CT) and angiography revealed thrombosed varicose SOV in association with pure intraorbital AVF. Direct surgical exposure of the thrombosed SOV through right fronto-orbital approach followed by excision of the fistulous lesion resulted in complete orbital decompression with disappearance of all symptoms by 2 months.

Conclusion: Spontaneous onset pure intraorbital AVF in association with thrombosed varicose SOV is extremely rare. In such cases, direct surgical exposure of the SOV followed by excision may accomplish complete closure of the fistula without significant risk for iatrogenic injury.

Key Words: Direct surgical exposure, pure intraorbital arteriovenous fistula, spontaneous onset, thrombosed varix

INTRODUCTION

Intraorbital arteriovenous fistulas (AVFs) that lie purely within the orbit and do not connect to the cavernous sinus are quite rare. DeKeizer’s series of 101 patients with carotid–cavernous and orbital fistulas included only 10 cases of intraorbital AVF. AVFs develop from a single communication between an artery and a vein. Because the bypass of capillary beds results in decreased vascular resistance, regional blood flows preferentially through the fistula, thereby exposing the veins to increased intraluminal pressure and varix formation. The optimal treatment of intraorbital AVFs is still a matter of debate because of the small number of reported cases. Successful treatment requires closure of the fistula. However, symptomatic AVF in association with thrombosed varices are only amenable to surgical excision under direct vision. We describe clinical and radiological findings in a patient with spontaneous onset purely intraorbital AVF with thrombosed varicose superior ophthalmic...
vein (SOV), which was treated by direct surgical excision and discuss the management strategy.

**CASE REPORT**

A 50-year-old woman, presented with 18-month history of painless progressive proptosis of right eye in association with upper lid swelling and conjunctival chemosis. Initially the proptosis was more marked during episodes of straining or prone/stooping positioning. Only in the last 1-month, she reported acute onset of retro-orbital pain, fixed proptosis, decreased vision, and mucopurulent discharge. There was no history of orbital trauma or family history of vascular disorders. Ophthalmologic examination of the right eye disclosed marked dilatation of conjunctival vessels, chemosis, and exophthalmos [Figure 1] with gross reduction of visual acuity (Perception of light). Ocular movements were almost normal. Intraocular pressure was 22 mmHg in the right eye and 10 mmHg in the left eye.

Computed tomography (CT) of the head and orbits demonstrated a nonenhancing, serpiginous soft tissue mass within right retrobulbar space, extending from the superior ophthalmic fissure to the anterolateral wall of the orbit [Figure 2] with no change in the size of the lesion on valsalva manoeuvre. Contrast enhancement was evident within the neighboring artery. It was clearly separated from the optic nerve and extraocular muscles. CT angiogram with 3D reconstruction revealed intracanal serpiginous vascular lesion arising from right ICA and ending at the level of SOV, which is grossly dilated and tortuous (2 cm) and filled with nonenhancing thrombus [Figure 3]. The SOV drained anteriorly into several small tributaries with slow outflow. No nidus and no communication into the cavernous sinus were observed [Figure 4]. These findings were consistent with the diagnosis of thrombosed varicose SOV in association with pure intraorbital AVF, which was further confirmed by high frequency (5-7.5 MHz) B-scan ultrasound with color doppler.

As the lesion was symptomatic due to dilated thrombosed SOV posing significant mass effect, open surgical excision was planned in our center. On opening the orbital roof through fronto-orbital craniotomy, a thickened serpiginous thrombosed SOV was found, which was resected [Figure 5] completely as far back toward the orbital apex as possible after ligation of the proximal feeding artery. Postoperative course was uneventful with good clinical outcome. CT scan obtained at the time of discharge demonstrated complete decompression of the orbit [Figure 6]. At 6 month follow, she was doing well with no recurrence.

**DISCUSSION**

AVFs in the orbit are quite rare, and most are part of facial arteriovenous malformations (AVMs).[7] In contrast to AVM, AVF develops from a single communication between an artery and a vein without having nidus.[8]
Most AVFs in the orbit occur after injury to an ethmoid artery because of fracture of the ethmoid bone with subsequent rupture of the artery into the ophthalmic venous system. They are usually low-flow fistulas and can occur spontaneously due to degeneration of vessels from hypertension, atherosclerosis, and other vascular diseases. In the absence of trauma, the orbital AVF in our case might have occurred spontaneously.

Having the same hemodynamic characteristics, AVFs of the orbit must be considered in the differential diagnosis of carotid-cavernous sinus fistulas (CCFs) and orbital AVMs. Increased orbital venous pressure and signs of orbital congestion, such as proptosis, dilation of conjunctival and retinal vessels, ocular hypertension, dilation of the SOV, and extraocular muscle enlargement can be found in all the three conditions. Orbital symptoms are frequently related not only to the degree of shunt but also to the adequacy of external drainage of the SOV. Very slow flow shunts can result in severe proptosis and chemosis because of poor or nonexistent external drainage. When orbital AVF produces varix, engorgement may occur through a valsalva manoeuvre or similar action (e.g., bending, coughing, and straining) resulting in intermittent periorbital pain, varying degrees of proptosis, thrombosis, and hemorrhage. Vascular complications such as variceal thrombosis or hemorrhage usually result in acute unilateral anopia due to optic nerve compression. Acute onset of retro-orbital pain and decreased vision as described by our case suggests variceal thrombosis.

Orbital varices in association with AVF are easily identifiable on either CT or duplex ultrasound imaging. Being a static modality, CT is less reliable in demonstrating flow across fistulae. Fine cut contrast-enhanced CT scans usually show ill-defined, heterogeneous multiloculated enhancing soft tissue mass(es) with connections to the orbital and extraorbital circulation. When thrombosed, orbital varices may or may not show patchy enhancement. Alternatively, duplex ultrasound using a high frequency (5-7.5 MHz) probe on B-mode with color doppler is a repeatable primary imaging modality for the orbit and reliably demonstrates flow across AVF of varying velocities. Angiographically, AVFs are characterized by a single arteriovenous connection within the vascular mass. Abrupt changes in the vascular caliber and/or the course of the feeding pedicle mark the location of the shunt between the artery and vein. In our patient, CT angiography revealed no evidence of a nidus; instead, a fistulous communication was found to exist between a minute artery from ICA and SOV. Accordingly, we made the diagnosis of an intraorbital AVF. Thrombosis of the dilated right SOV, as evident radiologically could have aggravated the clinical signs in our case. Though the role of catheter angiography is useful in evaluation and management of intraorbital AVF, due to unavailability in our center, neither it nor direct carotid stick angiography could be done in this case. On referral to another higher center, her relatives showed their inability due to poor economic status.

Treatment of intraorbital AVFs remains controversial. These orbital fistulas are usually managed by a conservative treatment, rather than by active vascular intervention. They are usually only treated if symptomatic (either due to mass effect or thrombosis/hemorrhage). Symptomatic AVF in association with thrombosed varices are only amenable to surgical excision under direct vision and the outcomes are usually satisfactory and associated with minimal morbidity. If thrombosed, the anterior part of the varix can usually be relatively easily excised surgically. Direct surgical removal by means of a fronto-orbital approach, could fully restore vision and correct proptosis. The surgical approach should be large enough to allow complete removal of the lesion. Sub-total excision may result in recurrence, and repeat treatment is often more complicated. Ideally the vein should be resected as far back toward the orbital apex as possible. In most of the published cases,
Surgery was the treatment of choice for our patient. In the present case, as the AVF in the orbit was associated with hard thrombosed SOV varix and caused mass effect, simple disconnection of the feeding artery might not have been sufficient to decompress the lesion. So the lesion was excised surgically by direct exposure of the SOV.

**CONCLUSION**

Spontaneous onset pure intraorbital AVF in association with thrombosed varicose SOV, as reported here, seems extremely rare. In such cases, direct surgical exposure of the SOV followed by excision may accomplish complete closure of the fistula without significant risk for iatrogenic injury.

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