

Proptosis Caused by Partially Thrombosed Orbital Varix of the Superior Orbital Vein Associated With Traumatic Carotid-Cavernous Sinus Fistula

—Case Report—

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Abstract

A 49-year-old female presented with proptosis and slight chemosis with diplopia on the right. The patient had a past history of skull base fracture occurring one year before the onset of the symptoms. Magnetic resonance imaging showed a partially thrombosed varicose aneurysm (varix) of the superior ophthalmic vein (SOV). Carotid angiography showed a pseudoaneurysm in the right cavernous sinus, associated with slow flow carotid-cavernous sinus fistula via the SOV. Proptosis was due to the direct mass effect of the thrombosed varix of the SOV. The thrombosed varix was removed after right carotid artery trapping with external carotid artery-middle cerebral artery (M₂) high flow bypass. The symptoms subsided after the surgery.

Key words: orbital varix, superior orbital vein, thrombosis, carotid-cavernous sinus fistula

Introduction

Orbital vascular diseases such as carotid-cavernous sinus fistula (CCF), arteriovenous malformation, aneurysm, and orbital varix are occasionally encountered in ophthalmologic and neurosurgical practices.⁴⁻⁶⁾ Orbital varices are generally considered to be a congenital venous pouch of the orbit, usually located on the superior ophthalmic vein (SOV), manifesting as intermittent proptosis,⁵⁻⁷⁾ but sometimes as sudden orbit pain, proptosis, diplopia, and slight conjunctiva congestion due to rapid thrombosis.²⁾ Orbital varix thrombosis may occur as a result of blood pooling in the varix and result in an acute orbital mass effect.²⁾ We recently experienced an extremely rare case of partially thrombosed orbital varix associated with traumatic CCF.

Case Report

A 49-year-old female was admitted to our hospital complaining of proptosis and diplopia. She had a past history of skull base fracture caused by a fall, and her ocular symptoms began one year later. She had remarkable proptosis in the right eye with only slight dilation of the ocular vessels (chemosis) (Fig. 1). The axis of the right eye ball was directed laterally. Extraocular movement of right eye was moderately limited in all directions. Visual acuity was 0.2 in the right and 0.7 in the left, similar to one year before. Intraocular pressure was 20 mmHg in the right (within



Fig. 1 Photograph showing prominent proptosis with slight chemosis.

normal limits) and 18 mmHg in the left eye.

Magnetic resonance (MR) imaging demonstrated a retrobulbar mass in the supero-medial portion of the right orbit with heterogeneous enhancement by gadolinium, causing a mass effect resulting in proptosis (Fig. 2). Computed tomography (CT) with contrast medium showed a tubular enhanced area inside the non-enhanced mass in the orbit, arising from the SOV (Fig. 3). The diagnosis was partially thrombosed varix of the SOV. Carotid angiography demonstrated an irregularly shaped aneurysm in the right cavernous sinus, with slow opacification of the dilated SOV and varix in the right orbit (Fig. 4). The inferior petrosal sinus was not opacified. Three-dimensional CT angiography demonstrated the hypoplastic anterior communication artery and the fetal type of the posterior cerebral artery. These radiological findings suggested the formation of pseudoaneurysm in the cavernous sinus, which had ruptured and caused slow flow CCF and par-

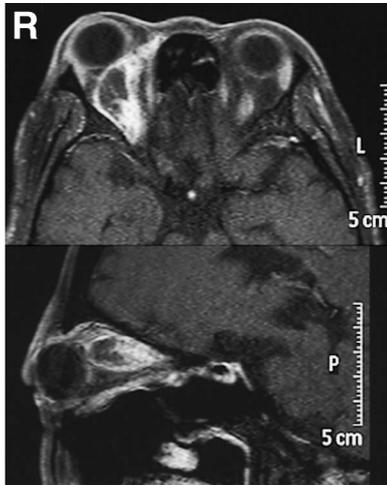


Fig. 2 T₁-weighted magnetic resonance images with gadolinium showing partial thrombosis of the varix in the right orbit.

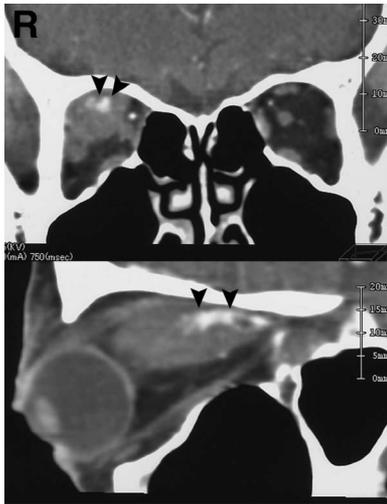


Fig. 3 Computed tomography scans with contrast medium showing a tubular enhanced area inside the non-enhanced mass in the orbit, arising from the superior ophthalmic vein (arrowheads).

tially thrombosed varix of the SOV in the orbit.

The proptosis was considered to result from the direct mass effect of the partially thrombosed SOV, rather than the more common cause of intraorbital congestion with blood reflux via the SOV. To improve the symptoms, treatment of both the blockage of the CCF and the thrombosed varix was necessary. External carotid artery-middle cerebral artery (M₂) high flow bypass with saphenous vein graft was performed. Then, the cervical internal carotid artery was ligated with 1-0 silk and the C₂ segment clipped proximal to the ophthalmic artery with a Sugita 10-mm clip. After unroofing and opening the periorbita, the partially thrombosed varix was removed.

Postoperative MR imaging showed complete removal of



Fig. 4 Right carotid angiograms showing a pseudoaneurysm in the cavernous sinus (upper, arrow) and a slow flow carotid-cavernous sinus fistula in the dilated superior ophthalmic vein and partially thrombosed varix (lower, SOV).

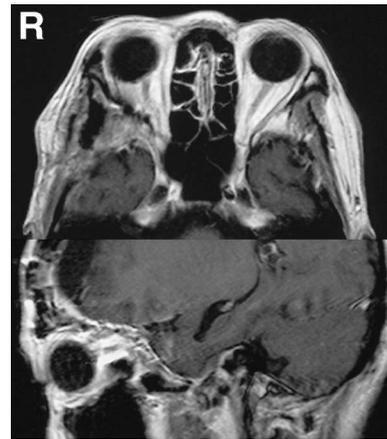


Fig. 5 Postoperative T₁-weighted magnetic resonance images with gadolinium showing complete removal of the intraorbital mass.



Fig. 6 Postoperative photographs showing complete recovery of the proptosis and extraocular movement.

the intraorbital mass (Fig. 5). Her symptoms completely subsided after the operation (Fig. 6). Histological examination showed varicose dilation of the SOV with intra-luminal thrombus formation (Fig. 7). Endothelial thickening and relatively thick fibromuscular wall were also recognized. Angiography confirmed patency of the high flow

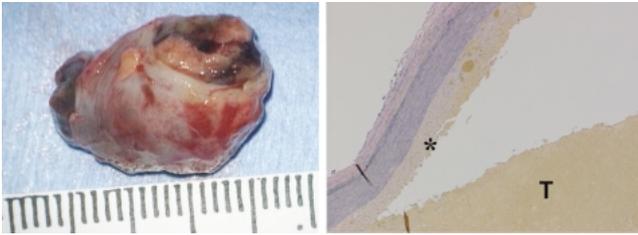


Fig. 7 Left: Photograph of the removed thrombosed varix in the orbit. Right: Photomicrograph of the specimen showing the varix contains thrombus (T), with endothelial thickening (asterisk) and thick fibromuscular wall. Van Gieson elastic stain, $\times 50$.

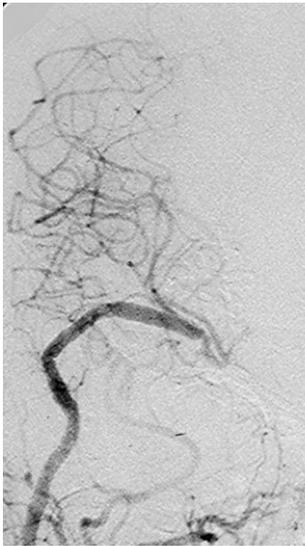


Fig. 8 Postoperative right carotid angiogram showing patency of the high flow bypass.

bypass at the time of discharge (Fig. 8).

Discussion

Three previous cases of orbital varix thrombosis have been reported, two due to thrombosis of the SOV, and one to thrombosis of the inferior ophthalmic vein.²⁾ The cause of thrombosis was probably stasis of pooled blood in the orbital varix for an extended period. These cases of orbital varix thrombosis manifested as the symptoms of acute mass effect in the orbit rather than orbital congestion as seen in cases of thrombosis of the SOV and cavernous sinus. The characteristic symptoms of thrombosis of the SOV and cavernous sinus reflect severe venous congestion, such as increased intraorbital pressure, retinal venous congestion, and eyelid edema.¹⁾ The three previous cases had good clinical outcomes after removal of the thrombosis via lateral orbitotomy.

In our case, the proptosis was prominent, but the chemosis was slight, and intraocular pressure was not significantly elevated. The global directional extraocular

movement limitation of the affected eye suggested the cause of the dysfunction was direct mass effect of the retrobulbar thrombosed varix rather than cranial nerve deficit due to venous congestion. These symptoms are compatible with those of typical orbital varix thrombosis. However, the etiology of the present case is different from previous cases of orbital varix thrombosis. The patient had a past history of skull base fracture and had developed a pseudoaneurysm in the cavernous sinus. Slow flow CCF was also recognized in the dilated SOV. We assume that the patient may have had an asymptomatic orbital varix, and the varicose vein was partially thrombosed and enlarged either by stasis of blood due to counter flow from the CCF or dislodging the thrombi from the pseudoaneurysm. Histological examination did not reveal the true cause of the thrombosed varix formation.

Modern methods of intravascular treatment have been used to successfully treat cases of traumatic pseudoaneurysm with and without CCF using balloons, coils, and stents.^{8,9)} However, endovascular treatment for pseudoaneurysm is still controversial because of the wall fragility, which, with the location and aging of the pseudoaneurysm, may determine the safety of endovascular treatment.^{3,8)} In the present study, the pseudoaneurysm in the cavernous sinus had an extremely irregular shape and the wall may have been fragile. Furthermore, surgical removal of the thrombosed large varix in the orbit was considered to be necessary to resolve the symptoms, so endovascular treatment was not selected in this case.

Our patient completely recovered from her orbital symptoms after removal of the varix, supporting our interpretation of the etiology of the orbital symptoms as the direct mass effect and the treatment modality of this rare clinical entity.

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