

Thrombosis of intraorbital arterio-venous fistula

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ABSTRACT

في تقرير الحالة هذا ، تعرض مريض يبلغ من العمر 65 عاماً لانسور شرياني وريدي نادر. تشمل الأعراض الاحتقان وانخفاض حدة البصر والجحوظ. يكشف مزيد من التحقيق عن انسور شرياني وريدي غير رضحي (AFV) مع تجلط الوريد العيني. خضع المريض لحجر القحف ووصفت مضادات التخثر للعلاج. تعافى المريض بعد شهرين من الجراحة. تم تقديم استراتيجية إدارة فعالة للمريض للتخفيف من الأعراض المتعلقة بالانسور الشرياني الوريدي النادر.

A 65-year-old male patient presented with a rare arterio-venous fistula (AFV). The symptoms included congestion, decreased visual acuity, and proptosis. Further investigation revealed a non-traumatic intra orbital AFV with ophthalmic vein thrombosis. The management strategy was craniotomy and the prescription of anticoagulants. The patient recovered 2 months after surgery demonstrating successful resolution of his presenting symptoms and an alternative approach to complicated cases of embolization.

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Earlier published studies have shown that intraorbital arteriovenous fistulas are related to the carotid-cavernous sinus. Our case report presents unique findings of a rare intraorbital arterio-venous fistula (AFV) with facial arteriovenous malformation where, as per clinical evidence, we found that it was not connected to the cavernous sinus. Non-traumatic AFVs

are rare, while traumatic arteriovenous fistulas are more prevalent due to trauma.

In this report, we found an increased risk of thrombosis due to higher intraluminal pressure that exposes the veins and increases regional blood flow through the fistula in cases related to an arteriovenous fistula. The patient presented with chemosis, conjunctival congestion, and visual disturbances as per magnetic resonance imaging and CT scans. These symptoms are linked to thrombosis associated with a superior ophthalmic vein. Management strategies and appropriate intervention is required for patients suffering from an underlying disorder associated with AVF.

Case Report. Patient Information. A 65-year-old man presented with a one-month history of painless increasing proptosis of the left eye, upper lid edema, and conjunctival chemosis. The proptosis was initially more noticeable during episodes of straining or prone/stooping positioning; however, 2 days prior, he had had an acute commencement of quick vision loss, conjunctival chemosis, fixed proptosis, and mucopurulent discharge. There was no history of orbital trauma or vascular disease in the family.

Clinical findings. On admission, an ophthalmologic examination revealed significant dilation of the conjunctival vessels, chemosis, and exophthalmos of the left eye, as well as a significant loss in visual acuity (Figures 1A & 1B). Ocular movements were virtually normal. Right eye intraocular pressure was 14 mm Hg, whereas the left eye was 42 mm Hg due to open-angle glaucoma. Increased venous pressure downstream of the episcleral anastomoses resulted in a decrease in venous blood outflow from the orbit, and the elevated episcleral venous pressure (EVP) further reduced aqueous outflow. Proptosis of 6 mm was detected in the left eye as per Hertel measurements. In addition, the left eye demonstrated a relative afferent pupil defect (RAPD) using the swinging light test. Both fundi were normal (Figure 1C).

On a computed tomography (CT) scan of the head and orbits, asymmetrical enlarged hyperdense left superior ophthalmic vein as well as partially visualized

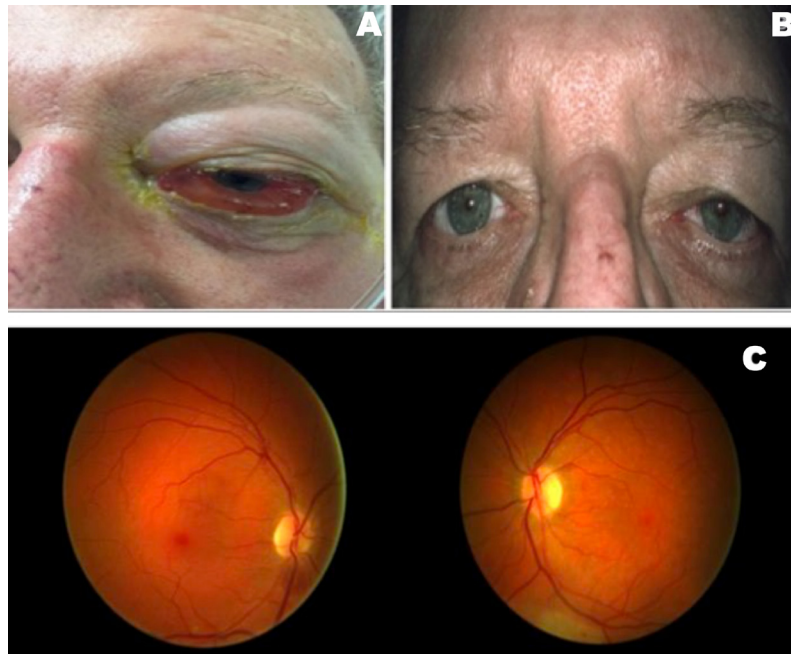


Figure 1 - Clinical photograph showing A) progressive proptosis of the left eye associated with upper lid swelling and conjunctival chemosis, B) Resolved proptosis, motility limitation, and dilated episcleral vessels. C) Fundus photo of both eyes showing normal appearance

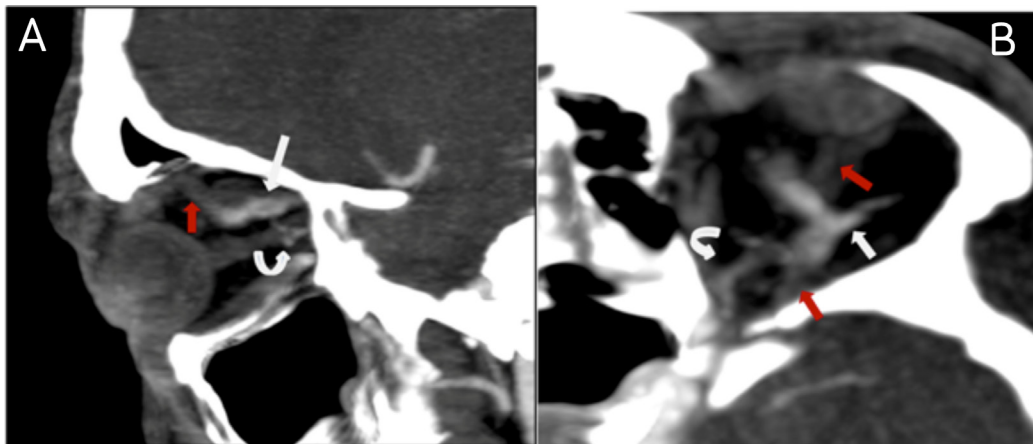


Figure 2 - CT angiogram showing A & B) early opacification of left ophthalmic vein (white arrow) through small ophthalmic artery (curved arrow) and the contrast confined to the enlarged tortuous superior ophthalmic vein with lack of opacification within the proximal and distal vein (red arrow) representing thrombosed varix

left periorbital soft tissue asymmetry and swelling was noted.

The CT angiography showed early opacification of left ophthalmic vein through small ophthalmic artery and a contrast confined to the enlarged tortuous superior ophthalmic vein with lack of opacification within the proximal and distal vein representing thrombosed varix (Figures 2A & 2B).

There was no direct drainage to the cavernous sinus, and there was no pathway into it. In the presence of

pure intra orbital AVE, these findings matched the diagnosis of thrombosed varicose superior ophthalmic vein (SOV).

Magnetic resonance angiography utilizing MRA TOF technique showed flow-related signal in the left superior ophthalmic vein in an arterial phase denoting the diagnosis of the fistula. The contralateral side appeared unremarkable.

Digital subtraction angiogram (DSA) of the left ICA artery shows early filling of the superior ophthalmic

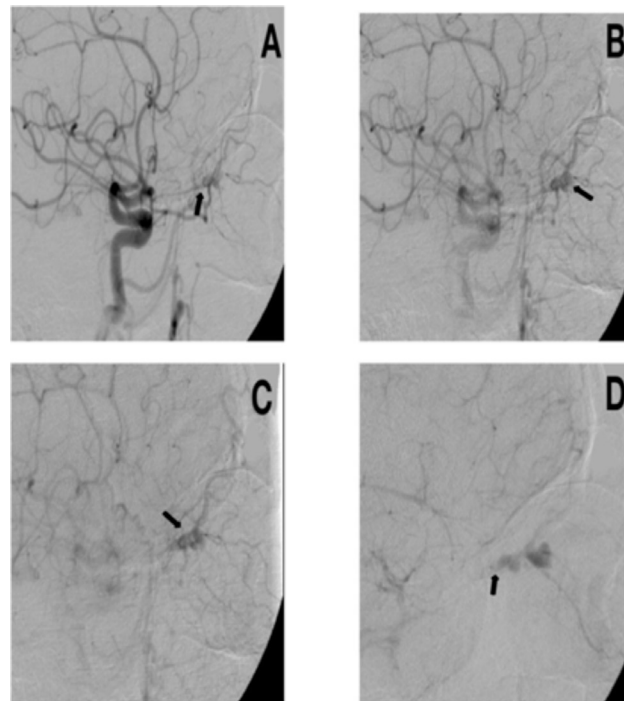


Figure 3 - Digital subtraction angiogram of carotid artery showing A) early faint filling of the superior ophthalmic vein. B) Late arterial phase clear opacification of arteriovenous fistula. C & D) Venous phase shows Tortuous dilated superior of the ophthalmic vein.

vein through small branches of the left ophthalmic artery and on venous phase shows persistent dilated superior of the ophthalmic vein and some flow drain anterior to the left facial vein. There was a lack of flow, and no drainage to the left cavernous sinus or cortical venous reflux noted which likely suggests thrombosed left superior ophthalmic vein (Figure 3)

Therapeutic intervention. A left pterional craniotomy was performed to accomplish immediate orbital and optic decompression. This was combined with rigorous anticoagulant therapy and local/systemic eye pressure lowering agents. The orbit had been entirely decompressed, according to a CT scan done at the time of discharge.

The DSA of the carotid artery (Figure 3) shows early faint filling of the superior ophthalmic artery, late arterial-phase opacification of arteriovenous fistula, and venous phase shows tortuous dilated superior of the ophthalmic vein. The funduscopy images of both eyes (Figures 1A & 1B) are shown respectively.

Follow-up and outcomes. After the operation, the patient's visual acuity increased considerably to 0.4 OS. Proptosis, motility limitation, and dilated episcleral vessels were all resolved. His symptoms had improved by the fourth week following surgery and had completely

disappeared two months later. Without medication, IOP in his right eye was 16 mm Hg OD and 18 mm Hg OS in his left eye, and his visual acuity was 0.8 OS after 2 months (Figures 1A & 1B). Patient's timeline was summarized in Figure 4.

Discussion. The combination of non-traumatic intra-orbital AVF with SOV thrombosis is a rare occurrence. Facial arteriovenous malformations are the most common intra-orbital AVFs (AVMs). AVF, unlike AVMs, begins with a single contact between an artery and a vein but no nidus.^{1,6,7} Intra-orbital AVFs are most usually caused by a physical lesion to the ethmoid artery, such as a fracture of the ethmoid bone and a rupture in the ocular venous system.^{6,7} Fistulas can develop rapidly in non-traumatic situations due to artery degeneration caused by atherosclerosis, hypertension, or other vascular disorders.⁷

Patient's perspective. The patient was concerned about the risk of thrombosis and the symptoms related to AVF. He stated that he did not want to lose vision in his left eye. He also mentioned that visual disturbances and conjunctival chemosis were major issues for him.

The presence of SOV thrombosis in combination with AVFs raises the risk of vision loss.⁸ Although the

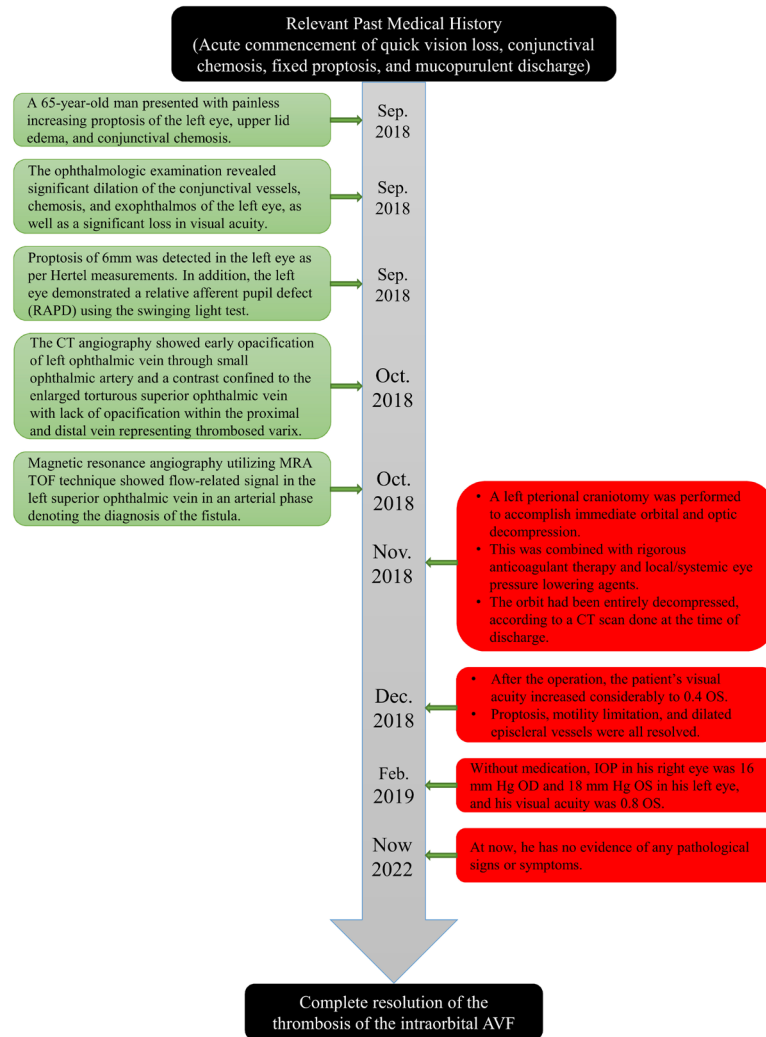


Figure 4 - Patient's timeline.

65-year-old man in the case study presented at the clinic with a painless progressive proptosis of the left eye and conjunctival chemosis, he had no history of orbital trauma nor family history of vascular problems, so we assumed he had non-traumatic intra orbital AVF. As previously indicated, CT examination assisted in the diagnosis of AVF with SOV thrombosis in our case.⁵

Mishra et al⁷ also reported a case of pure intra orbital AVF with thrombosed varicose SOV, which was similar to ours. Their patient, on the other hand, had a lengthy history of proptosis, chemosis, and a recent onset of retro-orbital pain and reduced vision, spanning 18 months. Mishra et al⁷ performed a frontal-orbital

craniotomy and removed the thrombosed SOV, even though the treatment is case-specific.

We also performed an emergency craniotomy because there was substantial danger of sight loss and the conservative method should only be used in moderate instances. Within four weeks, our patient began to heal, and two months after surgery, there was neither proptosis nor chemosis. Some case series and case reports of endovascular therapies for AVFs that do not require surgical excisions have been published. The use of trans-venous or trans-arterial embolization has yielded positive outcomes.^{6,9} Wang et al¹⁰ described a case of significant SOV thrombosis with dural AVF

of the ocular vein that was successfully treated by transvenous embolization. In our situation, medical care included the use of anticoagulants, which has been supported by other clinical trials.^{5,8}

Because AVFs and carotid-cavernous sinus fistulas (CCFs) and orbital AVMs have comparable hemodynamic characteristics, they are included in the differential diagnosis. Proptosis, ocular hypertension, extraocular muscle expansion, SOV dilation, and dilatation of retinal and conjunctival vessels are common symptoms of increased orbital venous pressure and congestion of the orbital arteries.^{1,9} The case study's ophthalmologic examination revealed substantial dilatation of conjunctival vessels, chemosis, and exophthalmos of the left eye, as well as a significant loss in visual acuity. This conclusion supports prior research that found hemodynamic similarities between AVFs and CCF features.

Notably, the degree of shunt and the adequacy of SOV's external drainage is linked to the symptoms of orbital venous pressure and orbital congestion. Due to insufficient or absent external drainage, a sluggish low shunt frequently causes severe chemosis and proptosis.^{6,10} The degree of a shunt in the case study had gradually developed and reached intense levels two days previously when the patient exhibited an extreme reduction in vision, fixed proptosis, conjunctival chemosis, and mucopurulent discharge.

Conclusion. Arteriovenous fistulas are rare but traumatic in origin. Ophthalmic vein fistulas are a rare form of AVF that emerge spontaneously as they mimic symptoms related to cavernous carotid fistulas. The case reported shows dilated SOV. Given that a transvenous approach from the skin is difficult to perform due to several small tributaries, we suggest obliteration of the fistulous point as the choice of treatment for arteriovenous fistula. The superior ophthalmic vein was directly exposed as it did not drain into the cavernous sinus and the orbital cavity was decompressed. This case report demonstrates successful resolution based on the patient's presenting symptoms and an alternative approach to complicated cases of embolization.

Written informed consent was obtained from the

patient for publication of their clinical details and/or clinical images.

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