

**CAROTID-CAVERNOUS FISTULA; DIAGNOSIS WITH MAGNETIC RESONANCE IMAGING.****Dr. Krishna Kumar M.\***

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Article Received on 29/12/2015

Article Revised on 19/01/2016

Article Accepted on 10/02/2016

**ABSTRACT**

A carotid-cavernous fistula (CCF) is an abnormal communication between the venous cavernous sinus and the carotid artery. The fistula may occur spontaneously but usually occurs following some sort of head trauma. Patients with CCF generally present with varied complaints, including unilateral visual loss, proptosis, lid swelling, pulsatile tinnitus and/or diplopia. A triad of clinical findings has been described as exophthalmos, orbital bruit, and dilated conjunctival vessels. We report a 59 years old male, presenting with persistent redness & painless bulging of Right eye since two months, not relieved by medicines, provisionally diagnosed as CCF, referred for Magnetic Resonance Imaging (MRI) evaluation.

**KEYWORDS:** Carotid-cavernous fistula, Dural AV fistula, Dural carotico-cavernous fistula, Endovascular treatment, Magnetic Resonance Angiography, Magnetic Resonance Imaging.

**INTRODUCTION**

Carotid cavernous fistula (CCF) is an abnormal communication between the carotid arterial system (ICA or ECA) and the venous cavernous sinus. Benjamin Taverns first described this condition in 1809 as unilateral pulsating exophthalmos in a patient who subsequently lost vision in the affected eye.<sup>[1]</sup> CCFs are classified as direct or indirect, traumatic or spontaneous, and high or slow flow based on arterial supplies, etiology, and shunt flow rate, respectively.<sup>[2]</sup>

Barrow *et al*<sup>[2]</sup> Classified CCFs into four angiographic types based on arterial supply. Type A fistulas are direct communications between the internal carotid artery and the cavernous sinus. Types B, C and D are indirect shunts (dural), because fistulas to the cavernous sinus arise from dural arteries and not directly from the internal carotid artery. Direct CCF (Type A) usually occurs in young men secondary to trauma. Indirect CCFs (dural CCF) usually occur in postmenopausal, hypertensive women arising from dural branches of either internal carotid artery (ICA) (Type B) or external carotid artery (ECA) (Type C) or both (mixed or Type D).<sup>[2]</sup>

Patients often complain of a swollen red eye, orbital pain, diplopia, headache and progressive vision loss.

Common clinical signs include proptosis, orbital bruit, chemosis, extraocular palsy, pulsating exophthalmos, ptosis, elevated intraocular pressure, anterior segment ischaemia and retinopathy.<sup>[3]</sup>

We evaluated the MRI features of CCF in a 59 year old male patient presenting with persistent redness & painless bulging of Right eye.

**CASE REPORT**

A 59 years old male, presenting with persistent redness & painless proptosis of Right eye since two months, not relieved by medicines was referred for Magnetic Resonance Imaging (MRI) evaluation with suspected diagnosis of right sided Carotid.Cavernous fistula (CCF) based on clinical suspicion. Patient admitted that he had minor head injury two months back, following which he developed the present problem.

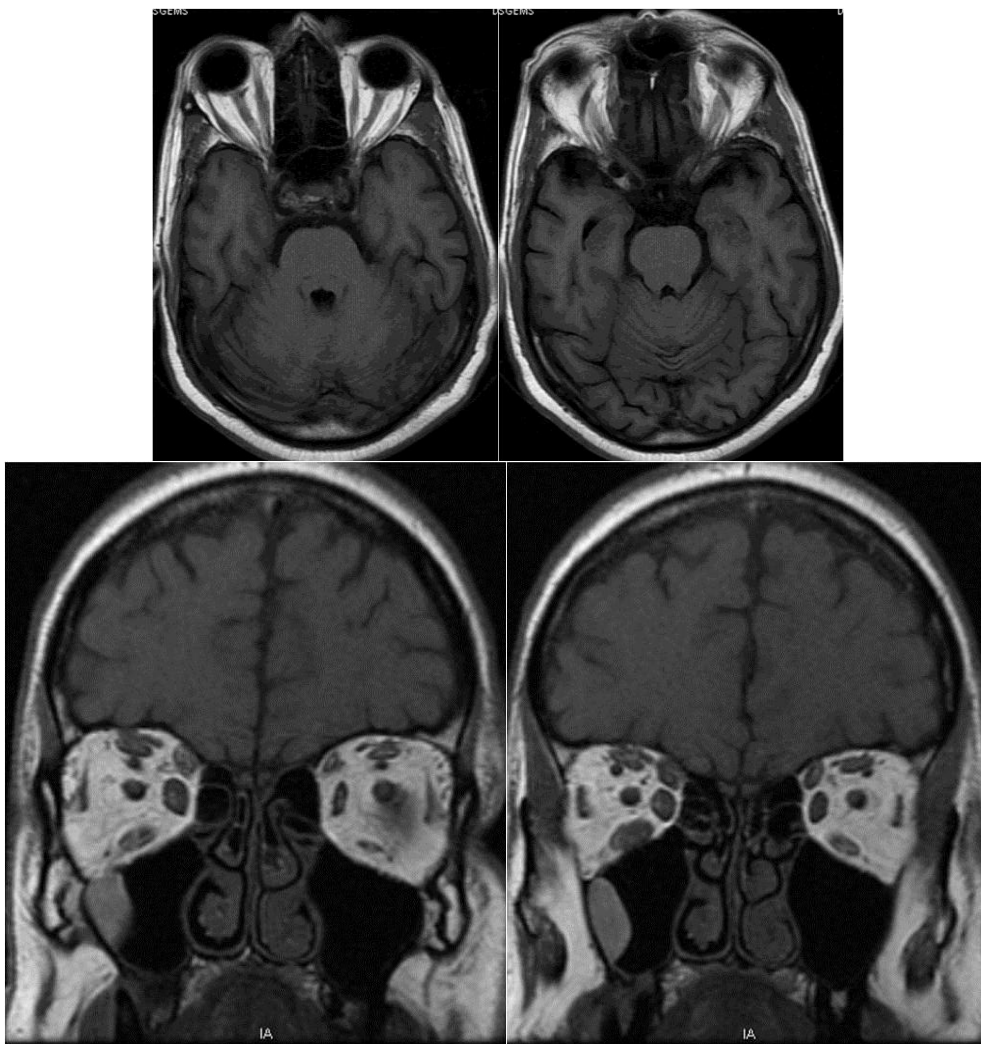
Physical examination revealed visual acuity of 6/6, normally reactive pupils with no relative afferent pupillary defect, raised intraocular pressure 14mm Hg and conjunctival injection .Dilated fundoscopic examination showed normal macula and vasculature. External examination showed venous engorgement of the right upper and lower eyelids and proptosis [Fig.1].

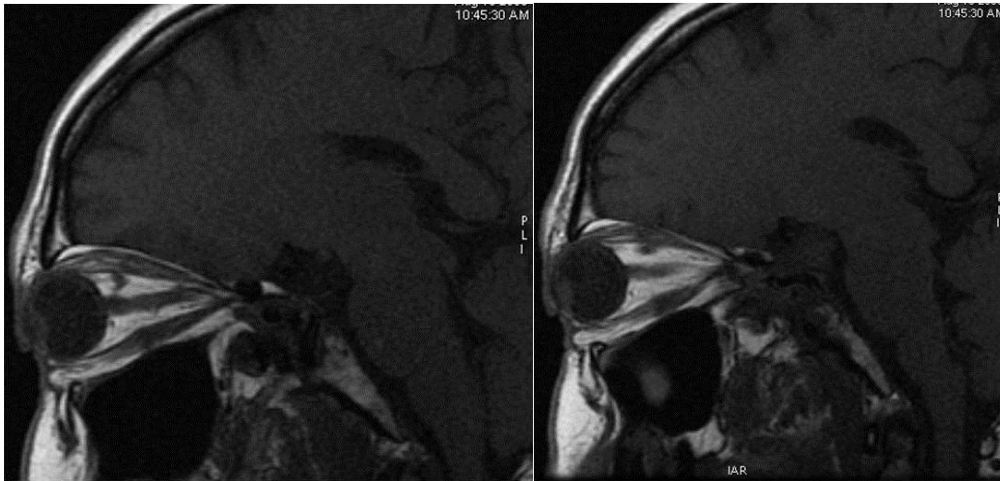


**Fig 1. External Examination two months after initial injury. Note the swelling of the upper and lower right eyelids, as well as the engorged conjunctival vessels associated with proptosis.**

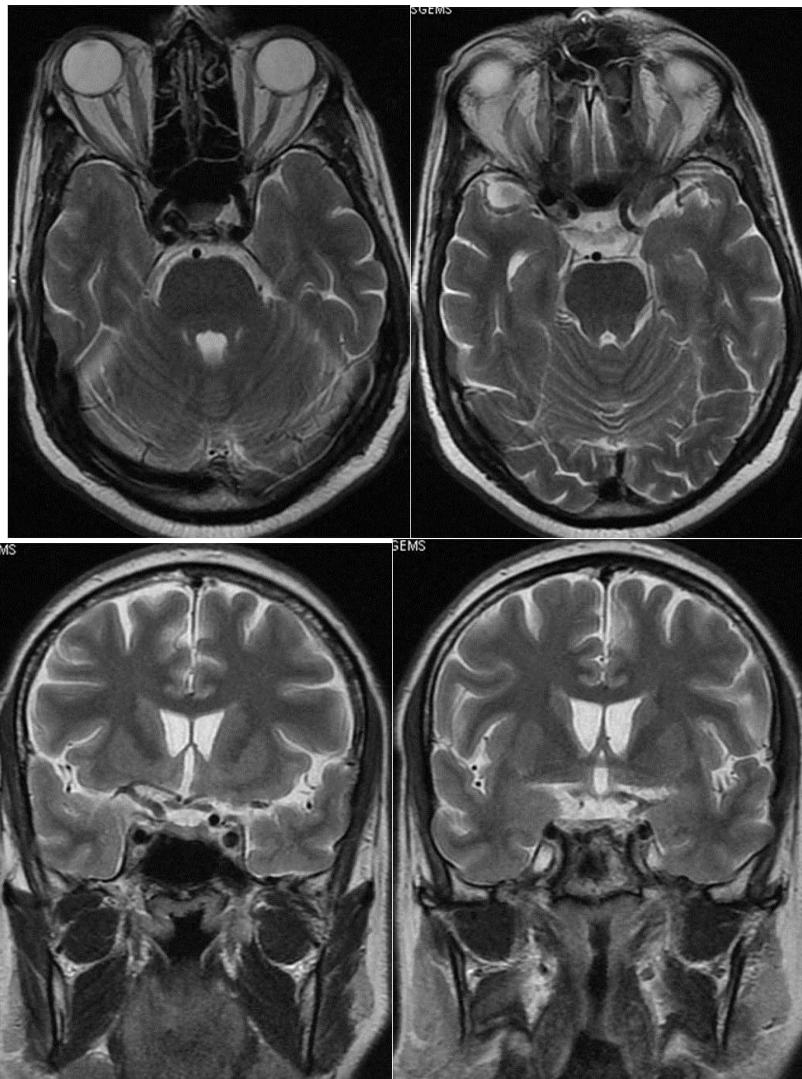
The patient underwent MRI on a 1.5T magnets (Signa GE medical systems). The images were processed by GE workstation. Standard T1 and T2 Weighted [Fig.2,3], Fat

saturated T2- Weighted [Fig.4], Fluid attenuation inversion recovery (FLAIR) [Fig.5], spin-echo sequences were used.

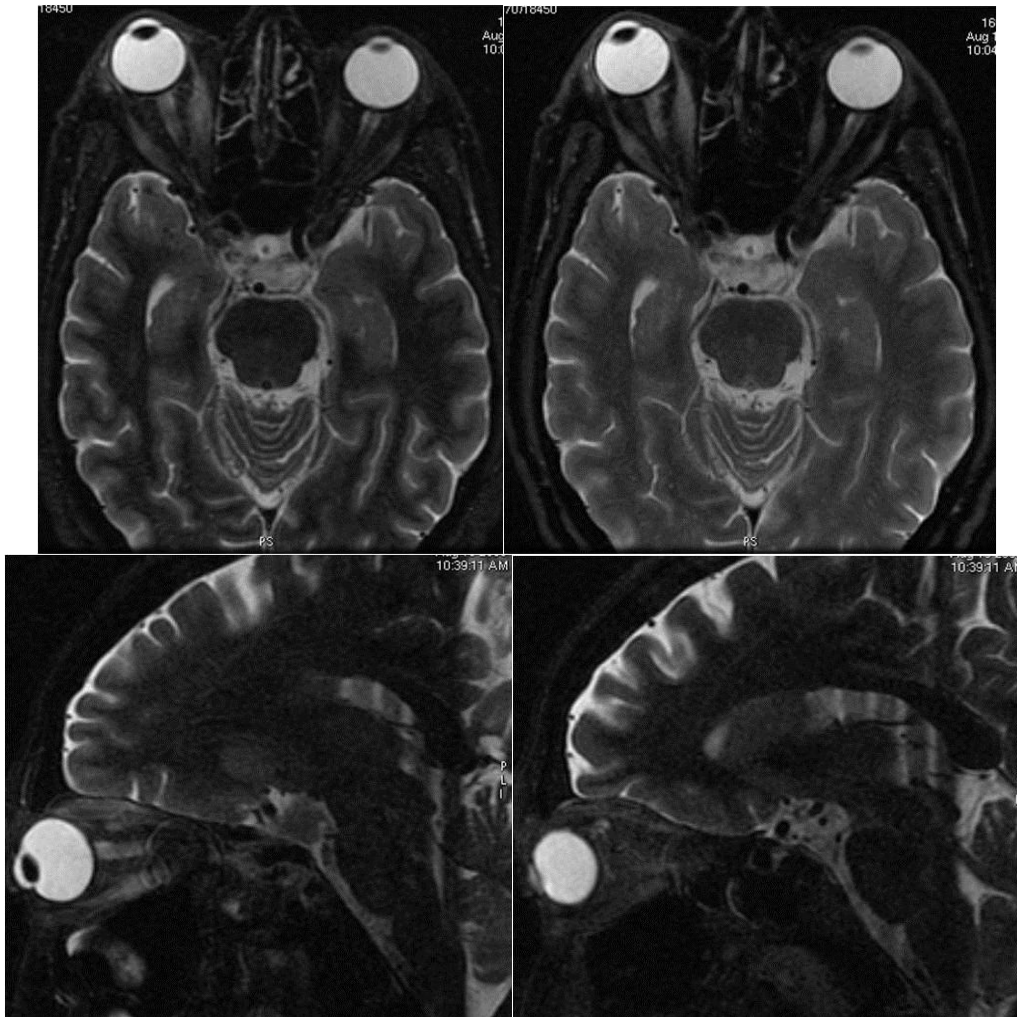




**Fig 2.**T1 Weighted Axial (A,B), Coronal (C,D) & Sagittal (E,F), images showing arterialization of cavernous sinuses (CS) and arterialization of the surrounding venous structures appearing as flow void.



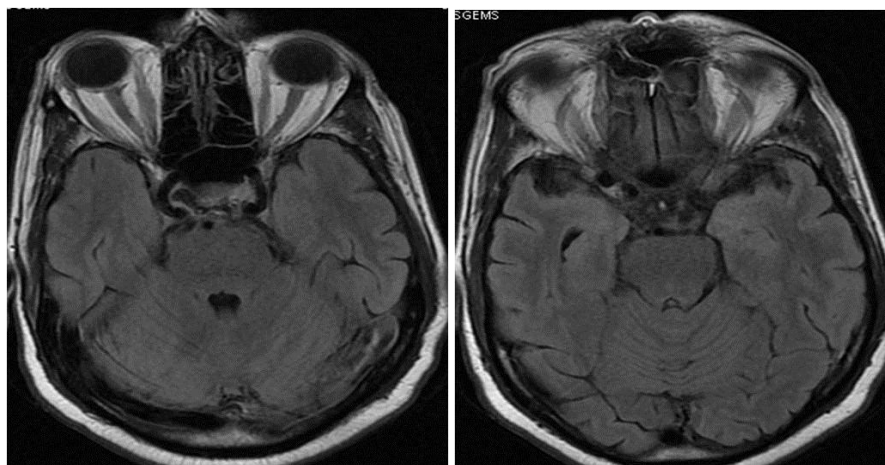
**Fig 3.** T2 Weighted Axial (A,B) & Coronal (C,D) images show Arterialization of cavernous sinuses (CS) with flow void appearance.



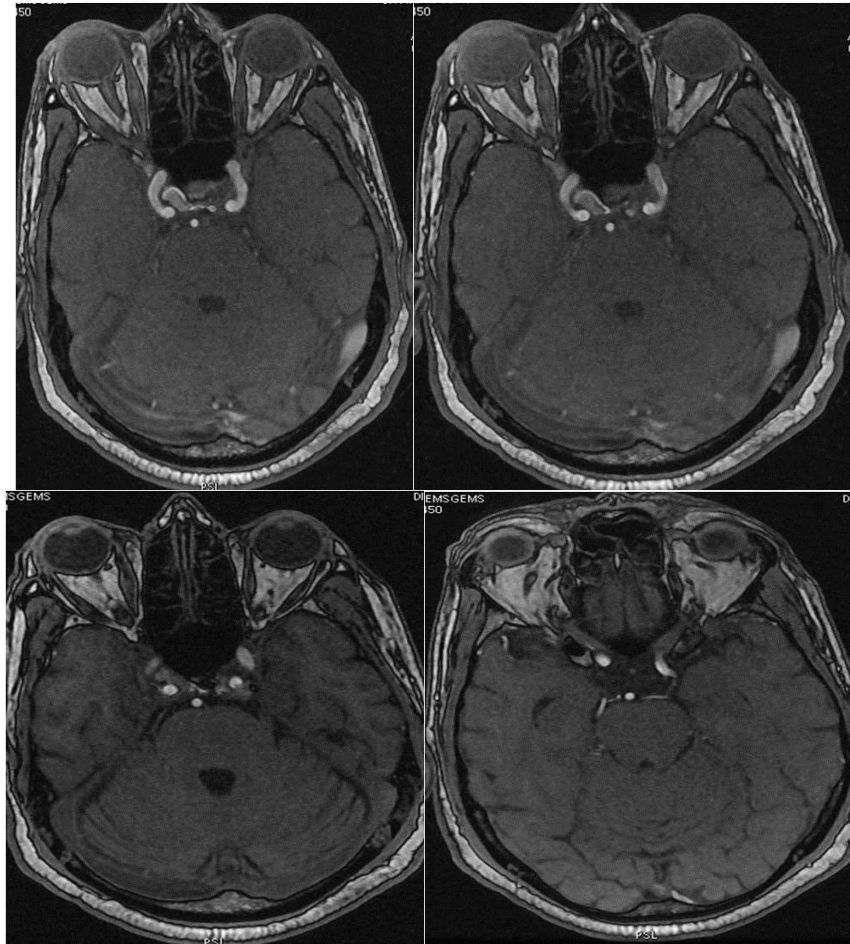
**Fig 4.** T2 Weighted Fat Saturation Axial (A,B) & Sagittal (C,D) images reveal right orbital congestion, thickened extraocular muscles and exophthalmus with dilated SOV and no evidence of flow void appearance of the right CS.

T1-weighted post contrast enhanced (0.1mmol/kg gadopentete dimeglumine) axial Images [Fig.6] were taken.

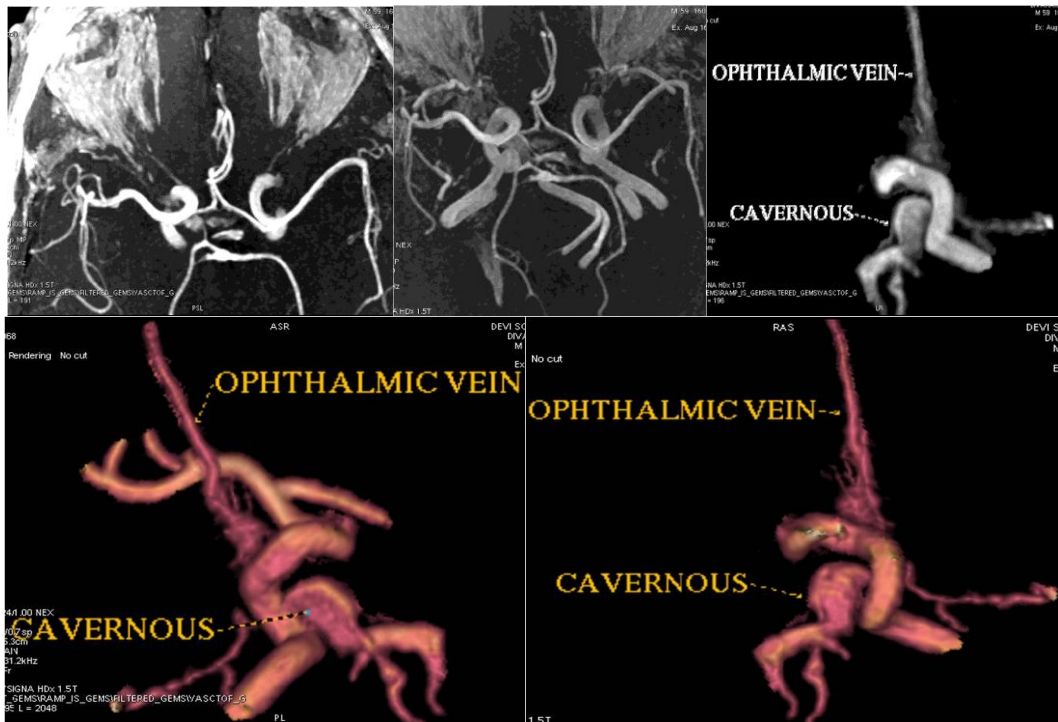
For the Magnetic Resonance Angiography (MRA) studies, a two-dimensional time-of flight (TOF) was used with a neurovascular phased array coil. The images demonstrated a right CCF as well as a markedly dilated right superior ophthalmic vein (SOV) (Figures 2-7).



**Fig.5.** Fluid Attenuated Inversion Recovery (FLAIR) images reveal Arterialization of cavernous sinuses (CS) with flow void appearance.



**Fig 6. MRI of the brain with contrast. T1W Axial images (A,B,C,D) show rapid increased flow in the Right cavernous sinus and ectatic dilated right ophthalmic vein.**



**Fig 7. Magnetic Resonance Contrast Angiogram (MRA) image demonstrating a flow related signal enhancement in cavernous sinus (CS) on maximal intensity projection (A,B) and enlarged Right superior ophthalmic vein (SOV) and its early filling on arterial imaging(C,D,E).**

Findings suggestive of CCFs on noninvasive MRI were orbital congestion, thickened extraocular muscles, exophthalmos, dilation of SOV, asymmetric enhancement of the cavernous sinus and surrounding venous system, and arterialization of the surrounding venous structures appearing as flow void on MRI or flow related signal on MRA.

## DISCUSSION

Carotid cavernous fistula (CCF) is an abnormal communication between the carotid arterial system and the venous cavernous sinus (CS). Increased pressure within the CS seems to be the mainstay of pathophysiology, although arterial steal and increased flow plays an important role.

The abnormal communication results in high-pressure arterial blood entering the low-pressure venous cavernous sinus, which interferes with the normal patterns of venous drainage and which compromises blood flow into the cavernous sinus and globe.<sup>[3,4]</sup> Direct CCFs usually occur suddenly, resulting from a tear in the arterial wall connecting the internal carotid artery to the cavernous sinus.<sup>[3-6]</sup> The fistulas typically have a high flow rate. Indirect CCFs have a more gradual onset over days, weeks, or months, a less fulminating course and generally lower flow.<sup>[2,7]</sup>

Traumatic CCFs are usually high flow and direct type fistulas with sudden onset of symptoms. They predominantly occur in young men because of higher incidence of trauma in this population. Underlying mechanisms are direct injury from the skull base fracture or injury from torsion or stretching of the carotid siphon upon impact and impingement of the vessel on bony prominences. Direct CCFs following surgical procedures such as endoscopic nasal surgery and vascular neurosurgery or spontaneously from aneurysm rupture have also been reported. Spontaneous CCF's occur secondary to hypertension, atherosclerosis, neurofibromatosis, and collagen vascular disorders. They are usually slow flow indirect type fistulas with insidious onset, commonly seen in elderly, postmenopausal, and hypertensive women with another peak of incidence during pregnancy.<sup>[2,8,13-15]</sup> Bilateral CCF cases comprise 12–15% of all and are usually indirect in variety.<sup>[8]</sup>

CCFs drain toward anterior via ophthalmic veins, inferior via pterygoid plexus and inferior petrosal sinus (IPS), contralateral via intercavernous connections, posterior via deep venous system, superior petrosal sinus (SPS), and cerebellar veins, and superior via superficial middle cerebral vein (SMCV). Mostly patients present with orbital symptoms secondary to anterior drainage but clinical presentation may change according to venous drainage pattern. Less commonly, headache, altered mental status, and other neurological deficits such as ischemia or infarction secondary to venous hypertension or steal phenomena may occur.<sup>[8-13]</sup>

Patients with a direct Type A fistula generally present with varied complaints, including unilateral visual loss, proptosis, lid swelling, pulsatile tinnitus and/or diplopia. A triad of clinical findings has been described as exophthalmos, orbital bruit, and dilated conjunctival vessels. Clinical findings include venous congestion of the eyelids, conjunctiva and episcleral vessels, cranial nerve palsies,<sup>[16, 17]</sup> visual loss, proptosis, elevated intraocular pressure, optic disc edema, and dilated and tortuous retinal vessels.

Complications include vision loss and, in rare cases, ischemic ocular necrosis.

Non-ocular symptoms are though fatal are less common and include epistaxis, subarachnoid hemorrhage and intracerebral hemorrhage due to rupture of fistula.

When a dural CCF is suspected, CT scanning, CT angiography, MR imaging, MR angiography, orbital ultrasonography, duplex carotid sonography, transorbital and transcranial color Doppler imaging, or a combination of these tests may be of benefit in confirming the diagnosis.<sup>[18]</sup> The gold standard diagnostic test, however, as in the case of the direct CCF, is a catheter angiogram.<sup>[19]</sup>

Doppler ultrasound may show reverse flow or arterialized flow within the SOV and asymmetric enhancement of cavernous sinuses and surrounding venous system.

Morphologic changes such as dilation of the SOV, cavernous sinus, and protrusion of the globe have been well demonstrated by contrast-enhanced CT and MR imaging. CT and MR angiograms are also able to depict abnormal vascular changes, including engorged venous sinuses or cortical vein drainage, arterial or venous aneurysm formation, and arterial dissection. The differences between the modalities in detecting CCFs, however, were dependent on the location of the fistula.<sup>[20]</sup>

It is important to distinguish between direct (type A) and indirect (Types B-D) fistulas because of the prognostic implications. Urgent treatment is usually needed for direct and high flow fistulas in which endovascular embolization is mostly applied. Indirect, slow flow CCFs usually close spontaneously without treatment.<sup>[2,8,11,13]</sup>

Among endovascular treatment of direct CCFs and symptomatic indirect CCFs, transarterial embolization is a more common approach. Transvenous embolization is employed in the presence of multiple arterial feeders and inability to occlude indirect CCFs by the arterial route. The inferior petrosal sinus is the most common route in transvenous approach. If the sinus is impassable, alternative routes are the pterygoid venous plexus, superior petrosal sinus, facial vein, or ophthalmic veins.<sup>[21]</sup>

MR Imaging findings of Direct CCF in our case- include proptosis, engorgement of the superior ophthalmic veins, cavernous sinus distention, and abnormal flow voids within the cavernous sinuses.

In our Case, direct CCF in a male patient, was attributed to previous history of trauma and drainage pathway were toward anterior via ophthalmic veins. Our findings so far are in accordance with the literature.

Our patient underwent coil embolisation of the right internal carotid artery. During the procedure, a high flow, expansive connection between the artery and the cavernous sinus and collateral veins was noted. Post procedure was uneventful.

### CONCLUSION

Non-invasive MR imaging is helpful for diagnosing CCF's, by depicting uni- or bilateral exophthalmus, orbital congestion, dilation of SOV, arterialization of the surrounding venous structures appearing as flow void on MRI or flow related signal on MRA.

Although non-invasive radiologic techniques can aid in the diagnosis of the CCFs, DSA is still a gold standard for diagnosis, valuable for categorization of CCFs and verification of their drainage patterns and allows for the planning and performance of the therapeutic procedure.

### Consent

The patient has given consent to present the case and for the use of images of diagnostic procedures.

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