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A rare case of caroticocavernous fistula

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Abstract
Carotico cavernous fistulae (CCF) are rare, abnormal arteriovenous fistulae (AVF) connecting the carotid arterial system and cavernous sinus (CS) [1, 2]. The increase in cavernous sinus pressure resulting from this is the main reason for the clinical features and complications of this entity [3]. The classic triad of Dandy includes chemosis, pulsatile proptosis and ocular bruit. Imaging plays a key role in the diagnosis of a CCF. Use of multimodality approach including orbital doppler, CT, MRI with respective angiography techniques help in the diagnosis. However, digital subtraction angiography (DSA) is the gold standard investigation for diagnosis of CCF as it can identify fistula angioarchitecture whilst allowing simultaneous planning and intervention. Transarterial or transvenous embolization of the fistula by endovascular intervention is the first line treatment modality for the treatment of most CCFs.

Keywords: Carotico cavernous fistula, arteriovenous fistula, barrow's classification, digital subtraction angiography, endovascular intervention

Introduction
Carotico cavernous fistulae (CCF) are rare, abnormal arteriovenous fistulae (AVF) connecting the carotid arterial system and cavernous sinus (CS) [1, 2]. The increase in cavernous sinus pressure resulting from this is the main reason for the clinical features and complications of this entity [3]. The classic triad of Dandy includes chemosis (red eye), pulsatile proptosis (abnormal protrusion of the eyeball) and ocular bruit (blood flow sounds coming from the eye) [4]. It can be associated with head ache, nausea, vomiting, blurring of vision and diplopia. Direct fistulas are more common than the indirect fistulas. They are usually due to trauma but sometimes can be spontaneous. About 0.2% of patients with traumatic brain injuries go on to develop direct fistulas [5]. The demographics of direct CCF reflect the distribution of head trauma, most commonly seen in young male patients. The presentation is acute and symptoms develop rapidly. In contrast, indirect CCF has a predilection for postmenopausal female patient and the onset of symptoms is often insidious [6].
We present a rare case of spontaneous direct carotico cavernous fistula with aneurysm of cavernous part of internal carotid artery.

Materials and methods
A 20 year old male presented to the emergency OPD with complaints of severe right sided headache, nausea, vomiting, blurring of vision in right eye since 5 days and drooping of right upper eyelid since one day. He is a known case of migraine since 10 years on irregular medication. Similar episodes of headache associated with nausea and vomiting were treated with migraine medications but results were not satisfactory. No history of trauma and not a known case of hypertension.
On examination, there was complete ptosis of right eye with conjunctival congestion, mild proptosis and mild restriction of extra ocular movements in all nine gazes (Fig - 1). Visual acuity was 6/18 with no pinhole improvement. Intra ocular pressure was raised with IOP of ~ 28 mmHg. Further, B-scan of right eye and contrast enhanced CT of brain with angiography was advised.
B-scan of right eye showed mild dilatation (~ 3.5 mm) of superior ophthalmic vein (SOV) with arterialized flow and raised velocity (Fig - 2).

Fig 1: Complete ptosis of right eye with conjunctival congestion, mild proptosis and mild restriction of extra ocular movements.

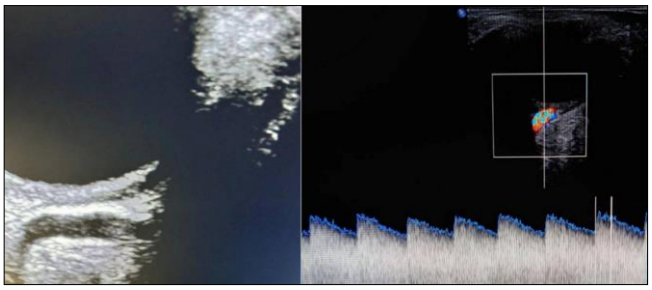

Fig 2: B-scan of right eye showing dilated SOV with arterialization of flow and raised velocity of SOV

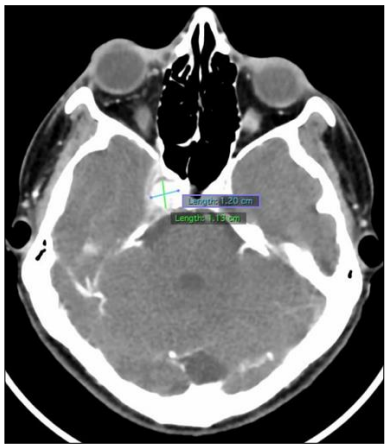

Fig 3: Axial CECT brain showing enlarged and dilated right cavernous sinus with intense homogenous enhancement in arterial phase, similar to that of carotid arteries with saccular dilatation of cavernous part of right ICA within the cavernous sinus

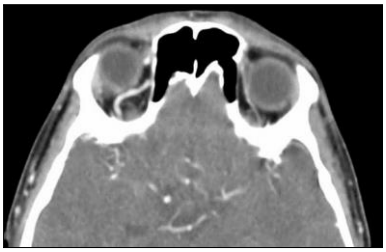

Fig- 4: Axial CECT brain showing dilated, tortuous right superior ophthalmic vein and cortical veins of right temporal lobe

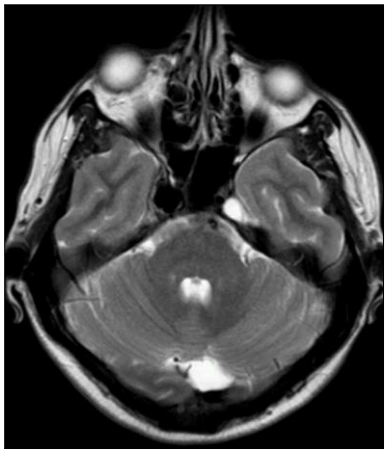

Fig 5: Axial T2WI showing prominent flow void in right cavernous sinus, suggestive of internal carotid artery aneurysm with dilated right cavernous sinus

On CECT brain with angiography, right cavernous sinus appeared enlarged and dilated with intense homogenous enhancement in arterial phase, similar to that of carotid arteries with saccular dilatation of cavernous part of right internal carotid artery (ICA) within the cavernous sinus (Fig - 3).

Dilated, tortuous right superior ophthalmic vein with dilated cortical veins of right temporal lobe (Fig- 4) and dilated right pterygoid plexus of veins showing enhancement in early arterial phase was noted. Proptosis of right eye with mildly bulky and edematous right extraocular muscles was noted.

The above features were suggestive of right internal carotid artery aneurysm with suspicious communication between cavernous part of right internal carotid artery and right cavernous sinus leading to carotico-cavernous fistula (Barrow's type A) and causing dilatation of right superior ophthalmic vein, right cortical veins and right pterygoid plexus of veins.

MRI brain showed prominent flow void in right cavernous sinus, suggestive of internal carotid artery aneurysm with dilated right cavernous sinus (Fig - 5).

Right superior ophthalmic vein appeared dilated and tortuous with few prominent cortical veins in right temporal region.

Right optic nerve appeared mildly enlarged with mildly bulky and edematous right extra ocular muscles. Right Meckel's cave, basal cistern and perioptic sleeves appear prominent and hyperintense on T2 weighted images, suggestive of raised intra cranial pressure.

In keeping with the above findings, diagnosis of right internal carotid artery aneurysm with suspicious communication between cavernous part of right internal carotid artery and right cavernous sinus leading to carotico-cavernous fistula (Barrow's type A) was made.

The patient was advised digital subtraction angiography (DSA) for confirmation, exact localization and characterization of the carotico-cavernous fistula.

Discussion

Carotico cavernous fistulas are abnormal, spontaneous or acquired arterio venous communications between the carotid circulation and the CS, resulting in increased CS pressure and siphoning of the cerebral arterial supply [7]. They are classified as direct (Barrow type A) and indirect (Barrow types B, C and D) based on the anatomic features seen on angiograms [8]. Further, they are also categorized according to etiology (spontaneous or post-traumatic) and hemodynamic effects (high or low flow) [3]. The angiography findings form the basis of Barrow's classification and are helpful to plan a therapeutic treatment strategy [3].

The high flow fistulas with direct communication between the intra-cavernous ICA and CS are direct fistulas and are classified as type A, while those fistulas with

communication between the meningeal branches of ICA, ECA or both and CS are classified as types B, C and D respectively and these are low flow indirect fistulas [9]. Direct CCFs are most commonly traumatic in origin but may also be caused due to rupture of internal carotid artery aneurysm into the cavernous sinus, Ehlers–Danlos syndrome type IV or iatrogenic intervention. Indirect CCFs occur due to hypertension, Ehlers–Danlos type IV, fibromuscular dysplasia and dissection of the internal carotid artery [8].

CCFs have a wide spectrum of clinical presentation depending on the type, size, location, flow, route of venous drainage and duration of the fistula [5]. The presenting features involve ocular symptoms, cranial nerve neuropathies, headache or pulsatile tinnitus. Fistulas may complicate and result in intracranial hemorrhage and ischemia. Smaller fistulas may remain asymptomatic. Complete ophthalmologic workup is necessary and includes visual acuity, pupillary function, intraocular pressure, fundoscopy (direct and indirect) and gonioscopy [8].

Imaging plays a key role in the diagnosis of a CCF. Orbital doppler sonography may show arterialization with low resistance flow, reversal of flow or thrombosis of superior ophthalmic vein. CT and MRI with angiography reveal dilated tortuous superior ophthalmic vein (SOV), enlarged ipsilateral CS with bulging of its lateral wall, proptosis and extra-ocular muscle thickening. Non-contrast cranial CT scan is helpful in detecting skull fractures. Orbital edema and abnormal flow voids in CS corresponding to shunted blood are better seen on MRI. 3D TOF MRA detects CCF by depicting flow-related enhancement in the involved CS and the arterial feeders in an indirect fistula [9].

Digital subtraction angiography (DSA) is the gold standard investigation for diagnosis of CCF as non-invasive imaging does not exclude the diagnosis of CCF and the ability of DSA to identify the angioarchitecture of the fistula which helps in planning and intervention. Assessment of the arterial supply in low flow fistulas, breech in the vessel wall in high-flow fistulas and venous drainage pattern can be done at DSA. Early contrast enhancement of the affected CS usually indicates the presence of a fistula. Specific assessment for cortical venous drainage at angiography is needed as urgent treatment is indicated, if present, because of its association with potentially life-threatening sub-arachnoid/intracerebral hemorrhage [5].

The aim of the treatment is to occlude the fistula completely while normal flow of blood through the ICA is preserved. Transarterial or transvenous embolization of the fistula by endovascular intervention is the treatment of choice for most of the CCFs. Transvenous access to cavernous sinus is preferred when the CCF originates from branches of the ICA as transarterial embolization is difficult and carries risk of stroke due to embolic reflux into ICA. Transarterial access is used in few cases of direct fistulas and when the CCF originates from branches of the ECA. The fistula can be occluded using either metallic coil or liquid embolization of the cavernous sinus. Few complications of endovascular treatment are cerebral infarction, decreased visual acuity, ophthalmoplegia, diabetes insipidus and femoral vein thrombosis. Other complications include subarachnoid or intracerebral hemorrhage, sinus rupture, extradural extravasation of contrast and cranial nerve palsies. Transient worsening of symptoms after cavernous sinus embolization is seen in few patients which usually resolves overtime. Most of the patients undergoing endovascular treatment for direct and indirect CCFs ususally experience complete cure. This can be seen clinically by reversal of the signs and symptoms of the CCF and angiographically by obliteration of the fistula with reversal of retrograde cortical venous flow [11].

Conclusion

Direct CCFs are unlikely to have spontaneous closure and upfront treatment is needed due to risk of neurological deficits and worsening of venous congestion. Indirect CCFs that fail conservative therapy or demonstrate progression of symptoms can be considered for endovascular treatmentet [12]. Although DSA is the investigation of choice, high suspicion on CT/ MR angiography helps us clinch the diagnosis of CCF. Endovascular intervention by transarterial or transvenous embolization of the fistula is the treatment of choice for most of the CCFs.

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