Unilateral Orbital Proptosis Secondary To A Carotico-Cavernous Fistula: A Case Report

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Citation

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Abstract

Carotico- Cavernous Fistulas (CCF) are caused by the laceration of the intercavernous portion of the carotid artery, or one of its intercavernous branches resulting in arterialisation of blood within the cavernous sinus. There are four types (A,B,C,D) of CCF out which type A (Direct type) and types B, C, and D (Indirect types) occur spontaneously. Prompt diagnosis and treatment of this condition can prevent visual loss, development of secondary glaucoma and intracranial hemorrhages.A unilateral orbital proptosis associated with sinusitis is an indication for endoscopic decompression. In our case even though there was unilateral orbital proptosis along with sinusitis, endoscopic surgery was unnecessary and did not significantly effect the course of patients illness. We present a case of unilateral orbital proptosis in a middle aged male with an indirect type of CCF detected on Magnetic Resonance Angiogram. The role of MR imaging and indications for endoscopic surgery in a unilateral orbital proptosis with review of relevant literature is discussed in this paper.

INTRODUCTION

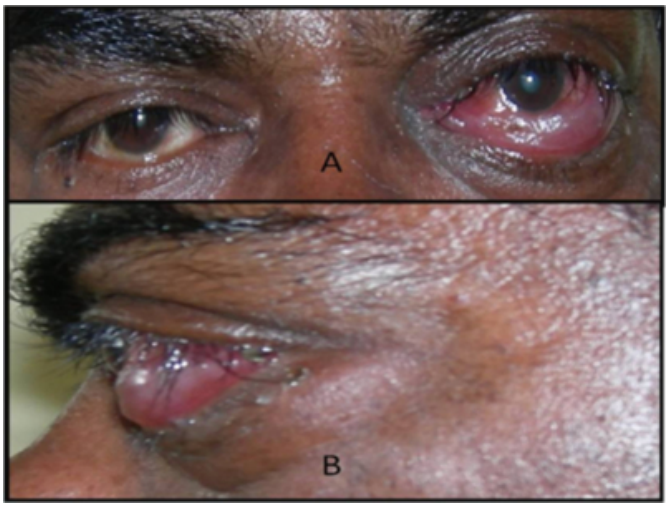
Carotico- Cavernous Fistulas (CCF) are caused by the laceration of the intercavernous portion of the carotid artery, or one of its intercavernous branches resulting in arterialisation of blood within the cavernous sinus.[1] It can occur spontaneously (25%) or following trauma (75%).[3] There are four types (A,B,C,D) of CCF out which type A (Direct type) occurs in traumatic cavernous arteriovenous fistulae where the fistula is fed directly by the trunk of the internal carotid artery. This is seen predominantly in young males.[4] Types B, C, and D (Indirect types) occur in spontaneous cavernous dural arteriovenous fistulae. They are more common in women more than 50 years, with a 1: 7 male to female ratio.[5] We present a case of unilateral orbital proptosis in a middle aged male with an indirect type of CCF detected on Magnetic Resonance Angiogram. The role of MR imaging and indications for endoscopic surgery in a unilateral orbital proptosis with review of relevant literature is discussed in this paper.

CASE

A 54 year old male presented to the Otolaryngology outpatient department at a tertiary referral centre in Bangalore, India with a history of painless proptosis of the left eye associated with non progressive decrease in vision since 5 months [Fig 1].

Figure 1

Figure 1: Left axial proptosis and chemosis of the left eye & Left axial proptosis in lateral view.

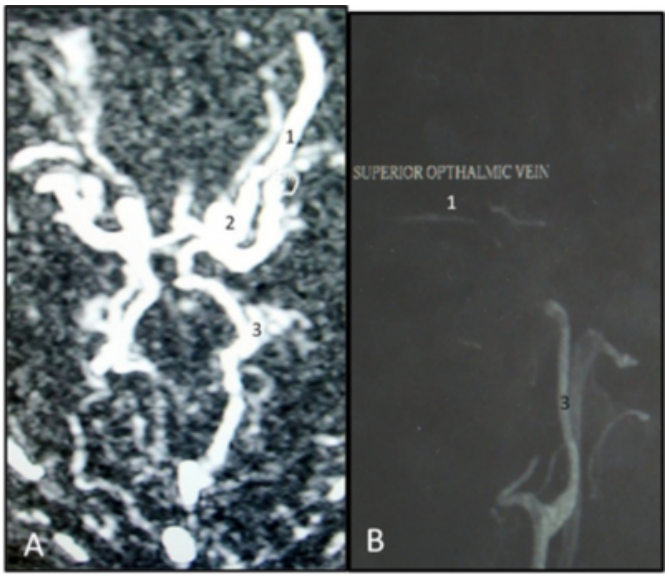


Old records show that he underwent a functional endoscopic sinus surgery 3 months ago for an initial suspicion of a complication secondary to left maxillary and ethmoidal sinusitis. However, after the surgery the proptosis persisted. CT and MRI reports from the past indicated proptosis of the left orbit with thickening of left extra ocular muscles and prominent left superior ophthalmic vein. On re-evaluation at our centre, on examination an evident left orbital proptosis with chemosis and epiphora associated with restricted

movements of the left eye was noted. Vision was 6/24 in the left eye with no loss of color vision. A repeat MRI and MR Angiogram with gadolinium contrast (a paramagnetic substance which enhances on T_1) was taken that revealed left superior ophthalmic vein opacification in arterial phase indicating a small carotid cavernous fistula on the left side in addition to the earlier mentioned findings.

Figure 2

Figure 2 Â– A-Magnetic resonance imaging(MRI) post contrast venogram showing the contrast from the left superior ophthalmic vein(1) into the internal carotid artery(3) through the carotico cavernous fistula(2) & B-Dynamic post gadolinium contrast study showing contrast enhancement of superior ophthalmic vein(1)in a post contrast study signifying a communication from the internal carotis adrery(3)



In view of the vision loss, intravenous glucocorticosteroids were administered while waiting for definitive diagnostic studies and management. The definitive management of a caroticocavernous fistula is obliteration of the fistulous connection with restoration of normal arterial and venous flow. This was achieved through an endovascular approach which was performed by an intervention radiologist along with a neurosurgeon at the national institute of neurosciences in Bangalore. After complete delineation of the fistulous tract, an approach to close the fistula was planned and performed. This fistula was approached through the internal carotid artery. A detachable balloon was positioned to occlude the fistula while maintaining patency of the internal carotid artery. The patient recovered following the procedure and is maintaining continuous follow-up.

DISCUSSION

Carotico- Cavernous Fistulas (CCF) are caused by the laceration of the intercavernous portion of the carotid artery, or one of its intercavernous branches resulting in arterialisation of blood within the cavernous sinus.[1]A CCF thus allows shunting of blood from the high pressure internal carotid artery to a low pressure system, usually resulting in the engorgement of the opthalmic venous system with congestion and edema of the orbital tissues.[2] It can occur spontaneously (25%) or following trauma (75%).[3] Based on the etiology, there are several entities responsible for a CCF such as closed or penetrating head trauma, Surgical trauma, rupture of an intracavernous aneurysm, connective tissue disorders, vascular disease and dural fistulas.[6,7]

Clinically, cavernous dural arteriovenous fistulae are classified into four types, namely Type A,B,C,D on basis of the fistula being fed by the main trunk of the internal carotid artery, branch of the internal carotid artery , branch of the external carotid artery and branches of both the internal and external carotid arteries respectively.[4] The type A (direct type) occurs in traumatic cavernous dural arteriovenous fistulae, and is more common in young males.[4] Types B, C, and D (Indirect types) occur in spontaneous cavernous dural arteriovenous fistulae. They are more common in women more than 50 years, with a 1: 7 male to female ratio.[5]Clinical Findings of Indirect fistulae are mild compared to direct ones, and they progress slowly. The classical triad of chemosis, pulsatile exopthalmos and ocular signs are only observed in direct fistulae. Patients may also present with chronic red eye, diplopia, headache and opthalmoplegia. CCFs from cavernous carotid aneurysm can have an audible pulsatile bruit.8 The CCF may also present with massive and life threatening epistaxis.[9]

Indirect fistulae improve spontaneously in 20-50% of the cases. Interestingly they may regress spontaneously after angiography. Unfortunately in 20-30% of the cases visual loss can result as a consequence if they are left untreated. On the other hand, symptomatic direct fistulae almost always require immediate treatment.[10]Indications for management are proptosis, visual loss, abducens paresis, severe pain, angiographically increased corticovenous filling, and increased intracranial pressure.[10]

A carotid cavernous fistula can be confirmed on an MR Angiogram. Imaging modality preferred for orbital proptosis secondary to sinusitis is Computerised Tomography of the Para nasal sinuses with orbital cuts. However the role of

Magnetic Resonance Imaging [MR] and Venography/Arteriography in orbital proptosis secondary to sinusitis is unclear with insufficient literature regarding its application.

The aims of the treatment are to correct the orbital signs and symptoms, to prevent neurological damage due to pressure effects within the cavernous sinus, and to avoid subarachnoid hemorrhage or catastrophic epistaxis. [2]Observation alone may be considered for a spontaneous fistula in an older patient, but traumatic fistulas require active management. Treatment options have included ligation of the internal carotid artery, with or without muscle embolisation and direct surgical closure of the fistula.[2] The use of detachable intravascular balloon has curently become the treatment of choice. [11]Prompt diagnosis and treatment can prevent visual loss, development of secondary glaucoma and intracranial hemorrhages. [12]However on many occasions a unilateral orbital proptosis associated with sinusitis is an indication for endoscopic decompression. In our case even though there was unilateral orbital proptosis along with sinusitis, endoscopic surgery was unnecessary and did not significantly effect the course of patients illness.

CONCLUSION

A unilateral painless orbital proptosis which is not attributed to complications of acute sinusitis or intraorbital causes should be considered for Magnetic resonance imaging and Magnetic resonance angiogram to rule out conditions such as a carotid cavernous fistula. An unnecessary endoscopic sinus surgery and decompression can be avoided which may not improve the course of patients illness if a fistula is detected and an accurate treatment modality can be selected without expenditure of time to avoid complications. The need to detect CCF at the earliest is therefore necessary to prevent complications such as catastrophic epistaxis, visual loss, development of secondary glaucoma and intracranial hemorrhages.

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