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## Diagnostic Dilemma of Dural Carotid Cavernous Fistula

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**Abstract-** We report a case of dural carotid cavernous fistula (CCF) pose a diagnostic dilemma with initial symptoms of the arteriovenous shunt. A 56 year-old man presented with right eye diplopia, then subsequently developed ptosis, congestion of conjunctiva, dilated episcleral vessels, and gradual rise in intraocular pressure. Initial diagnosis of pseudotumour was made based on negative finding of CCF by computed tomography angiography (CTA). In view of persistent clinical manifestations in spite of steroid therapy, and with the presence of new ocular signs; cock-screw conjunctival vessels, dilated retinal veins, and proptosis, digital subtraction angiography (DSA) was performed and confirmed the diagnosis of dural CCF. The ocular symptoms resolved completely post embolization of the fistula.

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### 1 INTRODUCTION

Dural carotid cavernous fistula (CCF) is a rare condition characterised by a communication between the cavernous sinus and one or more meningeal branches of the internal carotid artery (ICA), external carotid artery (ECA) or both [1]. The classical triad of pulsating exophthalmos, conjunctival chemosis, and pulsatile tinnitus [2,3] are well established clinical symptoms of the disease but are not usually present in majority of patients as early indicators [4]. Dural shunts have been misdiagnosed as chronic conjunctivitis, episcleritis, orbital cellulitis, orbital pseudotumour or thyroid eye disease [4,5].

We illustrate the dilemma in arriving to a correct diagnosis in the absence of characteristic ocular signs and absent positive imaging finding; hence the importance of a revision of diagnosis when there is no improvement of patient's condition.

### 2 CASE REPORT

A 56 year-old man with no known medical illness presented to ophthalmology clinic with right horizontal diplopia for 2 days duration. Four weeks later, the diplopia was associated with right eye ptosis and generalised eye redness. The patient also complained of headache. However, there were no symptoms of raised intracranial pressure such as nausea and vomiting. Vision was 6/6 in both eyes. Right eye examination

showed features of incomplete right 6<sup>th</sup> nerve palsy shown by mild restriction in abduction and 3<sup>rd</sup> cranial nerves palsy seen as mild ptosis, diffuse conjunctival congestion with dilated episcleral vessels (Figure 1). Intraocular pressure were within normal limits in both eyes. There were no proptosis and papilloedema. Other cranial nerves examination were normal with no abnormality detected in the left eye. Computed tomography (CT) scan of the brain and orbit showed mild proptosis of the right orbit with bulkiness of right recti muscle and dilated right superior ophthalmic vein. There was no space occupying lesion noted.



**Figure 1:** Right eye shows dilated episcleral vessels and diffuse conjunctival congestion.

Hence, a provisional diagnosis of thyroid eye disease was made based on CT scan finding of bulkiness of recti muscle with an orbital pseudotumour as a differential diagnosis. CCF was also in the list of differentials due to presence of dilated right superior ophthalmic vein however there were absence of risk factors such as hypertension, arteriosclerosis, connective tissue disease and head trauma. Furthermore other clinical signs of CCF were also absent especially the classical triads. The systemic features of thyroid dysfunction and blood investigation for thyroid function test values were in the normal range. Therefore, the diagnosis was revised to orbital pseudotumour and planned to treat with oral prednisolone.

Before the initiation of oral prednisolone, his right visual acuity reduced to 6/9. The intraocular pressure raised to 26 mmHg in the right eye and 19 mmHg in the left eye. In view of features suggestive of early optic nerve compression, intravenous methylprednisolone 250 mg 6 hourly for 3 days was administered along with topical timolol 0.5% eye drop twice a day for the right eye. There was improvement of his visual acuity to 6/6 on day 2 of intravenous methylprednisolone. Right eye examination showed resolution of conjunctival congestion but persistent dilated episcleral vessels with new signs of cock-screw vessels of the conjunctiva and dilated retinal vein of the right eye. A revised diagnosis of CCF was made with the presence of new ocular findings. Further assessment of the orbit showed absence of orbital bruit. Urgent CT angiography (CTA) of the brain was performed and revealed dilated right superior ophthalmic vein but abnormal flow voids in cavernous sinus was not detected to support the diagnosis of CCF. His treatment was continued with oral prednisolone 40 mg 12 hourly (1 mg/kg/weight) for 3 weeks and then tapered down 5 mg per week over the period of 6 weeks. An anti-inflammatory dose of steroid 1.0 to 2.0 mg/kg/day are usually started and continued with slow tapering when improvement is noted [6].

His condition improved temporarily after completion of steroid therapy. There was complete resolution of 3<sup>rd</sup> cranial nerve palsy with residual 6<sup>th</sup> cranial nerve palsy. Two days after completion of his oral steroid, he experienced another attack of optic nerve compression with reduced vision, impaired optic nerve function, proptosis, conjunctival congestion and raised intraocular pressure of the right eye. He was treated with second cycle of intravenous

methylprednisolone and continued with tapering dose of oral prednisolone. However, there was persistence of ocular findings. In view of temporarily response to steroid therapy, residual right 6<sup>th</sup> cranial nerve palsy, proptosis, cock screw vessels and dilated retinal vein, a differential diagnosis of low flow CCF was reconsidered. Retrospectively, history of motor vehicle accident 30 years ago with right lower limb injury was discovered. However, the patient was unable to recall any history of head or eye injury.

A diagnostic cerebral angiogram, digital subtraction angiography (DSA) was performed and showed features of dural CCF. There was communication between the cavernous sinus and the right middle meningeal artery (branches of right ECA) and also communication of the cavernous sinus with bilateral meningo-hypophyseal trunk (from bilateral ICA). Finally, the patient was diagnosed to have dural CCF. He was successfully managed with embolization of fistulous communication with diluted glue and showed complete resolution of ocular symptoms within 24 hours post embolization.

### 3 DISCUSSION

Dural CCF are characterised by a communication between the cavernous sinus and one or more meningeal branches of ICA, ECA, or both. Several small branches of the cavernous portion of the ICA (tentorial, dorsal meningeal and inferior hypophyseal arteries) traverse the cavernous sinus to anastomose with meningeal branches of the external carotid and vertebral arteries [7].

These fistulae usually have low rates of arterial blood flow and almost always produce symptoms and signs spontaneously, without any antecedent trauma and commonly seen in middle aged to elderly women with atherosclerosis, systemic hypertension, collagen vascular or connective tissue disease [7]. However our patient was medically fit and he had developed delayed ocular features of dural CCF possibly secondary to trauma. The common signs are mild dilatation of conjunctival and episcleral veins without chemosis, ipsilateral proptosis, rise in intraocular pressure, inconstant often high pitched and focal bruit heard over ipsilateral closed eye or temple. The transient 6<sup>th</sup> nerve palsy and unilateral headache usually antedate orbital signs by few months [7].

The classical triad of pulsating exophthalmos, conjunctival chemosis, and pulsatile tinnitus [2,3] are well established clinical symptoms of the CCF, but are not usually present

in majority of patients as early indicators [4]. Dural CCF patients usually do not show the classical triad, rarely dural CCF that drain posteriorly presents with only cranial nerve palsies [4]. However, some patients with dural CCF may have more severe symptoms such as proptosis, ophthalmoplegia, visual impairment or intracranial haemorrhage [8]. Patients with dural CCF are often diagnosed with delay, because the clinical symptoms seem to be usually mild and non-specific. In addition, spontaneous occlusion of the fistula may occur [1]. Patients often have been initially misdiagnosed when the classical triad is missing. Dural shunts have been misdiagnosed as chronic conjunctivitis, episcleritis, orbital cellulitis, orbital pseudotumour or thyroid eye disease [4,5,9]. In our case, he was treated as pseudotumour initially due to absence of classic triad; pulsating exophthalmos and pulsatile tinnitus and overlapping clinical features; conjunctival congestion.

Orbital pseudotumour or idiopathic orbital inflammation is an uncommon disorder characterised by non-neoplastic, non-infective, space occupying orbital infiltration with inflammatory features. Periocular redness, swelling and discomfort, reduce vision, diplopia are the symptoms and they can mimic CCF [6]. Idiopathic orbital inflammation typically responds favorably to systemic corticosteroid treatment [10]. Although our patient had responded well to the initial treatment, there was reduced sensitivity towards corticosteroid therapy on subsequent attack, hence revision of diagnosis was made.

Traumatic CCF involves either direct communication between the ICA and the cavernous sinus or between dural branches of the ICA or ECA and the cavernous sinus [7]. The angiographic demonstration of a traumatic CCF occurs usually within weeks or months after trauma [11]. Our patients was medically fit and presumed to have traumatic dural CCF with delayed presentation as he had history of trauma 30 years ago. Symptoms of traumatic dural CCFs are essentially the same as those of spontaneous CCFs.

Non-invasive imaging techniques are used in initial evaluation of CCF such as Doppler ultrasound, CT scan, magnetic resonance imaging (MRI) [12] and CTA [13]. In CCF, there is retrograde drainage from the cavernous sinus and appear as early filling of superior ophthalmic vein on CTA. However, dilated superior ophthalmic vein can be found in other disorders such as thyroid eye disease, orbital

pseudotumour and parasellar meningioma [14]. In our patient, the only findings of CCF in his initial CT scan was dilated right superior ophthalmic vein. CTA showed similar finding of dilated right superior ophthalmic vein but failed to evaluate dural fistulas. It is important to note that normal results in these tests do not exclude the diagnosis of CCF [7]. The dural fistulas usually have low rates of arterial blood flow and may be difficult to diagnose without invasive angiography and can be missed. In cases with variety of characteristic signs, higher index of diagnostic suspicion is essential. An invasive imaging is diagnostic and can be therapeutic for CCF. Finally DSA, an invasive imaging, confirmed the presence of CCF in our patient.

The severity of the condition versus the possibility of spontaneous resolution should be taken into account, especially in dural forms [15,16]. Dural CCF may close spontaneously, but, for those lesions causing progressive or unacceptable symptoms and signs, standard endovascular embolization performed with synthetic and natural materials such as absorbable gelatin (Gelfoam); Silastic; platinum coils; low-viscosity silicone rubber; autogenous clot, muscle, or dura; tetradecyl sulfate (a sclerosing agent); polyvinyl alcohol particles (Ivalon); ethanol; oxidized cellulose (Oxycel); and isobutyl-2-cyanoacrylate glue (Bucrylate) [15]. Other treatment options are irradiation, stent placement and some difficult cases require surgical techniques (eg, sinus isolation and resection) in combination with interventional procedures. Recent studies of stereotactic radiation therapy showed a relatively high occlusion rate (70%-88%) several months after treatment without significant complication. Higher occlusion rate (80% - 100%) was seen immediately after the endovascular embolization with glue; however, serious complications such as intracranial haemorrhage and cranial nerve deficits were also reported [17]. Our patient was successfully managed with embolization of fistulous communication with diluted glue and showed immediate complete resolution of ocular symptoms post embolization.

Majority of cases had been treated successfully. Clinical features usually begin to improve within hours to days after successful closure of a dural CCF and most patients have a normal or near-normal external appearance within 6 months [15]. Dural CCF have no direct cause of mortality. However, there were morbidities which are related to treatment and

ocular complications due to elevated venous pressure causing secondary glaucoma, central retinal vein occlusion, retinal detachment, choroidal effusion, optic disc swelling, ocular ischaemia leading to visual loss and exposure keratopathy caused by proptosis [15].

#### 4 CONCLUSION

Dural CCF should be considered in patients initially presented with limitation in abduction with generalized eye injection but not fulfilling the classical symptoms of pulsating exophthalmos, conjunctival chemosis, and pulsatile tinnitus. Thus treated as other conditions utilising standard treatment protocols, however results in unsatisfactory outcome which should prompt clinicians to revise the initial diagnosis.

#### CONFLICTS OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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