A Case of Carotid-Cavernous Fistula

By:

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A 19 year old male patient came to our outpatient clinic, complaining of:

- Severe conjunctival redness,
- Pain,
- Photophobia,
- Watering,
- Exposure of his both eyes and
- Annoying aching sound in the head 2 weeks ago.
**History:**

**Medical**
irrelevant medical history.

**Surgical**
- history of car accident 2 months ago with hematoma in his face which resolved by the time of presentation.

**Examination:**

**General**
- Patient looks conscious, alert and cooperative.

**Local**
- Patient presented with bilateral pulsating exophthalmos.
Ophthalmological Examination

**Right Eye:**

**Visual Acuity**
6/60

**Eyelid**
edematous

**Conjunctiva**
Severe hemorrhagic chemosis

**Cornea**
Infected exposure keratitis with ulcer 2x5 mm

**Pupil**
Round reactive

**AC**
Hypopyon 2mm

**E.O.M**
Limited Abduction

**I.O.P**
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**Fundus**
Couldnot be assessed


Ophthalmological Examination

**Left Eye:**

**Visual Acuity**
1/60

**Eyelid**
Edematous

**Conjunctiva**
Severe hemorrhagic chemosis

**Cornea**
Exposure Keratitis

**Pupil**
Round slightly reactive with APD

**E.O.M**
Total External Ophthalmoplegia

**I.O.P**
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**Fundus**
Pale optic disc
Investigations:

- CT brain.
- CTA brain.
- US orbit.

Non-Contrast Axial CT Scan
Coronal CT Section With Contrast

CT Angiography
Coronal Section          Axial Section
CT Angiography With Digital Subtraction

US of Left Orbit Showing The Dilated Superior Ophthalmic Vein.
**Diagnosis:**

Left Direct High Flow Carotid-Cavernous Fistula with Intercavernous Communication

→ Bilateral Pulsating Exophthalmos

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**Treatment**

- **Treatment of the keratitis:**

  **Surgical** → Lt. Lateral Tarsorrhaphy.

  **Medical** → Rt. And Lt. eye drops:
  - Antibiotic.
  - Lubricant.
  - Cycloplegic.

- **Regular Follow Up.**

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- Patient prepared for neurosurgical operation.

Endovascular Coiling done at Neurosurgery Department.

10 days Postoperative
14 days
Postoperative
One Month Postoperative
2 Months Postoperative

Patient regained VA of 6/24 with inferior superficial vascularized corneal opacity and normal EOM and fundus in his right eye.
Although he regained normal EOM in his left eye, the vision did not improve over 1/60 because of the established optic atrophy.
**Carotid-cavernous fistula**

An arteriovenous fistula is an acquired abnormal communication between an artery and a vein. The blood within the affected vein becomes ‘arterialized’, the venous pressure rises, and venous drainage may be altered in both rate and direction.

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A carotid-cavernous fistula is one such communication, between the carotid artery and the cavernous sinus. Ocular manifestations occur because of venous and arterial stasis, increased episcleral venous pressure and a decrease in arterial blood flow to the cranial nerves within the cavernous sinus.

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Carotid-cavernous fistulae can be classified on the basis of:
(a) **aetiology** *(spontaneous and traumatic)*,
(b) **haemodynamics** *(high and low flow)* and (c) **anatomy** *(direct and indirect)*.

**Direct carotid-cavernous fistula**

Representing 50% of all cases, direct fistulae are high-flow shunts in which carotid artery blood passes directly into the cavernous sinus through a defect in the wall of the intracavernous portion of the internal carotid artery as a result of the following:
- **Trauma** is responsible for 75% with sudden and dramatic onset of symptoms and signs.

- **Spontaneous** rupture of an intracavernous carotid aneurysm or an atherosclerotic artery. Usually have lower flow rates and less severe symptoms.

**Presentation** may be days or weeks after head injury with the classic **triad** of pulsatile proptosis, conjunctival chemosis and a whooshing noise in the head.

**Signs** are usually ipsilateral to the fistula but may be bilateral or even contralateral because of the vascular connections across the midline between the two cavernous sinuses.
• Severe epibulbar injection.
• Ptosis (due to 3rd nerve).
• Haemorrhagic Chemosis.
• Pulsatile proptosis.
• Increased intraocular pressure.
• Anterior segment ischaemia.
• Ophthalmoplegia.

• Fundus examination may show optic disc swelling, venous dilatation and intraretinal haemorrhages.
Vision

Immediate visual loss may be due to ocular or optic nerve damage at the time of head trauma.

Delayed visual loss may occur as a result of exposure keratopathy, secondary glaucoma, central retinal vein occlusion, anterior segment ischaemia or ischaemic optic neuropathy.

investigations

CT and MRI
may demonstrate prominence of the superior ophthalmic vein and diffuse enlargement of extraocular muscles.

Definitive diagnosis involves CT and MR angiography.
Treatment

Most carotid-cavernous fistulae are not lifethreatening; the organ at major risk is the eye.

A post-traumatic fistula is much less likely to close on its own than a spontaneous one.

The current treatment of choice involves endovascular embolization with coils or balloons which may be transvenous or transarterial.

Indirect carotid-cavernous fistula

*('dural shunt'), the intracavernous portion of the internal carotid artery remains intact.

Arterial blood flows through the meningeal branches of the external or internal carotid arteries indirectly into the cavernous sinus.

Due to slow blood flow, the clinical features are more subtle than in a direct fistula.
Causes

- *Congenital malformations*, in which the onset of symptoms is precipitated by intracranial vascular thrombosis.

- *Spontaneous rupture*, especially in hypertensive patients, which may be precipitated by minor trauma or straining.

Presentation is with gradual onset of redness of one or both eyes caused by conjunctival vascular engorgement.
Signs are variable.
• Mild epibulbar injection with or without chemosis.
• Exaggerated ocular pulsation.
• ‘Corkscrew-like’ epibulbar vessels.
• Raised intraocular pressure.
• Mild proptosis.
• Ophthalmoplegia
• Fundus may be normal or manifest moderate venous dilatation.

Differential diagnosis
- Conjunctivitis.
- Thyroid eye disease.
- Glaucoma.
- Orbital arteriovenous malformations.
Treatment

if required, usually involves endovascular embolization, although not via the transarterial route.

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THANK YOU

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