Contents lists available at ScienceDirect

Journal of Acute Disease

journal homepage: www.jadweb.org

Document heading doi: 10.1016/S2221-6189(13)60099-2

Bilateral indirect carotid cavernous fistula post trivial injury- A case report

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ARTICLE INFO

Article history:
Received 25 November 2012
Received in revised form 15 January 2013
Accepted 15 March 2013
Available online 20 March 2013

Keywords:
Indirect carotid cavernous fistula
Proptosis
Corkscrew vessels
Venous stasis
Retinopathy

ABSTRACT

Fifty-seven years old Malay lady, post menopausal with co-morbid of diabetes mellitus and hypertension presented with three months history of bilateral painful red eyes associated with double vision. Examination revealed both eyes proptosis, corkscrew vessels with present of bruit, secondary narrow angle with raised intraocular pressure, 6th cranial nerve palsy, and bilateral venous stasis retinopathy. CT angiogram showed bilateral dilated superior ophthalmic veins with cerebral angiogram findings of bilateral indirect carotid cavernous fistula involving small meningeal vessels. Indirect or dural cavernous sinus Fistula can easily be missed or misdiagnosed. Trivial injury especially in the predisposing patient can initiate the occurrence.

1. Introduction

Carotid cavernous fistulas (CCFs) are typically divided into direct and indirect types of arteriovenous shunts. Direct CCF is a communication between the cavernous segment of the internal carotid artery and the cavernous sinus, usually due to trauma or ruptured aneurysm. Indirect CCF is an acquired communication between dural (meningeal) branches of the internal or external carotid arteries through the wall of the cavernous sinus. The clinical manifestations may overlap, although the natural history is often significantly different. We report the indirect CCF post trivial injury which resolved without surgical intervention.

2. Case report

A 57 years old Malay lady was referred to us with complaint of bilateral generalized eye redness and tearing for three months duration. It was associated with periorbital swelling and fullness but no periorbital redness. Initially patient claimed it was painless but later she started to develop pain and described the pain as dull ache pain. However the pain does not awaken her from sleep and there was no pain during eye movement. No history of headache, nausea or vomiting. Patient also complained of mild blurring of vision, double vision and hearing whooshing sounds about one month prior to presentation.

On further questioning, patient gave history of trivial injury where she involved in motor vehicle accident two months prior to symptoms. Her head was knocked over the dashboard during that time. However there was no history of loss of consciousness or head injury. She denies history of eye discharge. No history of fever or unwell. No history of ocular surgery. No loss of appetite or loss of weight, no heat intolerance, no tremors or palpitation. There were no lethargic, no proximal muscle weakness and no neck swelling.

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Patient was a known case of diabetes mellitus and hypertension for more than twenty years under follow up and claimed the medical status were well control. She was post menopausal but not on hormone replacement therapy. She was non smoker and had history of allergy to sea foods but no allergy to any medicines.

On examinations, there were bilateral fullness of periorbital region with slight axial proptosis and left mild ptosis (Figure 1). Exophthalmometer measurement were 22 mm for both eyes at interpalpebral measurement of 120 mm. There were present of bruits over both eyes. There was no lid lag, no red or light desaturation and Ishihara test was normal for both eyes. Her right eye vision was 6/24 correctable to 6/18 with pin hole and left eye vision was 6/18 correctable to 6/12 with pin hole.

There was no relative afferent pupillary defect and no anisocoria noted. Both conjunctiva were generally congested with present of corkscrew vessels (Figure 2). Cornea was clear with intact corneal sensation. Right anterior chamber was shallow and quiet and left eye was moderately shallow and quiet. Both eyes showed mild nucleosclerosis cataract. The intraocular pressure of the right eye was 30 mmHg and left eye was 26 mmHg with Goldman Applanation Tonometer.

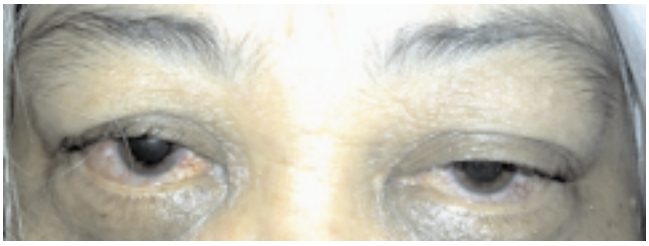

Figure 1. Bilateral periorbital fullness with proptosis and left mild ptosis.

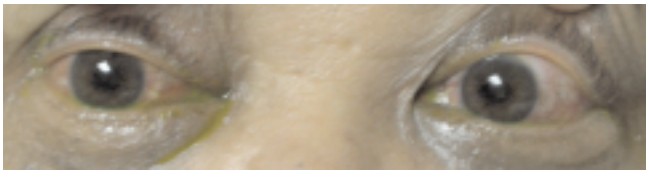

Figure 2. Generalized conjunctival congestion with corkscrew vessel.

Extraocular muscles movement showed some limitation. Movement on abduction of both eye were −3, elevation in abduction and depression in abduction were −2. Adduction of left eye was −2, but full for right eye. Elevation of both eyes were −3 and depression of both eyes were normal (Figure 3).

Gonioscopy of the right eye showed grade 2−3 in all quadrant with no peripheral anterior synaechiae but present of blood in Schlemms canal. Left eye gonioscopy showed grade 1 nasally, grade 2−3 in superior, temporal and inferiorly with present of blood in Schlemms canal. Both eyes vitreous were clear, disc pink and no disc swelling. Cup disc ratio was 0.4 in both eyes. Fundus examinations revealed both eyes vessels were slight dilated and tortuous with mid-peripheral blot haemorrhages. There was no cotton wool spots (Figure 4). Systemic examinations revealed stable vital signs. There was no neck swelling, no excessive sweating, no proximal myopathy and no fine tremor.

On auscultation, the lungs were clear with equal air entry. Cardiovascular was normal. Other cranial nerves examinations were normal.

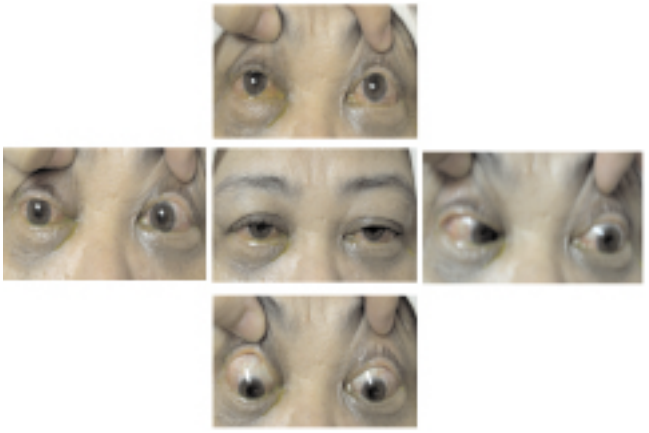

Figure 3. Limitation of all extraocular muscle movement except depression.

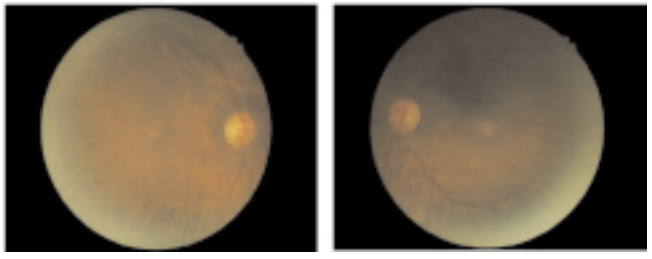

Figure 4. Multiple blot haemorrhages at mid periphery on both eyes secondary to venous stasis and mild venous dilatation.

Several investigations were done. B scan shows retinal flat with no choroidal effusion, no sclera thickening or T sign. Full blood count result was normal except for borderline Hb level of 11.9 g/dL, Random blood sugar was 11.9 mmol/L, Renal profile and thyroid function test were normal. Fasting serum lipid showed borderline result. ECG was normal and Chest X ray showed borderline cardiomegaly. CT brain and CT Angiogram then was performed and result showed bilateral dilated and engorged superior ophthalmic veins with prominent

cavernous sinus and normal extraocular muscles (Figure 5). Both internal carotid arteries are preserved. Calcification noted at the cavernous and intracranial portion of both internal carotid arteries. The petrous portion is preserved. Both vertebral, basilar and posterior cerebral arteries are normal. Both posterior communicating arteries are patent. Both globes are proptosed and the recti muscles are preserved. No intraconal or extraconal mass.

Patient was diagnosed to have bilateral carotid cavernous fistula with bilateral 6th cranial nerve involvement, left 3rd nerve involvement, secondary narrow angle with raised intraocular pressure and venous stasis retinopathy. Her intraocular pressure was well control with three anti-glaucoma drops and she was referred to neurosurgical team in other hospital.

Cerebral angiogram was done and findings showed bilateral indirect carotid cavernous fistula involving small meningeal vessels. Left superior ophthalmic vein is opacified. There are atherosclerotic changes at posterior circulation. Surgical intervention or embolization was not done in view of mild ocular symptoms and well control intraocular pressure. The dural CCF resolved spontaneously after few months follow up.

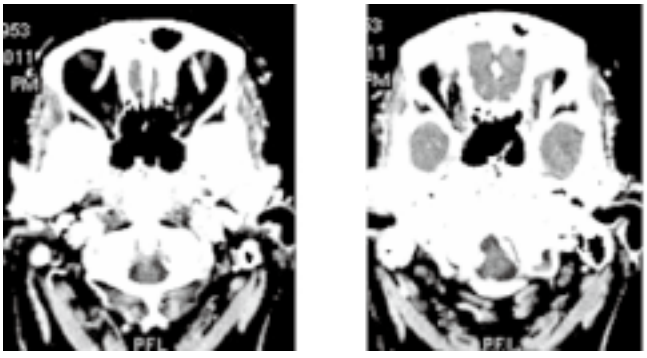

Figure 5. CT angiogram shows bilateral dilated/prominent superior ophthalmic veins

3. Discussion

Carotid-cavernous sinus fistula is an abnormal communication between the internal or external carotid arteries and the cavernous sinus. Because of this communications there will be some changes; includes the blood within the affected vein becomes arterialized, raises in venous pressure and venous drainage will be altered in rate and direction. In direct fistula, blood will flow directly into the cavernous sinus. This will cause increase blood flow and elevated pressure to be transmitted through the sinus to its tributary veins including ophthalmic vein[1].

Direct carotid-cavernous sinus fistula, represent 70%-90% of all carotid-cavernous sinus fistula. It is characterized by a direct connection between the intracavernous segment of the internal carotid artery and the cavernous sinus[2]. These fistulas usually have high rate of arterial blood flow and most commonly are caused by a single traumatic tear in the arterial wall.

Cerebral trauma accounts for approximately 75% of carotid-cavernous fistula with motor vehicle accidents, fights and falls representing the most common settings. The injuries may be penetrating or non penetrating and may be associated with basal or facial skull fracture. Other cause of direct carotid-cavernous fistula is spontaneous cause without any antecedent trauma or manipulation. This account approximately 25% of carotid-cavernous sinus fistula. It is due to rupture intracavenous carotid aneurysm or an atherosclerotic artery.

Post menopausal hypertensive women are at particular risk. These fistulas usually have lower flow rates and less severe symptoms than traumatic cases[2].

Indirect or dural carotid-cavernous fistulas are characterized by a communication between the cavernous sinus and one or more maningeal branches of internal carotid artery, external carotid or both. In this type of fistula the intracavernous portion of internal carotid artery remains intact. Arterial blood will flow through the meningeal branches of external or internal carotid arteries indirectly into cavernous sinus. Due to slow blood flow, the clinical features are more subtle than in a direct fistula.

The condition therefore be misdiagnosed or missed altogether. The lesions may represent congenital arteriovenous malformation, in which the onset of symptoms is precipitated by intracranial vascular thrombosis or develop spontaneously in association with atherosclerosis, systemic hypertension, collagen vascular disease, pregnancy and during or after childbirth[2]. Spontaneous rupture also can happen, which may be precipitated by minor trauma or straining especially in hypertensive patients[1].

As in our patient, she only presented to us after three months having symptoms most probably due to subtle clinical features which make the patient tend to ignore. This patient also is a known case of hypertensive and had history of minor trauma which may cause spontaneous rupture of one of the thin-walled dural arteries that transverse the sinus and lead to indirect CCF.

Ocular signs of carotid-cavernous fistulas are related to two main pathogenesis; venous congestion and reduced arterial blood flow to the orbit. Diminished arterial flow to cranial nerve within the cavernous sinus may cause diplopia. Stasis of venous and arterial circulation within the eye and orbit may cause

ocular ischaemia, and increased episcleral venous pressure may cause glaucoma. Our patient had all this pathogenesis, venous congestion changes, diplopia with 3rd and 6th nerve involvement and raised episcleral venous pressure caused blood in Schlemm canals with narrow angle and increase intraocular pressure.

The presence of ptosis may be helpful in differentiation from acute thyroid eye disease. A bruit is not pathognomonic of carotid cavernous fistula as it can be heard in normal infants, in young children, and in patients with severe anaemia[4].

Typically, arterialization of conjunctival veins is associated with other ophthalmologic manifestations, particularly with proptosis and can be found in 82% to 100% of patient with intraorbital symptoms. This may lead to dilatation and tortuosity of conjunctival veins which often misdiagnosed as other inflammatory conjunctivitis. However, it can be differentiated from allergic, viral or bacterial conjunctivitis by bright-red, corkscrew veins and usually no inflammatory reaction except in infectious cause[3]. Due to variform clinical manifestations, considering differential diagnosis is essential for optimizing therapy of patient with chemosis. Differential diagnoses includes mainly thyroid related orbitopathy (most common cause of unilateral and bilateral proptosis in adult), neoplastic disease (lymphoma, primary or secondary CNS tumors), allergic reaction, inflammation (viral/bacterial conjunctivitis, myositis), vascular and pseudotumor of the orbit and intraorbital bleeding (post-trauma, para-neoplastic)[3].

The abducent nerve is affected most often because it lies in the cavernous sinus, itself. The third and fourth nerves situated in the lateral wall of the sinus, are less commonly involved. Ophthalmoplegia occur in 60%-70% of cases due to ocular motor nerve damage caused by initial trauma or an intracavernous aneurysm or by the fistula itself[1]. Engorgement and swelling of extraocular muscle may also contribute to defective ocular motility. Other sign includes anterior segment ischaemia which is characterized by corneal epithelial oedema, aqueous cells and flare, iris atrophy, cataract and rubeosis iridis which are not present in this case.

Kim *et al* reported that Dural arteriovenous shunts are a dynamic disease that may undergo spontaneous angiographic pattern conversion. According to a long-term follow-up of a large single-center database, approximately 16% of the patients showed angiographic pattern conversion without any treatment. Various theories have been proposed to explain the angiographic conversions including natural history of the disease, fibrosis of intraluminal thrombus and the inflamed sinus wall may cause occlusion of the fistulous communication. Concomitant hemorrhage may promote thrombosis of the shunt by mass effect or vasospasm[5].

Thus, proper diagnosis, reassurance, and conservative follow up usually suffice. Only in the rare case is embolization necessary, as for a patient who had unacceptable symptoms and signs of visual loss, central vein occlusion, diplopia, severe exophthalmos or intolerable bruits[4].

Although significant risks of neurological or visual sequelae from treatment must be considered, treatment of the dural fistula should proceed surgery in cases of high intraocular pressure. In this patient spontaneous improvement occur after few months follow up without surgical intervention.

Conflict of interest statement

We declare that we have no conflict of interest.

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