A case of central retinal artery occlusion secondary to untreated barlow type D carotid cavernous fistula

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Background: To report a case of central retina artery occlusion after failed coiling in a Barrow Type D carotid cavernous fistula (CCF).
Methods: Case Report
Results: A sixty-four-year-old male with no prior history of trauma, presented with insidious onset of right eye proptosis with corkscrew vessels and bruit with increased intraocular pressure (IOP) but no blurring of vision and normal retina. Radiological examination showed Type D CCF. Cerebral coiling was performed but failed and resulted in worsening vision and intraocular pressure and noted to have central retina artery occlusion (CRAO). Secondary embolization was performed which obliterated the fistula, in which the patient recovered favorable visual function.
Conclusion: CRAO is a rare complication of cerebral coiling, in which the patient regained favorable visual function. This report describes our experience of CRAO post cerebral coiling which recovered after obliteration of the fistula by embolization.
Conflicts of interest: The authors report no conflict of interest.
Keywords: Carotid cavernous fistula, Barrow D, central retina artery occlusion, chemosis, proptosis, secondary glaucoma.
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Case history

Patient is a 64 years old Malay gentleman, with underlying hypertension and diabetes mellitus (T2DM) on medications. He first presented to the eye clinic in November 2015, with chief complain of redness of his right eye, without pain nor blurring of vision. He denies any systemic symptoms such as weight loss, joint pain, skin lesion or cough. Vision at this time was 6/6 bilaterally. Examination showed diffuse injection with red nodule at the nasal aspect of the sclera which did not blanch with vasoconstrictor. No signs of necrotizing, thinning of sclera or intraocular inflammation noted. General examination showed unremarkable findings. There was no sclera thickening on B scan and T-sign was negative. Initial diagnosis was nodular anterior scleritis with a

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differential of nodular episcleritis. He was started on topical steroid (Dexamethasone 0.1%) and was blood investigation was sent to rule out connective tissue disease. His red eye improved over the course of two weeks but the antinuclear antibody test (ANA) was positive (1:40, speckle pattern). Other tests were unremarkable. He was then referred to rheumatologist for further management.

One month later, he presented again with progressive right eye redness and swelling associated with visual acuity of 6/18 in his right eye. No history of trauma and he denied hearing any whooshing sound. On examination of the right eye, proptosis with chemosis and cock screw vessels were noted. Ocular motility was restricted in all directions of gaze. There was a difference of 9 mm on exophthalmometer measurement. The right pupil was mid dilated with evidence of relative afferent papillary defect. Intraocular pressure was 35 mmHg. The eyeball was not retropulsible and there was no bruit or thrill noted. Fundus examination showed pink optic disc both eyes with cup to disc ratio of 0.7 bilaterally. There were flamed shape hemorrhages near the disc and macula area. B scan showed flat retina with no fluid collection in the choroidal spaces. Provisional diagnosis at that time was right carotid cavernous fistula. He was started on oral acetazolamide and topical timolol/azopt/xalatan on the right eye to reduce the intraocular pressure (IOP).

An urgent CT brain/orbit revealed evidence of right eye proptosis and dilated right superior ophthalmic vein measuring 4mm in diameter. CT angiogram showed right cavernous sinus enlargement and dilated right superior ophthalmic vein (Figure 1). Cerebral angiogram showed type D carotid cavernous fistula with contralateral draining into left superior ophthalmic vein (Figure 2). Cerebral coiling was attempted but was failed due to inability to cannulate the fistula. At this point, his right eye became more proptosed (Figure 3) and his vision reduced

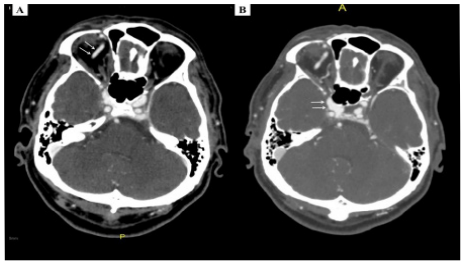

Figure 1: Computed tomography angiogram of the brain in axial view showing (A) dilated right superior ophthalmic vein and (B) enlargement of the right cavernous sinus (Source of Photo: Courtesy of Radiology Dept. Hospital of University Malaya).

to hand movement. His IOP was also not controlled (increased to 41 mmHg) despite being on 3 topical anti glaucomas. Right eye paracentesis was done to reduce the IOP and lateral canthothomy performed to help close the eyelids and protect the cornea. Fundus examination showed hazy media.

Second attempt of embolization of the fistula using liquid embolic system (PHIL 25-precipitating hydrophobic injectable liquid) was done through the external carotid artery (Figure 4). CT brain post

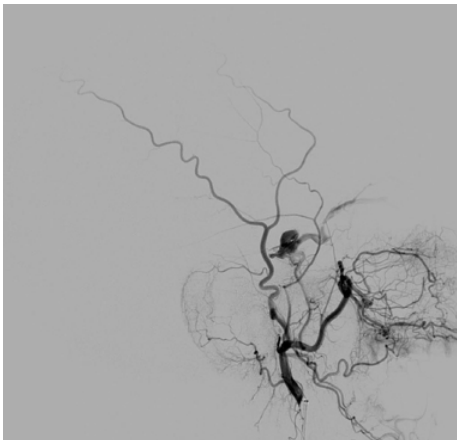

Figure 2: Location of the carotid cavernous fistula (arrow) Source of Photo: Courtesy of Radiology Department, Hospital of University Malaya).

procedure confirmed the material in the cavernous sinus. (Figure 5).

His condition improved later with

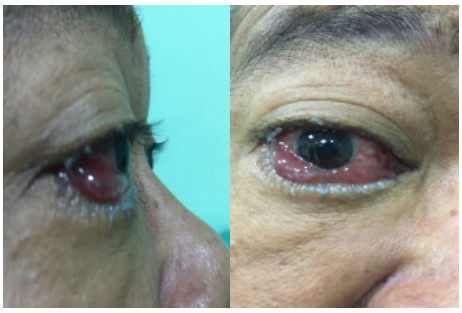

Figure 3: Lateral view and anterior view right eye

reduction of proptosis and chemosis 1-month post event (Figure 6). His vision however remained at hand movement in right eye. RAPD still present (Grade I) and IOP was 22mmHg on 3 types of antiglaucoma. Fundus showed cherry red spot with pale retina (Figure 7).

At 3 months post op, his condition was stable, and he was comfortable without any pain or redness to the eyes. His right eye visual acuity was 6/36, and the IOP was 18 mmHg, The Alphagan eye drop

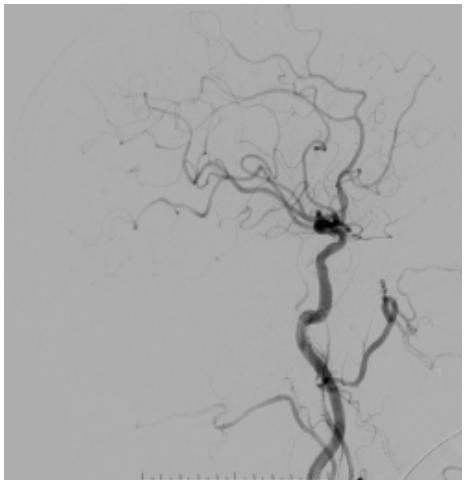

Figure 4: Obliteration fistula. Source of Photo: Courtesy of Radiology Department, Hospital of University Malaya.

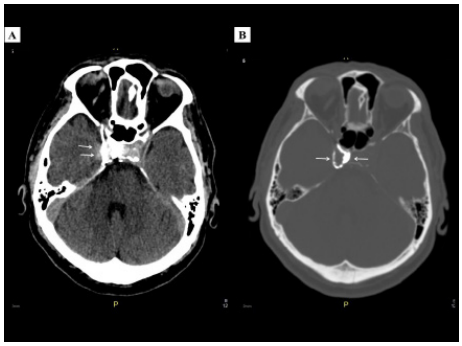

Figure 5: Non-contrast-enhanced computed tomography of the brain in axial images in (A) brain window showing artifact from the right cavernous sinus and in (B) bone window dense embolic material in the right cavernous sinus (Source of Photo: Courtesy of Radiology Department, Hospital of University Malaya).

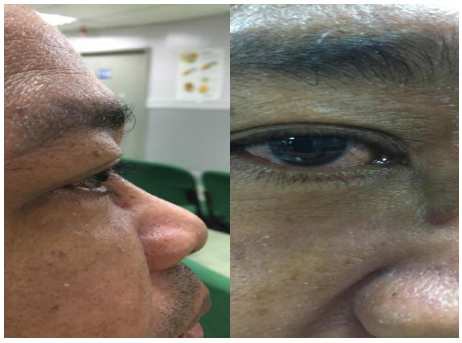

Figure 6: Lateral and anterior view of the right eye of the patient, 1 month after treatment, showing resolution of the chemosis and proptosis, but vision remains hand movement (HM)

was discontinued and he was seen again 4 months later (7 months post procedure). During this time, he denies any new complaints, the right eye visual acuity remained the same at 6/36 and his IOP came down to 14 and the timolol eye drop was discontinued. At 12 months follow up after the procedure, his best corrected visual acuity (BCVA) of the right eye improved to 6/24 and IOP is well controlled (15 mmHg) with single topical antiglaucoma

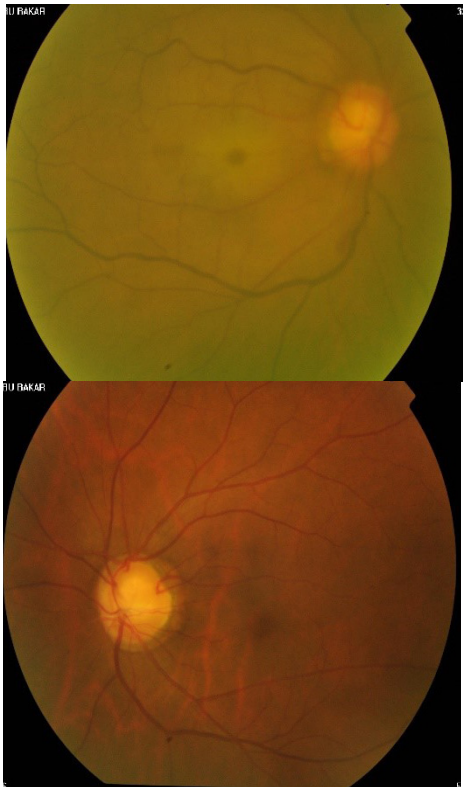

Figure 7: OCT macula of the right eye, showing cup disc ratio of 0.7, pale retina with cherry red spot as compared to the left eye.

Discussion

Carotid cavernous fistula (CCF) is an atypical communication between the carotid arterial system and the cavernous sinus. CCF can be classified on the basis of: (a) aetiology (spontaneous or traumatic), (b) hemodynamics (high and low flow) and (c) anatomy (direct and indirect). The most common classification is by Barrow et al,[4] who have divided CCF to 4 types according to its arterial supply:

Type A (direct): direct communication between the internal carotid artery (ICA) and the cavernous sinus.

Type B (indirect): supplied only by the dural branches of the ICA.

Type C (indirect): supplied only by dural branches of the external carotid artery (ECA).

Type D (indirect): supplied by dural branches of the ICA and ECA.

This classification is at present widely used and is of vital importance as the method of endovascular treatment may change. Angiography in this patient revealed that the fistula was supplied by dural branches of both ICA and ECA, therefore it was classified as Type D (indirect) CCF.

Unlike direct CCF, which mainly attributed by trauma, indirect CCF usually present with gradual onset of symptoms. Most of the symptoms are eye-related symptoms which include chemosis, exophthalmos, cranial nerve deficits, decrease in visual acuity, diplopia and ptosis.[2,3,5] This patient exhibited progressive sets of ocular of symptoms, which had threatened the vision of the right eye, and was complicated with secondary glaucoma.

In indirect CCFs, the goal of treatment is to reduce the pressure in the cavernous sinus by interrupting the fistulous communications. This can be achieved by obliterating the cavernous sinus that harbors the fistulous communication (transvenous embolization) which is more successful or obstructing the arterial branches that supplies the fistula (transarterial embolization).[1,6] Coils has been traditionally used in treatment of both direct and indirect CCFs. The benefits of coils are their radio-opacity and thrombogenicity which permits precise deposition and location into the cavernous sinus.[3] It is also easy to position and can be recovered if the initial location is not acceptable.[1] However, the coils need to be optimally packed to accomplish complete sealing which is difficult. Coils also tend to exert more mass effect, therefore dense packing of coils into the cavernous sinus may cause cranial nerve palsy. 'PHIL' (Precipitating Hydrophobic Injectable Liquid) (Microvention, Inc California, USA) is a recent advent of non-adhesive

liquid embolic agents. It has been recently used in treating indirect CCFs. Preliminary experience of its use in CCFs and spinal dural arterio-venous fistulas. appears to be an excellent embolic material with certain advantages compared with other available liquid embolic agents.[7]

In this patient, cerebral coiling was attempted but failed. Due to deterioration of the ocular signs (increasing proptosis, worsening vision, high IOP, presence of exudative retinal detachment and also CRAO), embolization using PHIL was then attempted to occlude the fistula, which was successful. However, the damage to the vision was severe and he had a vision of 3/60 in right eye 8 months' post event, which still need a single antiglaucoma eyedrop to maintain the IOP, although the proptosis and chemosis resolved.

Indirect CCF represent 10-30% of all CCFs, in which Type D is the commonest among the indirect CCF.[8] It is described as having a communication between one or more meningeal branches of external carotid, internal carotid artery or both and the cavernous sinus. In this form of fistula, the intra-cavernous portion of internal carotid artery remains intact. Arterial blood will flow via the meningeal branches of internal or external carotid arteries indirectly into cavernous sinus. Due to slow blood flow, the clinical features are subtler than in a direct fistula.[1]

The patients with CCF may present with chemosis, pulsatile exophthalmos and complaints of hearing a noise in the head. Clinically patients presented with corkscrew episcleral blood vessels in association with conjunctival chemosis, pulsating proptosis, thrill and bruit. These features should have a high index of suspicion of the diagnosis of arteriovenous fistula.[9] Resistance from the retrograde venous drainage into the ophthalmic vein may be causing proptosis, episcleral and conjunctival arterializations. Limited ocular movement and diplopia as a consequence of hypertrophied extraocular muscles; and exposure keratopathy as an effect of proptosis may present with a painful cranial nerve palsy with a white quiet eye in the absence of any proptosis.[10] The definite diagnosis can be confirmed by angiography, which may confirm a posterior draining CCF. Elevated episcleral pressure and vortex venous pressure may result in elevated intraocular pressure (IOP) and secondary glaucoma.[11] One of the often-observed ocular manifestation of CCF is secondary glaucoma, and closing the fistula is the crucial for satisfactory IOP control. There may be decreased ocular and retinal perfusion resulted by venous and arterial stasis. Retinal and choroidal changes may include, retinal hemorrhage, venous dilatation, CRAO, CRVO, cotton wool patches and serous retinal detachment. In addition, decreased perfusion to intra-cavernous sinus cranial nerves may cause anterior segment ischemia, causing ophthalmoplegia and diplopia mimicking Graves' ophthalmopathy. The visual loss may be secondary to corneal, retinal or optic nerve changes or may result from the accompanying glaucoma.[12] Rarely, a case of CCF after a cataract surgery has been reported.[13]

Embolization of CCF may carry a risk of inherent complication either from the procedure or due to reopening of the fistula. Complete ophthalmoplegia and visual acuity loss due to a central retinal vein obstruction (CRVO) after an attempt to close a CCF has been reported.[14] Ophthalmic artery occlusion and cerebral infarction post embolization are rare, which is one study reported 3 in 80 treated patients suffered such complications.[8] Other complications include increased proptosis, elevation of IOP, choroidal detachment, venous stasis retinopathy, ophthalmic vein thrombosis resulting in CRVO and neovascular glaucoma after embolization attempt.

Most CCFs are vision threatening, although they are usually not life threatening. Main indications for treatment include diplopia, glaucoma, intolerable headache or bruit, and severe proptosis producing exposure keratopathy. Spontaneous closure from thrombosis of cavernous sinus is unlikely. Non-surgical approach includes carotid compression therapy while surgical management includes ligation of the external and internal carotid arteries; and fistula embolization with particles, glue, detachable balloons and thrombogenic microcoils. Treatment of cavernous dural arteriovenous fistulas is usually done using a trans-arterial approach, in which in this patient done via the femoral artery and the external carotid artery, was successful and showed complete obliteration of the fistula.

The role of ophthalmogist is very important in this type of cases, as many patients with CCF may initially present to an ophthalmologist. High index of suspicion, appropriate diagnostic investigation is important to make a correct diagnosis. Proper follow up and care must be delivered to the patient as these patients might develop complications such as CRAO and secondary glaucoma.

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