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Bilateral Carotid Cavernous Fistula: To Wait or Not To Wait For Spontaneous Resolution

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Abstract: A 65 year-old Malaysian Chinese gentleman with history of hypertension presented with bilateral eye redness for three years associated with diplopia, mild blurring of vision, watery eyes, nausea and dizziness. On examination, the right eye best corrected visual acuity was 6/12 and left eye was 6/9. The right lateral gaze was slightly restricted and there was prominent corkscrew episcleral vessels on both eyes. CT Brain & Orbit showed mucosal thickening in left ethmoidal and left maxillary sinuses. CT angiogram showed bilateral symmetrical bulky and prominent cavernous sinuses with bilateral symmetrical enlarged and engorged superior ophthalmic veins. Bilateral carotid cavernous fistulas (CCFs) are extremely rare. Our case is the fourth (4^{th}) case of bilateral spontaneous CCFs that has been reported so far to achieve spontaneous resolution of ophthalmologic symptoms without neurosurgical or vascular intervention.

Keywords: Bilateral spontaneous carotid cavernous fistulas, spontaneous resolution, conservative management

INTRODUTION

While carotid cavernous fistulas (CCFs) are uncommon entities, bilateral CCFs are extremely rare, with only 67 cases reported in the literature [1]. Of the 67 cases of bilateral CCFs that had attainable CCF typing information, 26 (37%) were spontaneous and 41 (63%) were traumatic [1].

It is reported that seven (7) patients with bilateral spontaneous CCFs were managed conservatively [1]. Among the seven (7) cases, three (3) cases achieved spontaneous resolution with symptomatic improvement, one (1) case unresolved and refused treatment, while three (3) cases did not mentioned the outcome [1]. We report the fourth (4^{th}) case of bilateral spontaneous CCFs that achieved spontaneous resolution of ophthalmologic symptoms without neurosurgical or vascular intervention.

CASE REPORT

A 65 year-old chinese gentleman with history of hypertension presented with bilateral eye redness for three years, started with right eye followed by the left eye. The symptoms worsened over one week duration prior to the presentation. It was associated with diplopia, mild blurring of vision, watery eyes, nausea and dizziness. He denied history of fever or trauma to the eye or head. There were no symptoms suggestive of connective tissue disease or thyroid disease. There was no history of tuberculosis or connective tissue disease in the family. On examination, the right eye (RE) best corrected visual acuity (BCVA) was 6/12 and left eye (LE) was 6/9. There was no proptosisor anisocoria. The relative afferent papillary defect was negative. Right lateral gaze was slightly restricted. Prominent corkscrew episcleral vessels were noted on both eyes (BE) (figure 1). BE intraocular pressure was 18 mmHg. BE optic disc were pink with cup disc ratio of 0.3. Other ocular and fundus findings were unremarkable. Systemic examinations revealed stable vital signs. There was no neck swelling, no proximal myopathy and no fine tremor. The respiratory and the cardiovascular system examination were normal. Other cranial nerves examinations were normal. A series of blood investigations were done, including full blood count, erythrocyte sedimentation rate, renal function test, liver function test, and anti-nuclear antibody. Rheumatic factor was noted to be elevated with a level of 32. All other blood investigations results were unremarkable. Chest x-ray was normal and mantoux test was 2mm. CT Brain & Orbit showed mucosal thickening in left ethmoidal and left maxillary sinuses. No obvious enhancing large retro-orbital mass. No pseudotumour seen. The recti muscles of both orbits were not enlarged. Eyeballs were grossly intact (figure 2). CT angiogram showed bilateral symmetrical bulky and prominent cavernous sinuses, with patent contrast opacification. Right cavernous sinus measures 1.2cm

width and Left 1.1cm width. Bilateral symmetrical enlarged and engorged superior ophthalmic veins. No obvious large external carotid artery branches communicating into orbit. Both intracranial distal internal carotid arteries were grossly normal (figure 3). The findings were consistent with bilateral CCFs. He was subsequently referred to neurosurgical team and was planned for and magnetic resonance imaging and digital subtraction angiography. However, he refused further intervention and opted for conservative management. On Follow up, diplopia resolved after 2 months from the initial presentation. His BCVA improved to a visual acuity of 6/6 BE with full extraocular movements. However, the prominent corkscrew episcleral vessels persisted.

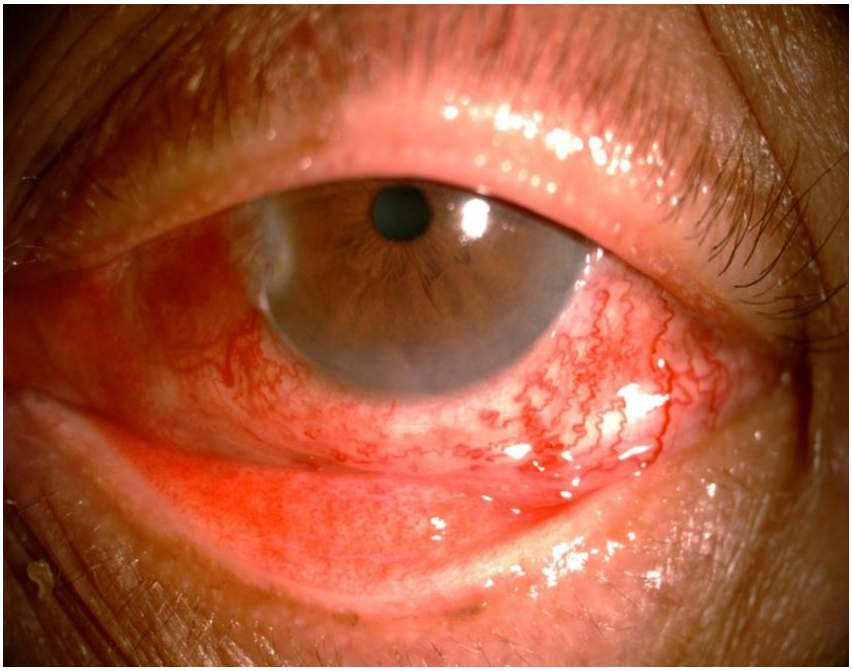

Fig-1: Prominent corkscrew episcleral vessels.

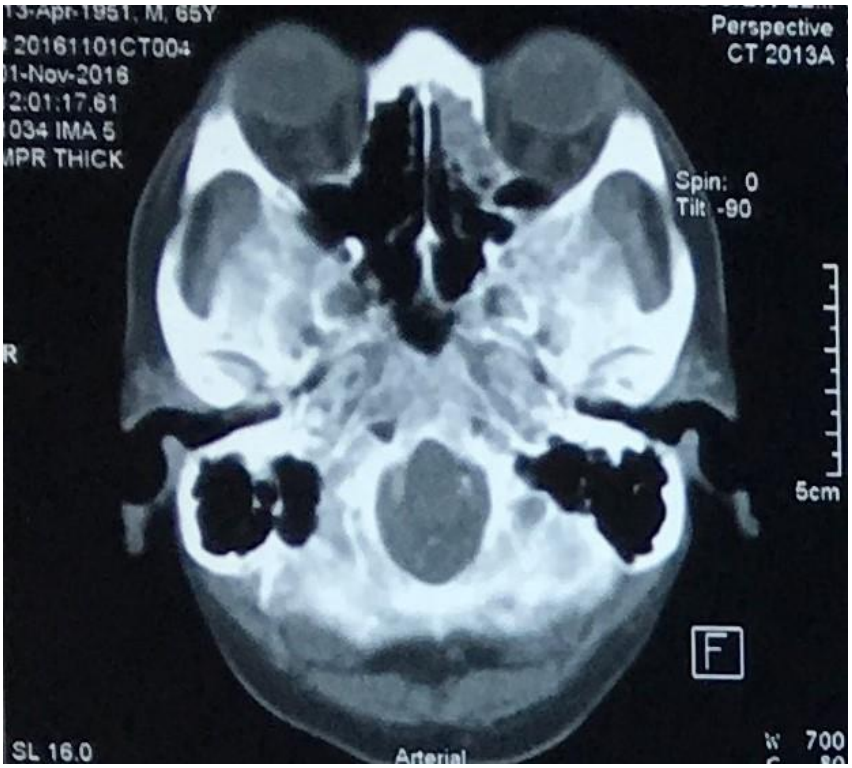

Fig-2: CT Brain & Orbit showed mucosal thickening in left ethmoidal and left maxillary sinuses. No obvious enhancing large retro-orbital mass. No pseudotumour seen. The recti muscles of both orbits were not enlarged. Eyeballs were grossly intact.

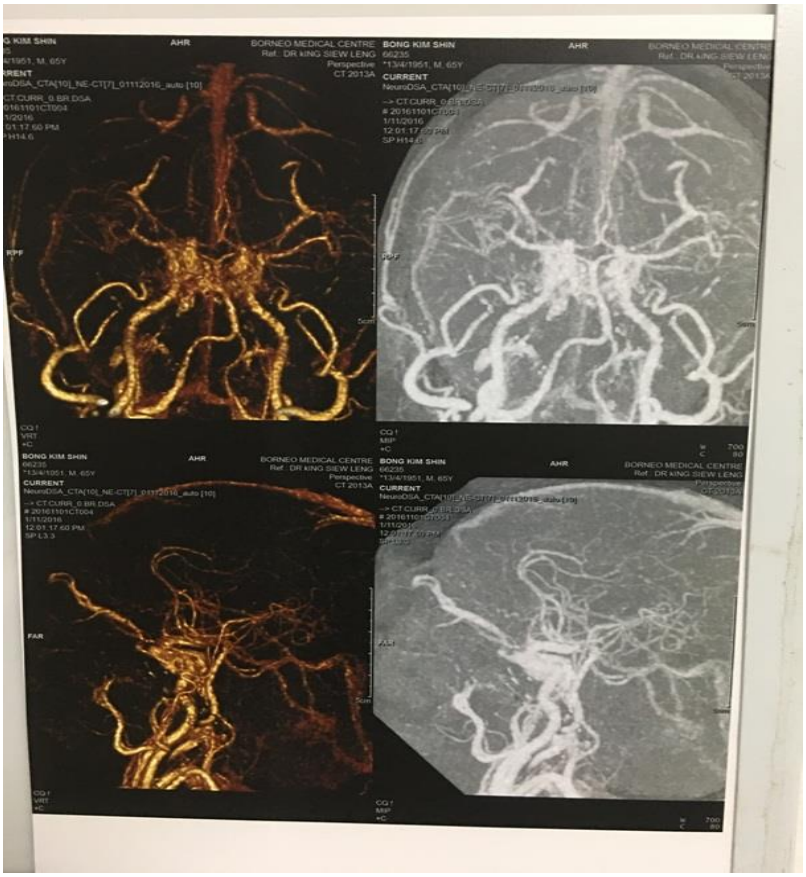

Fig-3: **CT angiogram showed bilateral symmetrical bulky and prominent cavernous sinuses, with patent contrast opacification. Right cavernous sinus measures 1.2cm width and Left 1.1cm width. Bilateral symmetrical enlarged and engorged superior ophthalmic veins. No obvious large external carotid artery branches communicating into orbit. Both intracranial distal internal carotid arteries are grossly normal**

DISCUSSION

CCFs may be categorized based on etiology, rate of flow, or by angiographic architecture [1]. The most commonly used classification scheme established by Barrow *et al*. [2] divides the CCFs into 4 types, depending on the arterial supply. Direct (typeA) are direct communications between the internal carotid artery (ICA) and the cavernous sinus, usually associated with high flow rates. Indirect fistulas (type-B, -C, and -D) are dural arteriovenous fistulas fed by the meningeal arteries of the ICA (type-B), or the external carotid artery (ECA) (type-C). Type-D indirect fistulas are supplied by the meningeal branches of both the ICA and ECA [2].

Typically, CCFs are unilateral, and true spontaneous indirect bilateral CCFs are extremely rare, with only 67 cases reported in the literature [1]. Of the 67 cases of bilateral CCFs that had attainable CCF typing information, 26 (37%) were spontaneous and 41 (63%) were traumatic [1].

Spontaneous CCFs may fall into any of these four categories [2]. The majority of spontaneous CCFs is idiopathic and tends to appear in middle-aged women. These CCFs generally present with insidiously progressive glaucoma, proptosis, or a "red eye" -- signs and symptoms that are usually less severe than those seen in direct fistulas [3]. These idiopathic CCFs are usually low-flow and can be angiographically divided into Types B, C, or D [2].

The cause of spontaneous CCFs is still unclear. Congenital dural fistulas have been reported in infants as young as 5 weeks of age [4].Taniguchi *et al*. [5] speculated that indirect CCFs may represent a collateral response to thrombosis of the cavernous sinus and may be a manifestation of a thrombotic tendency, which may lead to multiple dural fistulas. Newton and Hoyt[3] postulated that spontaneous low-flow CCFs form after the rupture of one of the thin-walled dural arteries that normally traverse the cavernous sinus. Different factors that may predispose patients to this rupture include hypertension, pregnancy, trauma and straining, atherosclerotic disease, and collagen vascular disease [6].

Spontaneous resolution of dural fistulas can occur independent of treatment. Its incidence ranges from 10% to 60% in the literature [2]. This resolution may be attributed to further thrombosis of the involved segment of the cavernous sinus [6]. Our case is the fourth (4^{th}) case of bilateral spontaneous CCFs that has been reported so far to achieve spontaneous resolution

of ophthalmologic symptoms without neurosurgical or vascular intervention. Our patient's only lingering sequela was dilated episcleral vessels while he retained a visual acuity of 6/6 with full extraocular movements in both eyes

If the decision not to treat CCFs is made, the patient must be carefully followed for progressive visual deterioration [4]. Barrow et al proposed the following indications for treatment of spontaneous CCFs: 1) Visual deterioration 2) Obtrusive diplopia 3) Intolerable bruit or headache. 4) "Malignant" proptosis with untreatable corneal exposure. The fact that spontaneous low-flow dural CCFs are rarely life-threatening emphasizes the importance of minimizing the morbidity and mortality of any therapeutic procedure [3].

CONCLUSION

CCFs usually present unilaterally. However, as demonstrated in our case, CCFs can uncommonly present bilaterally. Intervention is not without risk, and in cases which the potential complications associated with intervention are great, with the knowledge that spontaneous resolution is possible, the clinicians may choose to monitor patients that are stable and experiencing few and non-life-threatening symptoms.

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