CASE REPORT

A Rare Case Report of Bilateral Traumatic Carotid Cavernous Fistula

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ABSTRACT

Objective : To increase the awareness for ophthalmologist in diagnosing rare case of bilateral traumatic carotid cavernous fistula that could lead to serious complications.
Case presentation : A 44 years old woman came with chief complaint of squint and diplopia since one month after traffic accident. She had been underwent orbital bone fracture repair at previous hospital immediately. There was complaint of headache, blurred vision, double vision and pain on the eyes one month after trauma. There was no history of redness or proptosis. Examination revealed 6/6 for the right and 6/8.5 for the left eye corrected visual acuity, 30 degrees exotropia, normal intraocular pressures (IOP), restriction on eyes movement especially on the left eye, normal anterior and posterior segment, and no bruit. CT orbital revealed bilateral dilated and tortuous of the ophthalmica vein with suspicious of carotid cavernous fistula (CCF). Patient developed slowly progressive conditions such as proptosis non axial on the left eye, redness with corkscrew appearance on both eyes, restriction of the ocular movement and also founded bruit on both eyes within two months follow up. CT angiography revealed CCF bilateral type A. Patient underwent digital subtraction angiography (DSA) and balloon embolization four months after diagnosed.
Conclusion : Although bilateral carotid cavernous fistula is a rare case, performing a suitable diagnostic examination would help to prevent devastating outcome. Prognosis depends on severity and time span from symptoms to treatment.

Keywords : *Carotid-cavernous fistula, bilateral carotid-cavernous fistula, digital subtraction angiography, trauma*

Carotid-cavernous fistulas (CCFs) is a rare condition with a pathologic communications between carotid artery (internal or external) and the venous plexus of the cavernous.[1] There are two types of CCF based on etiology, which are traumatic (70-90%, mostly in man)[2,3] or spontaneous.[4,5] Commonly patients with traumatic CCF are unilateral,[6] while the bilateral types are uncommon (1-2%). [7-9]

The manifestation of direct CCF might not directly occur after the accident it could occurred several days to weeks related to rupture of a false traumatic aneurysm into the cavernous sinus.[8,9]

Symptoms and sign of bilateral CCFs are eyelid swelling, orbital bruit, pain, pulsating exophthalmos, chemosis, pupillary dilatation, increased IOP, diplopia, ophthalmoplegia, loss of visual acuity.[1,6] Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) are the modalities to depict the pathologies in the peripheral associated with CCFs.[10,11] The aim of treating patient with CCF is to occlude the fistula site, either manual or radiosurgical intervention, and endovascular embolization for high blood flow management.[2,10,11] Untreated fistula can cause a serious complication, such as loss

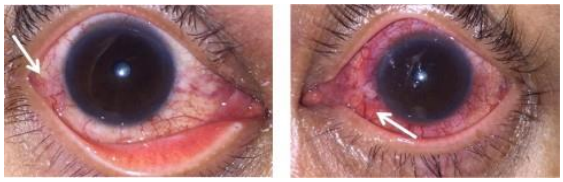
of vision and ischemic optic neuropathy.[11]

We reported a rare case of bilateral traumatic carotid cavernous fistulas in clinical practice and published articles. The purpose of this paper is to help ophthalmologist making an early diagnosis and prompt treatment to prevent it from serious complications.

CASE PRESENTATION

A 44 years old woman came with chief complaint of squint and diplopia since one month after accident. There was history of motorcycle accident on April 2017. She underwent operation for the orbital bone fracture at previous hospital one month after the accident. There was complaint of headache, blurred vision, double vision and pain on the eyes one month after trauma. Patient referred from previous hospital with diagnosis exotropia fixtus et causa suspicious parese of the nervous III with the differential diagnose of muscle loss/ entrapment of the rectus muscle with history of the orbital rime fracture of both eyes. The corrected visual acuity at admission was 6/6 for the right eye, and the left eye was 6/8.5. Hirschberg examination showed 30 degrees exotropia. Furthermore, the intraocular pressures of both eyes were within normal limit at the first visit. There was restriction on eyes movement especially on the left eye. There was no history of redness nor proptosis. The anterior and posterior segment was within normal condition. The bruit was not heard on the first admission. Patient then consulted to the plastic reconstructive division and diagnosed with ophthalmoplagia of the left eye et causa suspicious parese of the nervous III, IV and VI. Afterward, patient consulted to the neuro- ophthalmology division and was performed the computed tomography (CT) orbital.

CT orbital revealed bilateral dilated and tortuous of the ophthalmica vein with suspicious of carotid cavernous fistula (CCF). During the follow up at Cipto Mangunkusumo hospital, patient develop slowly progressive conditions such as proptosis non axial on the left eye, redness on both eyes with corkscrew appearance, restriction of the ocular movement and also bruit was heard on both eyes. From the CT angiography results patient diagnosed with CCF bilateral type A. Patient underwent digital subtraction angiography (DSA) and balloon embolization on December 2017.



(A)

(B)

(C)

Fig 1. (A) Corckscrew episcleral vessels of both eyes (B) Ophthalmological examination showed 30 degrees exotropia and limitation of eye movement and showed episcleral venous enlargement of both eyes (C) Funduscopy: both eyes showed round optic nerve with cup disc ratio 0.3-0.4

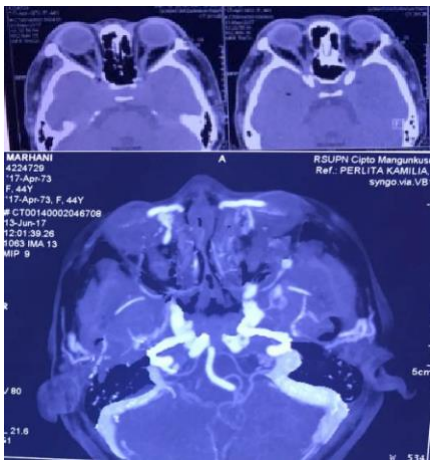


Fig 2. (Above) Computed Tomography orbital showed dilated superior ophthalmic vein in both eyes. (Below) Computed Tomography angiography (CTA) of the patient confirmed the connection between the internal carotid artery and the cavernous sinus in both sides

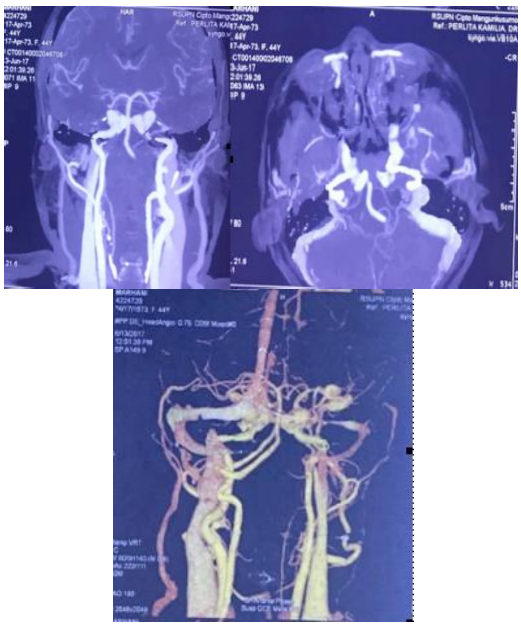


Fig 3. Enlarged of the ophthalmic vein and cavernous sinus visualized from CT angiography

Fig 4. Restriction of the eye movement to temporal sides of both eyes

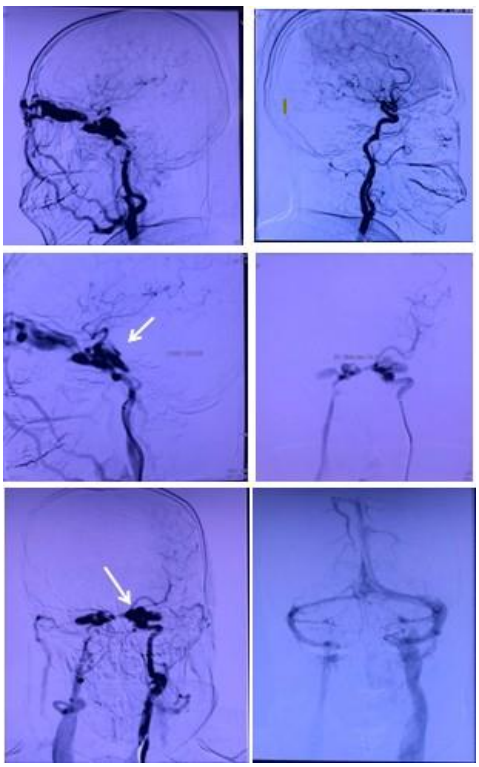


Fig 5. DSA and Ballooning embolization procedure. Catheter arteriography showed dilatation and tortuosity of the superior ophthalmic vein to the fascialis vein and dilatation of the inferior petrosus sinus that goes to the jugular vein

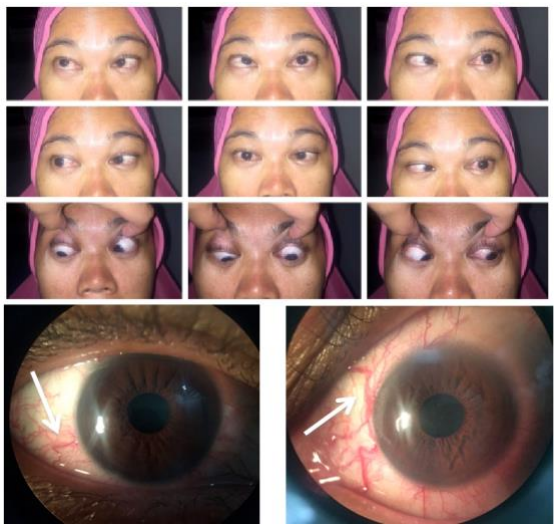


Fig 6. Post DSA and ballooning embolization. Both eyes still showed the cork and screw appearance (20th December 2017)

DISCUSSION

In this case report, patient came with chief complaint of squint in the left eye one month prior to the head trauma. The manifestation of CCF rarely occurred

immediate post-traumatic period, they usually manifest in days or weeks after the accident (the longest was 6 weeks).[8] Patient with bilateral type have the same symptoms to the unilateral type with the milder conditions. Moreover, the clinical appearance does not necessary occurred simultaneous bilateral manifestations due to difference in size and haemodynamics of the fistula.[6]

At the first admission, in our case there were no signs of proptosis, chemotic, corkscrew appearance and no bruit was found. Patient came due to squint eye, pain and restriction of the ocular movement and tinnitus one month after the trauma. Two months after accident, there was spasm, proptosis, hyperemia, ophthalmoplegia, and increased IOP. The delay manifestation could be related to rupture of a false traumatic aneurysm into the cavernous sinus, especially in non- communicable patient with impaired neurological state.[8] In this condition, the traumatic carotid dissection and pseudoaneurysm formation could develop late, within 2 -6 weeks after the initial trauma.[12]

She complaints of headache since one month post the trauma. Headache is usually found in 25 – 84% patient with CCF. There were also visual disturbance such as diplopia and also orbital pain. Diplopia reported happen in 88% of traumatic CCF case. Two months after the trauma she had proptosis non axial on the left eye which account as much 72 to 98% patient with CCF and also orbital bruits. These conditions could occur due to congestion of the draining pathway of the ophthalmic vein due to high flow from the ICA.[7,13] In addition, no redness or proptosis could be due to multi compartment of the cavernous sinus without connection to ophthalmic vein and blood flow were drained to the inferior and superior petrosal sinuses, sylvian venous plexus to the cortical veins.[13-15] Moreover, from our case there were also episcleral and conjunctival arterialization that could be the result of the resistance into the ophthalmic vein by the retrograde venous drainage.[7]

From the examination, patient right eye corrected visual acuity was 6/6, and the left eye was 6/8.5. CCF sometimes could make the visual acuity decreased and even vision loss if there is condition such as optic neuropathy, venous congestion with macular edema, retinal hemorrhage, corneal edema with combination of secondary glaucoma.[16] As patient complaint of visual disturbance such as diplopia and also orbital pain it is important to performed urgent intervention as this condition could appear due to retinal ischemia.[7]

There were also secondary glaucoma that might be due to the increased in the episcleral pressure and the vortex venous pressure, therefore for IOP control is important to performed closure of the fistulas. Furthermore, from our patient the orbital bruit identified few months after the accident. It is known that as much as 88% of patient with post trauma CCF experienced orbital bruit. Orbital bruit could be subjective, objective or both.[17]

Patient also complaint of restricted ocular motility and diplopia because of the enlargement of the extraocular muscles, and also could occurred due to suppression in the bulbus oculi or inferior venous drainage and if there were cranial nerve dysfunction due to the mechanical compression or ischemia which commonly involve the CN VI.

From the CT orbital examination there were bilateral dilated and tortuous of the ophthalmica vein with suspicious of carotid cavernous fistula (CCF). Patient diagnosed with bilateral traumatic CCF type A as there were signs of a high-flow that usually found in type A CCF such as acute onset headache, diplopia and also hyperemic.[18] The gold standard for diagnoses of direct CCF is catheter-based angiography.[13] Patient then undergone the non-invasive imaging such as CT angiography and the result

confirmed the diagnosis.

Ideally the principal treatment of direct CCF should involve occlusion of the pathological arteriovenous connection with preservation of the ICA that is known as transarterial or transvenous coil or liquid embolization. The purpose is to lower the venous hypertension and improving the cerebral perfusion and maintain the normal flow in the carotid artery. In managing patient with direct traumatic CCF it is important to manage as early as possible to decrease the risk of total ICA. From the literature stated that the size of the fistula was related with the injury to admission time. The chance of preservation of the ICA during embolization would diminished when the waiting time before admission was longer.[13,18]

DSA treatment could give high cure rate and low complication especially in patients with direct traumatic CCF. Moreover it could give flexible accesses to the fistulous site with alternatives embolic materials.[18] Post embolization there was decreasing size of the fistula. The clinical manifestation shows an improvement in both ocular movements. The balloon catheterization could also give the small trauma, good effect, high cure rate and also low disability rate.[1]

In our case, patient was scheduled for DSA and embolization ballooning therapy. The initial therapy for patient with bilateral traumatic CCF is transarterial detachtable balloon embolization.[19] From the DSA and ballooning embolization in this patient there was partial embolization with the fistula size was bigger than the maximal balloon expansion. Post embolization there was decreasing size of the fistula. By performing a suitable therapy the symptom resolution with low rates of recurrence could be expected.[6]

The consequences of patient with untreated traumatic CCF are decreased visual acuity, ocular necrosis, intracranial hemorrhage, epistaxis and cranial nerve palsy. Consequently, aggressive endovascular therapy is generally should performed.[17]

From this case we can learn that in patients with CCF as the manifestation sometime could be delayed, it is important to perform a suitable examination in order to establish suitable diagnosis. A proper management could prevent this condition from devastating complication

CONCLUSION

Carotid cavernous fistula is a rare condition. Our patient is presented with bilateral CCF that was triggered by trauma. The clinical manifestation showed diplopia and restricted ocular motility that manifest after two month prior to the accident. The end result of this case is satisfying as the definitive diagnosis was established in short time. The size of the fistula is decreasing post embolization with significant improvement in the clinical appearance. The outcome for this condition influenced by the time the patient underwent treatment. Therefore, the earlier the diagnosis is being established, the better the outcome.

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