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Case Report : Direct Carotid Cavernous Fistula : A Case Report
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Wednesday, March 24rd , 2021
07.30 a.m

Direct Carotid Cavernous Fistula : A Case Report

Abstract

Introduction : Carotid Cavernous Fistula (CCF) arises from abnormal connections between the carotid artery and the cavernous sinus. Direct CCF frequently are traumatic in origin and also may be caused by rupture of an ICA aneurysm within the cavernous sinus. The varied clinical presentation of CCFs depends on the anatomy, hemodynamic and size of the CCF fistula. Spontaneously resolved is rare in direct CCF.
Purpose : To report a case of spontaneously direct CCF: the clinical manifestation and treatment planning
Case Report : A 38-year-old female complained double vision, redness and swelling of her right eye (RE), headache, and heard of whooshing sound since 2 months before. There were episcleral injection, ophthalmoplegia, bruit, and upper right face hypaesthesia. The visual acuity was 0.32, contrast and colour sensitivity were within normal limit. The intraocular pressure was 39 mmHg on RE. She had hypertension and diabetes mellitus that treated with medication, and no history of trauma. The Computed Tomography Angiography revealed Type A CCF according to Barrow Classification. The patient planned to undergo Digital Subtraction Angiography by Neuro-surgery Department. She received medication and instructed to perform an external manual carotid compression while waiting for DSA.
Conclusion : Direct CCF can occur spontaneously due to rupture ICA aneurysm within the cavernous sinus. The clinical manifestations are swelling and redness of the eye, diplopia, ophthalmoplegia, bruit, headache, decrease sensation on ipsilateral face, and raised IOP. The mainstay treatment for direct CCF is endovascular treatment, because the spontaneously resolved is rare.

Keyword : carotid cavernous fistula, direct, aneurysm rupture

I. Introduction

A carotid-cavernous sinus fistula (CCF) is an abnormal communication between arteries and veins within the cavernous sinus. CCF can be classified based on the hemodynamic properties, the etiology, or the anatomy of the shunt. Barrow et al. classified CCFs angiographically into four types-type A, B, C and D. In Type A (high flow) CCFs, there is a direct communication between the internal carotid artery (ICA)-cavernous segment and the cavernous sinus. In Type B, C and D (low flow) CCFs, there is an abnormal communication between the cavernous sinus, and one or more meningeal branches of the internal carotid artery, external carotid artery (ECA) or both respectively.[1-3]

The most common etiology of direct CCF is trauma, such as basilar skull fractures, projectile or slash injuries, or iatrogenic injuries, account for 70% to 75%

of all CCFs. Spontaneous CCFs represent 30% of all CCFs and result from aneurysm rupture or genetic conditions that predispose the patients to vascular injuries. They are most commonly seen in older females. In addition, spontaneous fistulas may result from rupture of structural weakness of the arterial wall in patients with connective tissue disease. Opposite to indirect CCFs (types B, C, and D), direct CCFs are less likely to resolve spontaneously and usually require treatment in symptomatic patients. In some large series, the rate of spontaneous closure of direct CCFs was reported to be about 1.2–4%. The exact mechanism of these rare events remains a controversial topic. [2-4]

The characteristic clinical features seen in patients with CCFs are the sequelae of hemodynamic dysfunction within the cavernous sinus. Though most CCFs are not life-threatening, prompt treatment is necessary to prevent permanent injury to the involved eye. Vision loss, intracerebral hemorrhage, subarachnoid hemorrhage, epistaxis, or cranial nerve palsies are complications of CCF. Although CCF management is formally in the area of interest of neuroradiology and neurosurgery, ophthalmologists are frequently called to diagnose and manage the condition in cases first presenting with ocular features. [2-4] Here by, we report a case of spontaneous direct carotid cavernous fistula, its ocular manifestation and treatment planning

II. Case Report

A 38 years old female came to Neuro-Opththalmology outpatient clinic in National Eye Center, Cicendo Eye Hospital with complaining of swelling and redness in her right eye since 2 months before. She also complained of tearing, pain, and experiencing of horizontal double vision. The diplopia would be better if she closes one eye. She admitted of headache, and hearing of whooshing sound. There is no blurred vision or history of trauma. She had history of hypertension since 8 years ago, and currently on antihypertensive medication. The highest blood pressure that she had ever got was 200/105 mmHg. There is also Diabetes Mellitus history since 2 years ago. She had been using injection contraception for 2 years

and oral contraception for 6 years. There is no any history of dyslipidemia or any other systemic disease. The patient was never using any eyeglasses.

The physical examination revealed the consciousness was compos mentis, with the blood pressure measure was 163/103 mmHg, heart rate was 101 beats/min and other vital sign was within normal limit. The ophthalmology examination showed 7^{o} esotropia, the ocular motility in the right eye (RE) was limited with -2 to nasal, -1 to inferonasal and inferotemporal, -3 to lateral, and -2 to superotemporal and superonasal whereas the ocular motility of the left eye (LE) was full. The ocular bruit was audible in the right eye. The uncorrected visual acuity of RE was 0.32 pinhole 0.8, and LE was 0.4 pinhole 1.0f2. Refractometry of RE was S-0.50 C-0.75 x 79 and the LE was S-0.50 C-0.25 x 83. Intraocular Pressure (IOP) was 39 on the RE and 19 on the LE. Hertel examination revealed 17 mm on RE and 16 mm on LE.

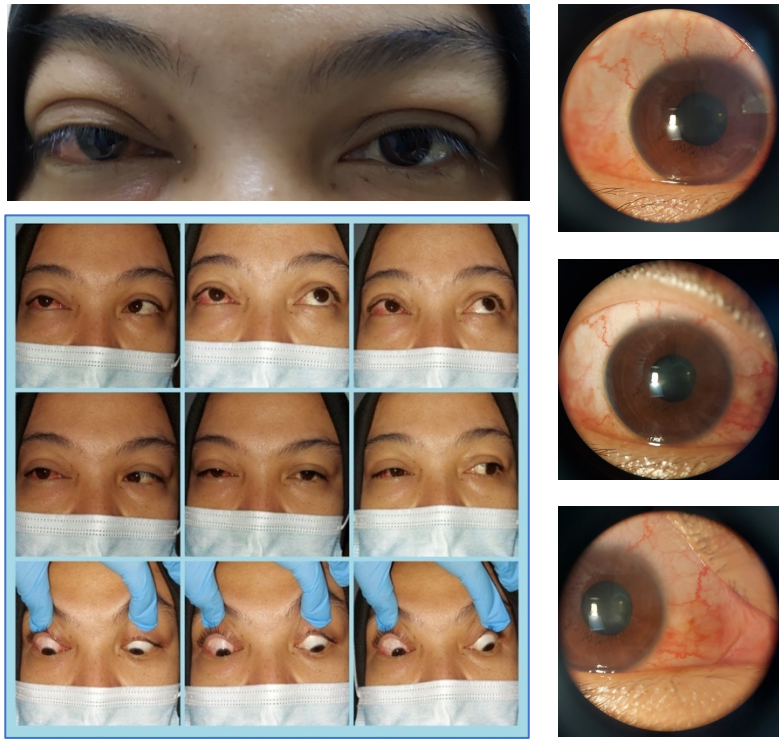
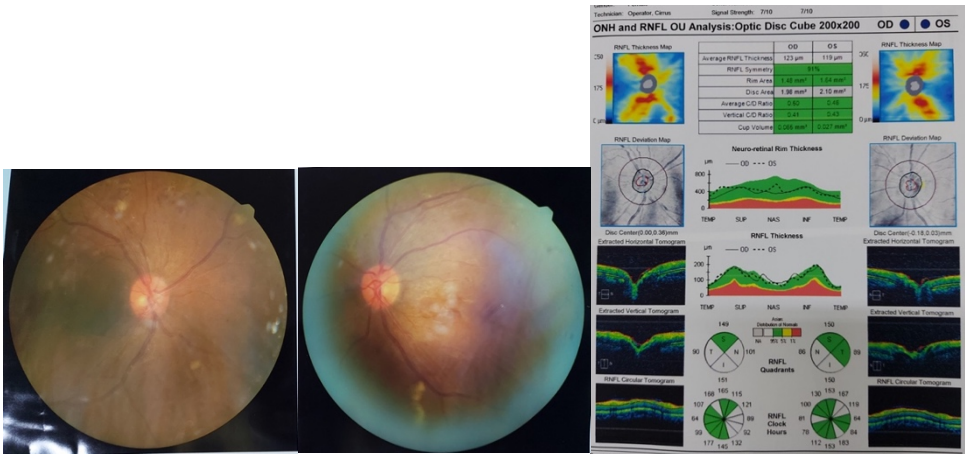


Figure 1. Ocular Motility and Anterior Segment of the Right Eye

Anterior segment examination on the RE showed the palpebra was edema and slight ptosis with Margin Reflex Distance (MRD) 1 : 1 mm, MRD 2 : 5 mm, and

Inter Palpebral Fissure (IPF) : 6 mm, and Levator Function (LF) : 17 mm. Palpebra of LE was quite with MRD 1 : 3 mm, MRD 2: 5 mm, IPF: 8 mm and LF : 17 mm. Conjunctiva bulbi on RE showed edema, the episcleral injection, corkscrew appearance on conjunctiva, clear cornea, normal light reflex on both eyes without relative afferent pupillary defect. Anterior segment on LE was within normal limit. Funduscopy examination on the both eyes showed mild swelling on inferior optic disc, cup to disc ratio 0.3-0.4, dot blot haemorrhage, cotton wool spot with artery to vein (A:V) ratio was about 1:3, without turtousity. Amsler grid, Ishihara colour plate and contrast sensitivity was within normal limit on both eyes. There was hypoesthesia in right upper facial, but other neurological examination was within normal limit.

Ocular Computed Tomography (OCT) of optic disc showed slightly increased Retinal Nerve Fiber Layer (RNFL) thickness on the right eye of inferior, nasal and temporal quadrant. Humphrey visual field 30-2 examination showed peripheral defect in RE but low test reliability due to high fixation loss.



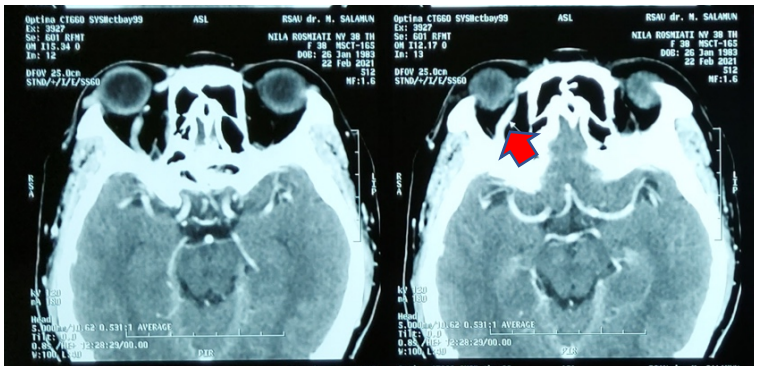
a. **b.** **c.**

Figure 2. Fundus Photography of a. Right eye; b. Left eye; c. Optic Disc OCT

The laboratory findings was fasting blood glucose 150 mg/dl, 2-hour-post-prandial blood glucose 178 mg/dl, HbA1c 8.7%, and normal lipid profile. The Contrast enhanced Computed Tomography (CT) Scan showed dilatation of the superior ophthalmic vein causing a mild proptosis of RE with swelling of the right

cavernous sinus, which suggests a "carotid cavernous fistula". CT Angiography revealed a "bulging" view of the right cavernous sinus directly connected to the right internal carotid segment C5 and dilatation of the right ophthalmic vein causing a mild proptosis of OD, suggesting a type A direct carotid cavernous fistula according to the Barrow classification of caroticocavernous fistulae.

a.



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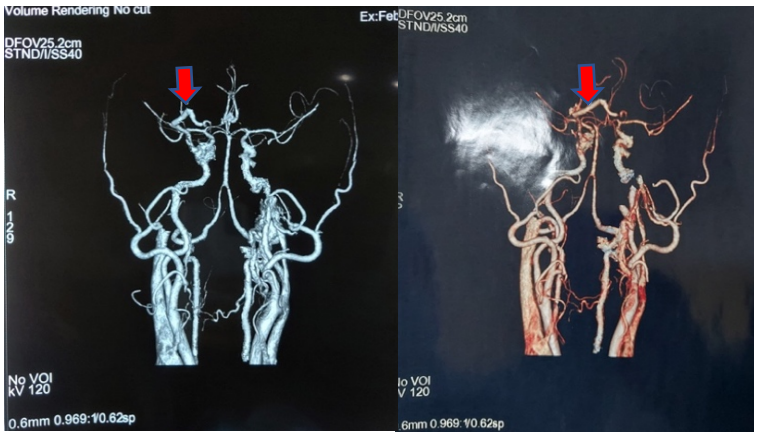

Figure 3. Contrast Enhanced CT Scan Brain-Orbita (a) and CT Angiography (b) of The Patient

Patient was diagnosed with Right Carotid Cavernous Fistula, Secondary Glaucoma on RE, Hypertensive Retinopathy grade III RLE, Moderate Nonproliferative Diabetic retinopathy RLE, Hypertension stage II and Diabetes Mellitus Type II. She was consulted to Glaucoma & Vitreoretina unit. She was treated with citicholine 1x1000 mg per oral, Timolol maleate 0,5% eye drop 2x1 drop on RE, acetazolamide 3x250 mg per oral, Kalium aspartate 1x1 tablet per oral

and advised for external manual carotid compression. The patient was referred to Neuro-surgery Department, Hasan Sadikin Hospital.

The next visit in the following 2 weeks, the patient came with improved visual acuity and the swelling of right eye felt a bit decreasing. The visual acuity was 0.8 pinhole 1.0 on RE and 0.63 pinhole 1.0 f2 on LE. The IOP decreased to 25 mmHg on RE. The other symptoms like double vision, ophthalmoplegia, redness of the eye, bruit were quite similiar as before. The Neurosurgery Department planned to do Digital Subtraction Angiography (DSA) to the patient. The external manual carotid compression was continued while waiting for DSA. Prognosis in this patient are *quo ad vitam dubia ad bonam*, *quo ad functionam dubia ad bonam*, *quo ad sanationam dubia ad bonam.*

Figure 4. Clinical Presentation in 2 Weeks Follow Up and Illustration of External Manual Carotid Compression

III. Discussion

Carotid cavernous fistula is an abnormal communication between the cavernous sinus and the carotid arterial system. The cavernous sinus also drain blood from the ocular veins. The ocular venous outflow begins in the arcade retinal veins, which exit into the central retinal vein (CRV), and in the choroidal veins, then exit the sclera through the vortex veins. Anteriorly, the episcleral venous plexus collects

both blood from the anterior uveal circulation and aqueous percolating through the Schlemm canal. These 3 primary venous drainage pathways empty mainly into the superior ophthalmic vein, which runs posteriorly within the superior medial orbit to the orbital apex, where it crosses laterally to enter the cavernous sinus posterior to the superior orbital fissure.[5-7] The CT Angiography in this patients showed that there is right superior ophthalmic vein dilatation, this condition could give rise to some ocular symptoms, such as the right palpebral oedema due to superior palpebral vein that drained to superior ophthalmic vein was also dilated, episcleral injection and corkscrew appearance, and also raised IOP.

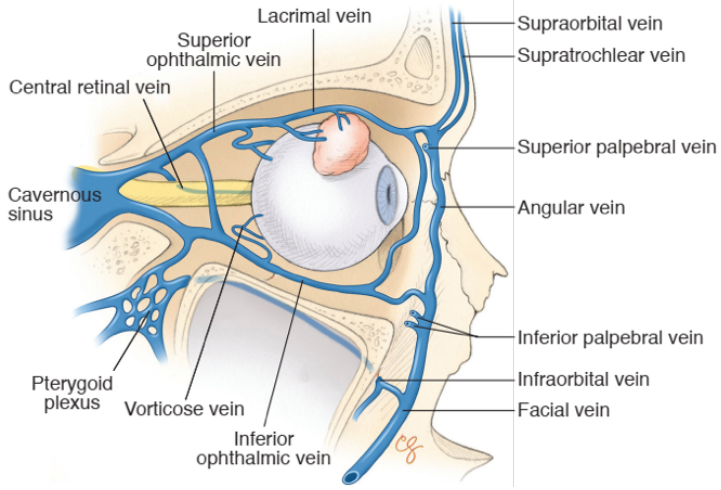

Figure 5. The Ocular Venous Drainage
Source : Cantor, LB[5]

The cavernous sinus comprises a network of venous channels located at middle cranial fossa, bilaterally to the sella turcica. The nerves of the cavernous sinus are cranial nerves (CN) III, IV, V1, V1, V2 and sympathetic plexus around the carotid artery. The CNs III, IV, and V (first and second divisions) run within the dura of the lateral wall of the cavernous sinus as seen in figure 6. The CN VI enters the cavernous sinus and surrounded blood, like internal carotid artery. Carotid cavernous fistula can cause increased intercavernous pressure and leads to compression in adjacent structure. The compression of these structures leads to total ophthalmoplegia and also decrease facial sensation on ipsilateral face due to CN V1, V2 compression.[6-8] [3,5,7] As seen in this patient, she presented with total

ophthalmoplegia due to injury of CN III, IV, VI and also right upper facial paresis due to injury of CN V1 and V2. The ophthalmoplegia gives rise to diplopia symptom.

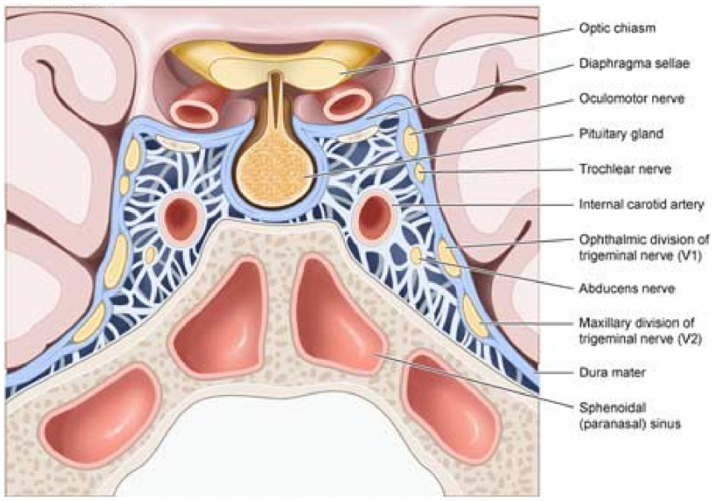

Figure 6. Coronal View Drawing of Cavernous Sinus Anatomy
Source : William, ZR[7]

Carotid Cavernous Fistula can be classified according to etiology as traumatic or spontaneous, according to speed of blood flow (hemodynamic behaviour) as high-flow or low-flow fistulas, and according to anatomy as direct or indirect. Direct CCF originates through direct communication between the internal carotid artery and the cavernous sinus. Indirect CCF originates through indirect communication through the meningeal branch of the ICA, ECA and cavernous sinus (thus not directly with the ICA). More precise classification is according to Barrow into four types as seen in figure 7. Barrow type A or direct CCF is a communication between the cavernous sinus and ICA. Barrow type B or indirect CCF is a dural connections between the cavernous sinus and meningeal connections of the ICA. Barrow type C - indirect CCF is dural connections between the cavernous sinus and meningeal branches of the ECA. Barrow type D - indirect CCF is a communication between the cavernous sinus and meningeal branches of the ICA and ECA. [2,9,10] The CT Angiography of this patient revealed that the CCF was suggested type A Burrow classification in which direct CCF.

The most common cause (70–90 %) of CCF is trauma in the intracranial or periorbital region, whereas indirect or spontaneous CCF occur secondary to spontaneous rupture of dural arteries wall which passed through the sinus. Direct carotid-cavernous fistulas usually have a high through-flow of arterial blood, and are most frequently caused by a traumatic defect in the arterial wall. Direct CCF are mostly caused by severe head trauma. Other cases are secondary to rupture of a cavernous ICA aneurysm, connective tissue disorder, and rarely, spontaneous dissection. [3, 6, 9] The patient had direct CCF but there was no history of trauma. She has risk factor of ICA aneurysm which is female and hypertension stage II, so we can assume that the direct CCF on this patient was due to rupture of ICA aneurysm.

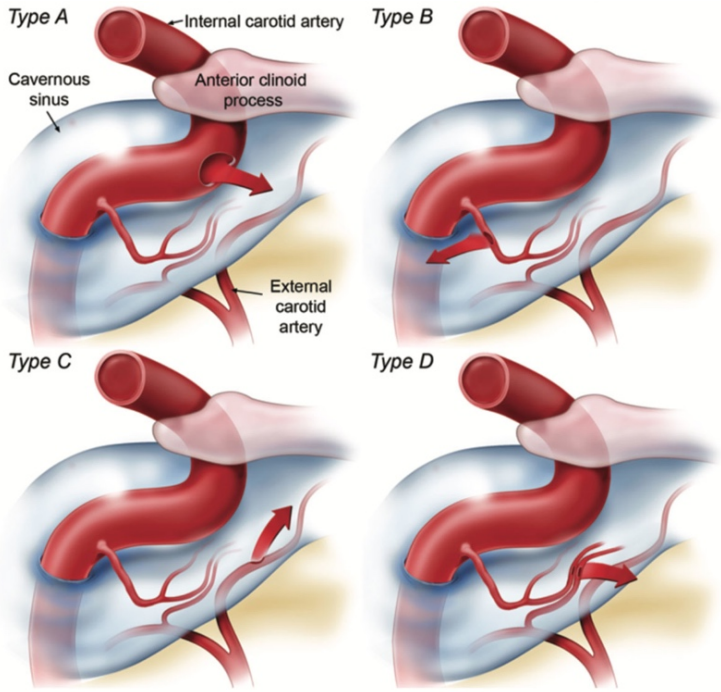

Figure 7. Barrow Classification of Carotid Cavernous Fistulas. Direct (A) and Indirect (B–D) Carotid Cavernous Fistula Subtypes
Source : Peng, TJ9

Clinical symptoms and signs usually present acutely in cases of direct fistula and are more indolent in dural fistulas. The varied clinical presentation of CCFs depends on the anatomy, hemodynamic and size of the CCF fistula. Clinical manifestations

of CCFs are due to progressive congestion of the venous draining into the cavernous sinus. Anteriorly draining fistulas are more likely to cause ocular symptoms. Patients with posteriorly draining fistulas may develop neurologic symptoms, such as confusion and expressive aphasia, as well as diplopia from isolated ocular motor nerve paresis. The clinical triad including proptosis, chemosis, and orbital bruit is the most commonly encountered, subsequent to the arterialization of the superior ophthalmic vein. Other presenting symptoms of CCFs may include tearing, red eye, ocular foreign body sensation, blurred vision, and headache. Decreased visual acuity may be secondary to corneal, retinal or optic nerve changes or may result from the accompanying glaucoma. They can occur from combination of mechanism including exposure keratopathy, venous stasis retinopathy leading to ischemia, central retinal vein occlusion with macular edema or choroidal detachment due to venous congestion. The retinal manifestation ranges from mild stasis retinopathy to frank CRVO.[1,2,9,8] The orbital bruit was audible using stethoscope. The visual acuity in this patient was improved as the IOP decreasing, and the retinal ischemia was found.

The increase of IOP in CCF is mainly caused by increased episcleral and vortex vein pressure. In such cases, closure of the fistula and normalization of circulation has a favourable result in reducing IOP. In other cases, glaucoma may be caused by iris neovascularization due to decreased retinal perfusion or vascular engorgement and edema of the choroid and ciliary body, causing a forward movement of the iris/lens resulting in pupil block glaucoma or may also cause neovascular glaucoma.[2, 4]

Since most of the patients have unilateral presentation (91.7%) and are of low-flow CCFs (87.5%), the high prevalence of the retinal vein dilatation in CCF patients has important implications. Patients with low-flow CCFs have mild signs and symptoms and may be misdiagnosed and poorly responsive to attempted therapy. Additionally they may manifest with a variety of presentations including chronic conjunctivitis, blepharoconjunctivitis, ocular hypertension, secondary glaucoma, episcleritis, thyroid eye disease and others. It was recommended to do a careful dilated fundus examination and comparison with the other eye, which might

provide an important clue to the ophthalmologist to clinch the correct diagnosis.[1, 4, 8] This patient presented with unilateral CCF, swelling and redness of right eye as the initial clinical presentation of CCF.

The diagnosis of CCF is made by neuroanatomic and neurovascular imaging. First-line modalities include CT and CTA or magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) of the brain. Signs of CCF detected with non-invasive imaging include proptosis, dilatation of the superior ophthalmic vein, extraocular muscle enlargement, ipsilateral cavernous sinus enlargement, and skull fracture. Digital subtraction angiography (DSA) is the gold standard in the diagnosis of CCF and must be performed before any intervention. DSA can show filling of the cavernous sinus through the fistula, drainage pattern of the fistula, and presence of reflux into cortical veins following an injection of CCA (common carotid artery), ECA, or ICA.[3, 7, 11] We had performed CT scan and CT Angiography on this patient that revealed type A CCF, then referred the patient to Neurosurgery Department and planned to do the DSA.

The causes of the CCF in this patient was suspected due to aneurysm rupture in ICA. Aneurysms of the cavernous segment of the internal carotid artery usually become symptomatic by mass effect on the adjacent cranial nerves, causing ophthalmoplegia and, sometimes, decreased vision or trigeminal neuralgia. Rupture of an intracavernous aneurysm leads to carotid cavernous fistula (CCF), epistaxis, or subarachnoid hemorrhage if the aneurysm extends through the dura into the subarachnoid space. The primary goal of treatment of CCFs caused by ruptured cavernous sinus aneurysms is closure of the fistula by balloon or coil occlusion of the aneurysm or occlusion of the carotid artery by trapping the fistula. [3, 8, 12]

The main goal in the treatment of CCFs is to preserve flow in the ICA while occluding the fistula. Clinical indications for emergent treatment include progressive ptosis, visual decline, haemorrhage (intracranial or external), and increased intracranial pressure. Endovascular management is the current treatment modality of choice for CCFs. Transarterial embolization is the preferred access method for direct CCFs and transvenous embolization is preferred for indirect CCFs. In the transarterial approach, a microcatheter is passed through the fistula

into the cavernous sinus and coils and/or embolic agents are placed in the cavernous sinus. Cure rates after endovascular repair of CCFs approximate 80%.[5, 11,12]

Conservative management is an option for some low risk, low flow, indirect CCFs. External manual carotid compression of the ipsilateral carotid artery and contralateral jugular vein, several times per day over several weeks, achieves cure in ≈30% of patients. External manual carotid compression (EMCC) should always be performed using the contralateral hand, because this will ensure that the compressing hand falls away should cerebral ischaemia develop. [2, 13] The patient was instructed to do EMCC while waiting for DSA, even though the spontaneously resolved was rare in the direct CCF. In two weeks follow up, the patient had improved visual acuity and decreased palpebral swelling.

High-flow fistulas rarely resolve without treatment, and these lesions clinically manifest with abrupt ocular and cranial nerve features including rapid vision deterioration, proptosis, retro-orbital pain, ophthalmoplegia, ocular injection and chemosis. Spontaneous closure of direct CCFs is rare, with rates estimated to be 1.2%–4%. A 2019 literature review identified 37 patients with 43 direct CCFs that resolved spontaneously. [3, 4, 14]

Carotid cavernous fistula can cause complication such as loss of vision, intracerebral haemorrhage, subarachnoid haemorrhage, epistaxis, or cranial nerve palsies especially in high flow fistula. Patients with direct or indirect CCF usually has reassuring visual prognosis unless there is evidence of optic nerve or retinal ischemia before treatment. Embolization therapy has high successful rate and provide resolution of bruit and IOP. Proptosis, chemosis and ocular misalignment usually improved within weeks of CCF closure and resolve within 3 months. Recurrence may occur in young patients the risk is lower with complete occlusion of the fistula.[3, 6, 7,14] The prognosis *quo ad vitam* is *dubia ad bonam* due to mostly not life threatening condition, but the systemic condition of the patient has high risk of other complication, *quo ad functionam dubia ad bonam* because this visual acuity and visual function of this patient is preserved, but if left untreated this CCF can lead to visual loss. *Quo ad sanationam* is *dubia ad bonam*, because the endovascular treatment has high successful rate, but recurrence may still occur.

IV. Conclusion

Carotid cavernous fistulas (CCFs) are a rare but potentially devastating cause of orbital symptoms, visual loss, and periocular disfigurement. The ophthalmologist must be aware of the various clinical features of CCF. Carotid cavernous fistula patients typically present with proptosis, diplopia, ophthalmoplegia, ocular bruit, elevated intraocular pressure, prominent tortuous conjunctival vessels, and sometimes headache. Diagnosis may be straightforward in cases with direct CCF or may be difficult, especially in cases of dural draining CCF where symptoms are subtle. First-line modalities include CT Scan and CTA or MRI or MRA, but DSA is the gold standard in the diagnosis of CCF. Treatment options must be carefully considered and adopted in a timely manner, owing to the risks of vision loss associated with high IOP, vascular stasis, and ischemia of the retina and other ocular structures. Endovascular treatment is the modality of choice for carotid cavernous fistulas.

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