· Case report ·

Direct carotid cavernous sinus fistula in a 14 years old Malay boy

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Abstract

• A fourteen years old Malay boy was involved in a motor vehicle accident and suffered multiple injuries. The patient was referred to ophthalmology for right periorbital haematoma, ocular examination was normal but proptosis of right eye was detected which was later associated with increase in the intraocular pressure (IOP). Direct carotid cavernous sinus fistula (CCF) was diagnosed by angiography and treated with embolization.

• KEYWORDS: direct carotid cavernous sinus fistula; 6th nerve palsy

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CASE REPORT

A 14 years old Malay boy was involved in a motor vehicle accident while riding a motorcycle on 24th of April 2006 he had history of brief loss of consciousness and sustained multiple injuries, generally the patient was stable haemodynamically, not dyspnoeac but in severe pain. On examination the patient was conscious, oriented and his GCS score was 14/15. CT scan of the brain showed small right temporal subdural haemorrhage with suspicious transverse fracture at the clivus. His vital signs were stable. Abdomen was tender on palpation raising the suspicion of viscus rupture. This was confirmed by CT scan that showed laceration of the spleen. Examination of the limbs showed open fractures of both left femur and right radius, closed fracture of left radius and right clavicle fracture. Urgent laparotomy and splenectomy was done at the date of admission. Four weeks later the patient had another surgery for fixing fractured femur and radius.

The patient was referred to ophthalmology team on 29th of April 2006 for right periorbital haematoma and subconjunctival haemorrhage . On eye examination: Right eye visual acuity was 6/6 and left was 6/6, there was no pain on eye movement, relative afferent pupillary defect was negative. The eye was not proptotic. Extraocular movement testing showed mild reduction of right eye abduction. RE anterior segment showed subconjunctival haemorhage and chemosis. The corneae were clear, pupils round, reactive and regular. AC was deep, IOP reading was normal between 17mmHg and 19mmHg. Fundus examination was normal. There were no commotion retinae, no intraretinal haemorrhage, and no retinal tear. On follow-up of the patient noted that the RE was slightly proptosed associated with dilated and tortuous conjunctival blood vessels. Later proptosis became more prominent and high pitch bruit over right eye was noticed, IOP reading was 25-29mmHg in RE and LE 17-19mmHg. Exophthalmometer reading, RE 22mm LE 18mm base 112mm. There was no RAPD, and fundus examination was normal. Examination of cranial nerves 2nd, 3rd, 4th, 6th, 1st and 2nd divisions of trigeminal were normal. A clinical diagnosis of right direct carotid cavernous fistula (CCF) with secondary increase in RE IOP showing obvious proptosis of the right eye was made.

Medical treatment with antiglaucoma was started to control the IOP with eye drops 5g/L timolol twice daily. CT scan (Figure 1) was requested and showed proptosis of the RE (22mm). Right superior ophthalmic vein was dilated and tortuous and right side of the cavernous sinus is bulky with early left cavernous sinus involvement, to confirm the diagnosis of direct carotid cavernous fistula cerebral angiography (Figure 2) was performed, it revealed direct communication between intracavernous portion of right internal carotid artery and right cavernous sinus with dilatation of right superior ophthalmic vein and inferior petrosal sinus.

Daily checking of the optic nerve functions and IOP were done. It was noted that the IOP continued to rise and after three days Trusopt eye drops was added three times daily as IOP exceeded 30mmHg. However, IOP could not be brought under control and after one week Xalatan eye drops on night was added when IOP readings were 35-37mmHg. With triple therapy IOP ranged between 29-31mmHg and good control was still unachievable, so a decision to add Alphagan twice daily was taken aiming to lower the IOP to a safer level. In subsequent follow-up IOP measurements ranged between 21-32mmHg. Vision was normal there was no RAPD and there was hyperemia of the conjunctiva with prominent dilated conjunctival blood vessels while proptosis remained stable. As four antiglaucoma medications had been used and the IOP was still high and in order to prevent the vision threatening complications, a decision of embolization of the fistula was taken. Embolization (Figure 3) was performed under general

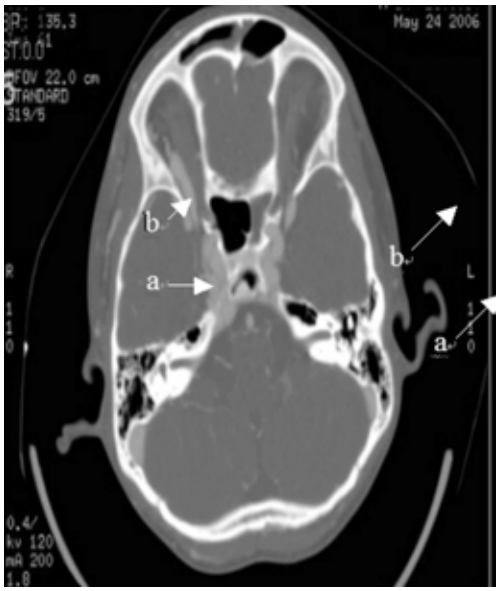

Figure 1 CT scan shows (a) Dilated cavernous sinus and (b) Prominent dilatation of right superior ophthalmic vein.

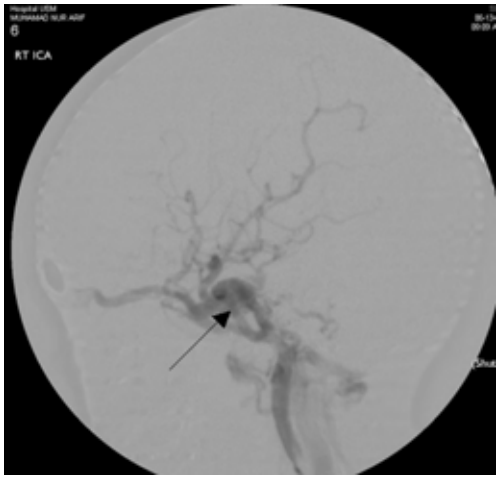

Figure 2 Angiography shows direct communication between intracavernous portion of internal carotid artery and right cavernous sinus.

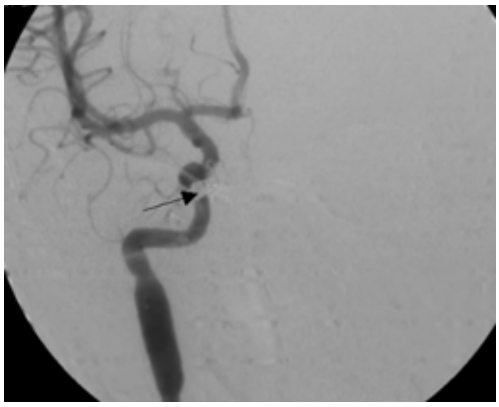

Figure 3 Near total occlusion of the fistula after embolization.

anaesthesia via right internal jugular vein and inferior petrosal sinus by using platinum coil. Post embolization angiography finding showed a near total embolization of the anterior compartment of the right cavernous sinus with no flow to the right superior ophthalmic vein and early shunting to the dilated inferior petrosal sinus is still present. Two days later IOP reduced to 14, 15 and 17mmHg and one week later it was stable on 15mmHg, so Alphgan eye drops was discontinued. On examination of the RE vision was 6/6, there was no RAPD, EOM reduced in out gaze and the patient complained of diplopia. Two weeks post embolization the proptosis reduced significantly but the patient still complaining of double vision which was attributed to abducent nerve palsy. ON next follow-up there was RAPD, visual acuity in RE 6/9 LE 6/6, proptosis resolved but there was extreme limitation of eye movement on abduction and the patient complaining of diplopia. As IOP maintained within normal range Trusopt was stopped. On subsequent follow-up the IOP was maintained within normal range so antiglaucoma drugs were gradually discontinued. Proptosis totally resolved but there was limitation of abduction and there was diplopia in lateral gaze. Glasses mounted with base in prism was prescribed to reduce the diplopia.

DISCUSSION

Direct carotid cavernous fistula (CCF) represent from 70%-90% of all carotid cavernous fistula[1]. Most of direct CCF result from head trauma mainly motor vehicle accidents fall and fight. The cavernous sinuses are a contiguous network of anatomically separate sinusoids between the dura mater and periosteum. The oculomotor, trochlear, abducens cranial nerves, the ophthalmic and maxillary branches of the trigeminal cranial nerve penetrate the cavernous sinusoids bilaterally. Damage to the intracavernous carotid artery or rupture of one of its branches in the cavernous sinus, without concomitant venous injury, may result in an arteriovenous fistula. Thus it is identical in anatomy and haemodynamics to traumatic arteriovenous fistulas else where in the body. The etiology of this traumatic arteriovenous fistula is unique, because other areas of the body require a contiguous injury to an artery and vein to produce an arteriovenous fistula. Once a fistula develops, its course depends on the specific venous composition of the sinus, the blood pressure, and the location and size of the communication. Because of the fistula's vascular communication, most large CCFs will result in bilateral clinical symptoms, however the predominant clinical symptoms are usually ipsilateral based on fistula location. The clinical picture also depends on the venous drainage from the fistula, for example a small fistula can produce significant ocular signs if adequate posterior drainage is not present. However, a large leak with adequate posterior drainage may produce minimal ocular changes. The classification scheme established by Barrow *et al*[2], which is used frequently, divides CCFs into 4 angiographic types, one direct (type A) and three indirect (types B, C, and D): Type A are direct communications between the cavernous segment of the intracavernous carotid artery and the cavernous sinus. Type B are shunts from the meningeal branches of the intracavernous carotid artery to the cavernous sinus. Type C are shunts from the meningeal branches of the external carotid artery to the cavernous sinus. Type D are shunts from the meningeal branches of both the intracavernous carotid artery and the external carotid artery.

The CCFs can be categorized based on their pathogenetic

origin, hemodynamic characteristics, or angiographic findings. The pathogenetic classification divides them into spontaneous or traumatic fistulas. High flow or low flow fistulas constitute the hemodynamic category. The angiographic classification divides the fistulas into direct or dural types. High flow or type A CCFs are high-pressure direct shunts between the internal carotid artery and the cavernous sinus that usually result from trauma or a ruptured aneurysm. More than 75% of CCFs result from trauma, however they occur in only 0. 17% of all patients with craniofacial trauma. Furthermore, penetrating trauma causes CCF less frequently than does blunt trauma. The incidence of traumatic CCF is highest in young men, with males outnumbering females by 2∶1. The CCFs from ruptured aneurysms are most common in middle-aged and elderly women. A traumatic CCF may occur from damage to the intracavernous carotid artery by penetrating objects or, less commonly from a fracture to the base of the skull. Helmke *et al* proposed that the most frequent cause of traumatic direct CCF is the precipitous increase of the intraluminal pressure of the internal carotid artery. Moreover, a CCF may develop immediately following trauma or days to weeks later. In one study by Dubov and Back, 43. 5% of the patients analyzed were diagnosed with CCF more than one month following the initial trauma. The direction of the blood flow through a direct CCF may be posterior into superior and inferior petrosal sinuses or into orbital veins. Although posterior draining fistulas may occasionally cause isolated ocular motor nerves paresis, the most severe ocular manifestations occur in patients with anterior redirection of arterial blood through normal orbital venous channels.

Immediate treatment should be initiated for patients with progressive visual impairment, an unbearable bruit or headache, extreme retro-orbital pain, cortical venous drainage of the fistula, or a traumatic aneurysm that expands beyond the cavernous sinus. Treatment goals include fistula elimination, vision preservation, bruit elimination exophthalmous resolution and cerebral ischemia avoidance. In addition, preserving the patency of the internal carotid artery should be attempted[3]. That many causes of retinal dysfunction associated with CCF, for example, retinal ischemia produced by stagnant anoxia, axonal conduction block produced by mechanical compression secondary to elevated intraocular pressure or compression of the optic nerve or chiasm by a distended cavernous sinus, are potentially reversible with obliteration of the CCF and normalization of cavernous sinus pressures. Optic atrophy, infarction and trauma, they state are more likely to be associated with irreversible optic neuropathy[4]. Post traumatic CCF may undergo spontaneous thrombosis of the with complete resolution of symptoms[5].

The complications of CCFs are not frequent, but potentially life-threatening. They may include intracranial haemorrhage, worsening pulsatile proptosis, exposure keratitis, neovascular glaucoma, blindness, cranial nerves deficits and fatal epistaxis. In minor cases, however, spontaneous healing, probably by thrombosis, can be observed. As many as 90% of patients with direct CCFs may lose vision if not treated. These manifestations are caused by a combination of diminished arterial flow to the cranial nerves within the cavernous sinus, stasis of both venous and arterial circulations within the eye and orbit and an increase in episcleral and orbital venous pressure. The ocular manifestations of a direct CCF are usually ipsilateral to the side of the fistula but they may be bilateral or even contralateral. The lateralization of ocular manifestations depend on venous drainage of the cavernous sinus especially the connection between two sinuses through intercavernous sinus and basilar sinus and presence or absence of the thrombosis within the sinus or superior ophthalmic vein on one side. The most common complaints of patients include:(1) Objective bruit 80%; (2) Visual blurring 59%; (3) Headache 53%; (4) Diplopia 53%;(5) Orbital or ocular pain 35%. Proptosis is one of the most common signs observed in patients with direct CCF. In most cases proptosis develops rapidly on the side of the fistula and becomes pronounced within few days. Some cases have been described in which proptosis has developed months or even years after head trauma. Once proptosis begins it increases slowly for several weeks until it finally becomes stabilized. The eye is usually pushed directly forward. In this case proptosis was detected after two weeks of the head trauma and four days later it became more pronounced with the eye bulged forward.

In early stages of direct CCF eyelids may become moderately or even severely swollen with the time swelling increases and associated with dilatation of periorbital vessels. Arterializations of the conjunctiva and episcleral veins occur as arterial blood is forced anteriorly into orbital veins; conjunctiva and episcleral veins become dilated, tortuous and filled with arterial blood. The presence of a frontal, temporal or orbital "bruit de diable" machinery type of murmur is pathognomonic[6]. The initial symptom is a buzzing sound synchronous with the heart beat. It is usually systolic in timing and decreases in amplitude when the patient is at rest or the affected carotid artery is compressed. Diplopia occurs in about 60%-70% of patients with a direct CCF. It may be caused by dysfunction of one or more of ocular motor nerves, the extraocular muscles or both and the degree of limitation of eye movement varies from mild limitation in only one direction to complete ophthalmoplegia. Ophthlmoplegia may be caused by damage to one or more of the ocular motor nerves by the fistula itself, this damage may be caused by compression of the nerve by the fistula. 50%-85% have isolated 6^{th} nerve palsy. The abducent nerve may be the only one affected or may be damaged along with the 3^{rd} and 4^{th}, Particular vulnerability of the 6^{th} nerve to damage is related to its location within the cavernous sinus. Glaucoma is a potential complication of untreated direct CCF and it develops in 30%-50% of patients at least four types of glaucoma can occur in patients with direct CCF. The most common type is increased episcleral venous pressure, in such cases usually it is mildly elevated as in our patient. In some patients with severe proptosis and chemosis glaucoma is caused by orbital congestion and IOP is usually very high. Neovascular glaucoma develops in some patients with direct

CCF and it is always associated with evidence of retinal hypoxia. Or rarely angle-closure glaucoma due to engorgement of choroidal bed that causes forward shift of the iris lens diaphragm. Glaucomatous optic neuropathy occurs in 20% of patients with untreated CCF. Intracranial haemorrhage occurs in 3% of cases with CCF. The patients with CCF who show dilatation of cortical veins in CT or MRI have a higher risk of cerebral haemorrhage or infarction and should be carefully observed [7].

Indications of treatment are: (1) Progressive vision loss; (2) Proptosis; (3) Ophthalmopegia; (4) Glaucoma; (5) Intracranial bleeding and epistaxis due to pseudo aneurysmal dilatation of ICA into sphenoidal sinus can cause life-threatening epistaxis but the prognosis is excellent[8]. A case has been reported of sudden death due to sever epistaxis that has been attributed to untreated CCF[9].

The gold standard diagnostic imaging study for a suspected CCF is cervical carotid angiography. Angiography provides supplemental anatomical information such as the size and location of the fistula, the presence of collateral circulation from the contralateral arterial blood supply, the blood flow into the fistula and the presence of anatomical anomalies of normal variation in the circle of Willis, and the venous drainage of the fistula. Alternately the presence of dilated orbital veins on a CT scan of the head supports the diagnosis of CCF. The usual CT finding in such cases are (1) Enlargement of ipsilateral cavernous sinus; (2) Enlargement and tortuosity of the superior ophthalmic vein; (3) Enlargement of the extraocular muscles and proptosis. If superior ophthalmic vein >4mm in diameter CCF is suggested. As many as 90% of patient may loss vision in the absence of treatment closure of the fistula is the optimum treatment and usually by endovascular closure using either platinum coils or detachable balloon and usually introduced via ICA they can be introduced by using micro catheter and after release they induce thrombosis and obliterate the fistula. After successful embolization of the fistula as in our patient which was confirmed by postoperative angiography most of ocular signs and symptoms resolved or at least improve and do not recur. The IOP reduced after two days to with normal limit and proptosis reduced significantly. Other signs like dilated conjunctival veins and lid swelling are also resolved. But faint bruit can still be heard which may be due to incomplete closure of the fistula. Ophthalmoparesis attributed to 6th nerve palsy in severe injury it may take months to years to recover or it may remain permanent.

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直接外伤性颈动脉海绵窦瘘 1 例

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摘要

一位14岁的马来男童发生车祸,身体多处受伤。患者因眶周血肿转交眼科,眼部检查正常,但发现右眼眼球突出,后来伴随眼压(intraocular pressure,IOP)增加。经血管造影诊断为直接型颈动脉海绵窦瘘(carotid cavernous sinus fistula,CCF)并给予栓塞治疗。

关键词:直接型颈动脉海绵窦瘘;第六神经麻痹