Digital Subtraction Angiography Therapy for Caroticocavernous Fistula: Case Series Report

Abstract: Objective: The treatment of caroticocavernous fistula is a challenge due to various anatomical and hemodynamic characteristics of a fistula. The most frequent complaints involve the orbital region. Carotid cavernous fistulas (CCFs) are rare potentially sight-threatening abnormal connections between carotid artery and cavernous sinus. We present the use of digital subtraction angiography for caroticocavernous fistula involving orbital region. Material and Method: We report two cases of caroticocavernous fistula involving orbital region. The first patient suffered swelling on his right eye in the last 4 months after he hit a tree. The second patient suffered from a massive swelling on her left eye after she underwent craniotomy procedure. Result: Both patients underwent digital subtraction angiography for diagnosing CCF and were planned for CCF occlusion. The first patient’s follow-up post-caroticocavernous fistula occlusion showed good results in the functional aspect and no significant neurological deficit. The second patient showed a spontaneous resolution of caroticocavernous fistula while waiting for occlusion. There was no recurrence of fistula during short term follow-up. Conclusion: Digital Subtraction Angiography is a safe option for treating caroticocavernous fistula which is manifested at ocular region. This technique has the advantage of preserving visual function without significant neurological deficit.

Keywords: caroticocavernous fistula, digital subtraction angiotherapy, ocular CCF.

INTRODUCTION

Carotidcavernous fistulae (CCFs) as a result of head injury and trauma are rare (Greenberg, 2010). However, they are a serious complication, potentially resulting in subarachnoid hemorrhage and, more commonly, loss of vision. Traumatic CCFs rarely occur in only 0.2% of cerebrocervical or maxillofacial trauma (Greenberg, 2010), but visual loss occurs in up to 90% of untreated cases (Guimarães, De Carvalho, Chone, & Pfeilsticker, 2014).

CCFs are reported to occur in 0.2% of patients with craniofacial trauma and in up to 4% of patients with basilar skull fractures. Regarding etiology, traumatic CCFs account for more than 70% of CCFs overall and are typically found in young male patients following closed head injuries, as observed in the present case. The remaining 30% of CCFs occur spontaneously and are mostly observed in older female patients. The effects of the artery wall are thought to be responsible for the occurrence of CCFs following minor stress (Zhu et al., 2018).

Carotidcavernous fistulae occur when there is a leakage of arterial blood into the venous cavernous sinus. They can be divided into direct and indirect fistulae according to the Barrow Classification System. Direct fistulae are high-flow shunts between the internal carotid artery (ICA) and the cavernous sinus. Up to 90% of direct CCFs occur after trauma (head or maxillofacial injury) or iatrogenic injury (especially after percutaneous trigeminal rhizotomy, rhinoseptoplasty, transphenoidal hypophysectomy, and endovascular procedures). There is a suggestion that CCFs are more likely to develop after closed head injuries (Kaplan, Bodhit, & Falgiani, 2012); they certainly can occur after what is initially perceived to be a minor head injury. The remainder are spontaneous CCFs, which are usually due to a ruptured cavernous ICA aneurysm (Guimarães et al., 2014).
The CCF can be classified by their cause (spontaneous or traumatic), hemodynamic behaviors (high flow or low flow), and angioarchitecture (direct or indirect). Because the clinical manifestations of CCFs are closely related to the anatomical and hemodynamic characteristics of a fistula, Barrow and Peeters et al., proposed a classification of CCFs into four types depending on the arterial supply in 1985, and this classification is preferred since it places more importance on the responsible blood vessels in the pathophysiology of CCF and has a therapeutic implication (Zhu et al., 2018).

The most frequent complaints involve the orbital region. The symptoms and signs of CCF always include eyelid swelling, proptosis, chemosis, and hyperemia, and the condition is commonly misdiagnosed as Graves’ ophthalmopathy or inflammatory conjunctivitis. The classical CCF triad consists of orbital bruit, chemosis, and pulsating exophthalmos, but CCFs can present with other signs and symptoms. These include orbital/retro-orbital pain, orbital frinmitus, proptosis, cranial bruit, pulsatile tinnitus, loss of visual acuity, raised intraocular pressure, and ophthalmoplegia (sixth cranial nerve palsy being the most common) resulting in double vision and pupillary dilatation (Pulhorn et al., 2016).

The evaluation with CT may show fractures, and magnetic resonance imaging of the head shows proptosis as well as engorged intracranial vessels and a convexity of the lateral wall of cavernous sinus. Extraocular muscles may be affected either by eye edema leading to mechanical limitations of movement or by compression of the cranial nerves in the cavernous sinus. Digital subtraction angiography or 4-dimensional computerized axial tomography shows shunting of blood from the ICA into the cavernous sinus (Pulhorn et al., 2016).

Recently, Alexander M et al., and Hamm KD et al. also concluded that CT and MR have a lower efficiency than DSA for the diagnosis of arteriovenous fistulas, which happen in cerebrovascular and microaneurysm in clinical research. DSA remains the gold standard for diagnosing CCF. It can not only help us diagnose CCF qualitatively but also detect the location and range of the nidus and provide detailed information on flow velocity, associated vascular injuries, and high-risk pathways since it can specially observe the blood vessels and detect hemodynamic processes. It can even reveal small dural feeding arteries that are missed on CT or MR. However, the DSA identified the feeding vessel and the topography of the shunt, and the patient was diagnosed with right direct CCF. Thus, DSA is necessary for evaluating the angioarchitecture of the fistula, assessing the feeding arteries, and planning the intervention surgery. Although DSA is an invasive test and has some complications such as thrombosis, cerebral vasospasm, nerve injury or hemorrhage, it is still the gold standard for diagnosing CCF (Zhu et al., 2018).

The therapeutic methods for CCF treatment include conservative management, endovascular intervention, open surgery, and radiosurgery. The current treatment of choice for a CCF is an endovascular approach, and all cases of direct CCF call for interventional treatment. The most important factor affecting treatment is the clinical presentation of the patient. Higashida et al. reported that conservative management can be effective in approximately 30% of indirect CCF and 17% of direct CCF cases. Interestingly, it has recently been reported that 20–60% of patients with indirect CCFs exhibit spontaneous fistula closure. However, transarterial or transvenous embolization has remained the first-line treatment modality for most CCFs because over 90% of cases can be cured successfully with this method (Zhu et al., 2018). There are many options available for the management of CCF, and the treatment is usually tailor-made. Endovascular embolization is an established modality in the management of carotid-cavernous sinus fistula, aimed at excluding the aneurysm and fistula from the circulation and preferably sparing the patency of the internal carotid artery (Yu et al., 2008). Therefore, the authors would like to discuss further about DSA therapy for CCF.

CASE
We report two cases of traumatic CCF. The first patient, a 21-year-old man, was admitted due to swelling on his right eye that occurred in the last 4 months after he hit a tree. The patient also complained of redness of the right eye, watery eyes, and pain when the right eyelid was raised upwards. Physical examination of the right eye showed 6/5 vision, normal vision field, normal intraocular pressure, bruit on auscultation, and no neurological deficit. The patient underwent head CT scan, and the result was dilation of the superior ophthalmic veins, leading to a CCF (carotico-cavernous fistula) image that extends to the posterior subarachnoid aspect, suprassellar and occipital lobe accompanied by bulbus rupture of the right oculi with right peribulboocular hematoma. Then, the patient was diagnosed of post traumatic OD Carotid Cavernous Fistula. The first patient underwent embolization for the occlusion of CCF.

Figure 1. First patient’s initial condition
The second patient, a 54-year-old woman, was referred from the previous hospital on the suspicion of artery or venous malfunction due to the swelling on her left eye. The patient had a history of craniotomy. The physical examination of the left eye showed massive swelling, vision could not be evaluated, intraocular pressure could not be evaluated, and bruit was found on auscultation. The patient underwent brain arteriography, and the result was CCF low flow to superior ophthalmic vein. After brain arteriography evaluation, the fistula disappeared spontaneously.
RESULT
For the first patient, the follow-up post-CCF occlusion showed good results in the functional aspect. The swelling was far decreased, and neither visual deficit nor neurological deficit was found. No recurrence of fistula during short term follow-up was noted.

DISCUSSION
The CCFs may occur spontaneously or following secondary causes (trauma, vascular aneurysm or malformations and venous thrombosis). Direct CCFs are often caused by head trauma or rupture of intra-cavernous aneurysm or head surgery (Marin et al., 2016). This theory is parallel with both patients’
etiology in this study. The first patient had a history of head trauma, and the second patient had a history of head surgery.

In this study, the clinical examination and imaging studies, especially head CT scan and brain arteriography, suggested CCF. Treatment decision depends on the severity of clinical presentations (Gemmete et al., 2010). The treatment of CCFs is suggested if there are worsening visual function, severe proptosis, cranial nerve palsies, intractable bruit, intraocular pressure of more than 25 mm Hg, and increased filling of cortical veins on angiography (Pulhorn et al., 2016). The first patient had bruit on his right eye while the second patient had bruit on her left eye, severe proptosis, and undetected intraocular pressure due to massive swelling.

The first patient underwent digital subtraction angiography procedure. Follow-up post-procedure showed good results in the functional aspect and no significant deficit. The prognosis of CCFs is excellent after endovascular treatment (Latt et al., 2018). One of the mechanisms that has been postulated is the effect of iodinated contrast media on vascular endothelium while angiography for diagnostic was performed which leads to thrombosis (Naragum et al., 2017).

**CONCLUSIONS**

Caroticocavernous fistula is rarely resolved spontaneously. Iodinated contrast media which lead to thrombosis, catheter manipulation, or spontaneous healing of an arterial injury during angiogram might trigger the closure. For most of the cases, caroticocavernous fistula requires adequate treatment for the closure. Digital Subtraction Angiography is a safe option for treating caroticocavernous fistula which is manifested at ocular region. This technique has the advantage of preserving visual function without significant neurological deficit.

**REFERENCES**