

Carotid cavernous fistula with bilateral thalamic infarct

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Abstract

Carotid cavernous fistula (CCF) is an abnormal communication between cavernous sinus and carotid arterial system. Diagnosis depends on clinical manifestations and MRI findings and angiography. Clinical presentation of CCFs is characterized with chemosis, orbital bruit and pulsatile proptosis. Cranial nerve dysfunction and orbital pain might accompany these symptoms. Although spontaneous remission might occur, sometimes CCF might develop life threatening complications. Here in this case we presented a patient with CCF who afterwards developed bilateral thalamic infarct. Cerebral ischemia in CCF is a rare complication and CCF together with bilateral thalamic infarct has not been reported before.

Keywords: Carotid cavernous fistula, Thalamic infarct.

Introduction

A CCF is an abnormal communication between the cavernous sinus and carotid arterial system. These fistulas are classified according to their etiology, anatomy and haemodynamics.¹ Due to etiological classification they are named as spontaneous and traumatic; where as due to haemodynamic classification they are divided into direct and indirect CCF's.¹⁻³ The shunt between the cavernous sinus and intracavernous part of internal carotid artery, is a high-flow fistula which cause direct CCF. The communication between cavernous sinus and meningeal branches of Internal Carotid Artery (ICA) causes indirect CCF which is a low-flow fistula. Post-traumatic CCFs account for 75% of all CCF cases and are commonly direct. Spontaneous CCFs are 25% which are commonly indirect and are rare.² Spontaneous CCFs are also called as Dural arteriovenous fistulas, they are usually associated with atherosclerosis, hypertension and collagen vascular disease. Although spontaneous CCFs are known to be indirect, a few number of direct cases are also reported in literature. Major trauma or rupture of pre-existing aneurysm are the most common reasons of direct spontaneous CCFs.³ CCFs are clinically presented with chemosis, orbital bruit and pulsatile proptosis. Cranial nerve dysfunction may also accompany these features.⁴ Here we are reporting a 65 years old female patient, admitted to the Hospital of Zonguldak Karaelmas University with isolated sixth cranial nerve palsy, orbital pain and chemosis and was diagnosed as CCF. An unexpected complication occurred during the follow up and the patient became unconscious due to bilateral

thalamic infarct. In this report we will highlight the relationship between CCF and bilateral thalamic infarct.

Case Report

A 65 years old female patient was admitted to the hospital of Zonguldak Karaelmas University with the complain of double vision, orbital pain and restriction of ocular motility. She had hypertension in her medical history. Her neurological examination was normal except abducens palsy of right eye. Also subtle chemosis and proptosis were the findings which were detected during the neurological examination. Her laboratory findings were normal. T1-weighted orbital Magnetic Resonance Imaging (MRI) showed signal increase in right lateral rectus muscle and was reported as orbital myositis. But control cranial MRI and MR angiography image showed dilated superior ophthalmic vein and a lesion congruent with CCF near internal carotid artery (Figure-1). Digital Substraction Angiography (DSA) was planned but the patient suddenly became unconscious before the procedure. Another control MRI was conducted, in which bilateral thalamic infarct on the

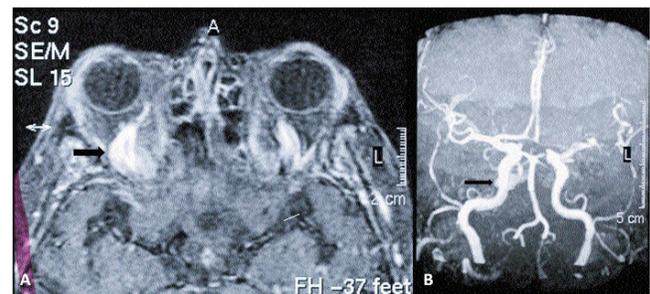


Figure-1:A) Contrast enhanced T1 weighted cranial MR image. Dilated superior ophthalmic vein. B) MR Angiography image. Carotid cavernous fistula.

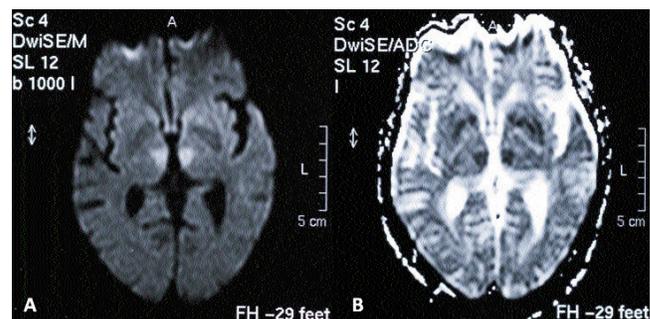


Figure-2: Diffusion weighted MR image.

diffusion weighed cranial MR angiography image was detected (Figure-2). At that moment patient had respiratory insufficiency and was intubated. After a long follow up period in intensive care unit for three months, she was discharged from the hospital bounded to mechanic ventilation device with a tracheostomy. Her last neurological examination (6 months later from discharge) was relevant with akinetic mutism.

Discussion

Clinical manifestations of CCFs might range from asymptomatic to life-threatening conditions. Subtle paralysis of the cranial nerves, together with a series of ophthalmologic alterations related to an increase in the intraocular venous pressure and ischaemia of the retina were frequently observed.⁵ Life threatening complications such as intracranial haemorrhage, increased intracranial pressure, steal phenomena, life threatening epistaxis might be seen, so early detection of fistula is important to prevent these complications.⁶ A sudden onset classical triad of pulsatile proptosis, conjunctival injection with chemosis, and orbital bruit is commonly seen in direct CCFs. Compared with direct CCFs indirect fistulas have gradual onset with a generally milder clinical presentation.^{4,7} Due to this slow onset clinic, indirect variety may be mistaken for chronic conjunctivitis, orbital pseudotumor, orbital cellulitis or thyroid disease. Diagnosing such cases requires a detailed clinical and radiological evaluation and a high index of suspicion.² The patients with CCF have usually ophthalmologic signs but also non-ophthalmologic features might be seen which are nonspecific, like severe headache localized to ipsilateral orbit and pulsatile tinnitus.⁴ Our patient also reported an intractable headache on the ipsilateral side of the affected eye from the onset of her symptoms. She had gradual onset of symptoms with sixth cranial nerve palsy, chemosis and proptosis. Since she did not have a trauma history before onset the type of fistula was considered to be a spontaneous CCF. Chemosis, proptosis and cranial nerve palsy were the clinical signs which support the diagnosis. The patients with CCF might have cranial nerve palsies up to 50%. Abducens paresis is the most commonest type of oculomotor paresis whereas 3rd and 4th cranial nerves may be involved in isolation or combination.⁴ Patients are usually referred to the neurologist due to cranial nerve palsy as in our case.³ The mechanism of cranial nerve paresis in a spontaneous CCF is the swelling of extra ocular muscles due to venous congestion, ischaemia of cranial nerve by vascular steal and compression of cranial nerve by distended sinus.⁸ In our case oedema in the lateral rectus muscle might explain the abducens paresis. For the diagnostic evaluation, CT and MRI studies help to detect proptosis, increased extraocular muscle size and a dilated or thrombosed superior ophthalmic vein.³ Especially as an initial neuroimaging study MR angiography is preferred.⁴ In our case the patient was also diagnosed with the help of MRI and MR angiography which showed dilated superior ophthalmic

vein and the CCF (Figure-1). However DSA is essential for confirmation.⁴ Unfortunately our patient deteriorated and became unconscious before the DSA was performed, hence a control MRI was repeated. Bilateral thalamic infarct was shown on that control MRI (Figure-2). In CCF related complications are variable. Intracranial haemorrhage, subarachnoid bleeding, increased intracranial pressure and cerebral ischaemia are amongst these complications and some of them might be life threatening.⁶ The CCF together with bilateral thalamic infarct had not been reported before as a coincidence or a complication in the literature before. A cerebral vein congestion, as a result of a dural arteriovenous fistula in the posterior cranial fossa, was reported to be the cause of bithalamic lesion only in one case.⁹

Thalamic infarcts are 11% of all vertebrobasilar infarcts. Usually bilateral thalamic infarcts are less common. The thalamus is supplied by paramedian arteries which can arise as a pair from each posterior cerebral artery. But in one third of people they arise equally from a common trunk which is called "percheron artery" and the thromboemboli of this artery causes bilateral thalamic infarct.^{9,10} Furthermore thrombosis of deep cerebral veins could also be the reason of bilateral thalamic infarct.⁹

Our patient had hypertension in her medical history and it's known that hypertension is the most common etiology of bilateral thalamic infarcts¹⁰ or thrombosis of deep cerebral vein might have caused the lesions. In our patient the etiology of bilateral thalamic infarct might also be a venous infarct due to cerebral vein congestion as a result of the CCF. It is known that sinus thrombosis was also reported as a cause of indirect CCF so this might also support the possible association between bilateral thalamic infarct and CCF as in our case.⁷

Conclusion

Spontaneous CCFs might present with subtle ophthalmologic symptoms. In these patients high index of suspicion is critical to diagnose. Such patients should be evaluated with a detailed cranial and orbital imaging modalities. A patient with CCF might have sudden deterioration due to a complication like bilateral thalamic infarct although presented with minor ophthalmologic signs. Hence prompt diagnosis is crucial to prevent life threatening complications in CCF.

References

1. Chaudhry IA, Elkhayry SM, Al-Rashed W, Bosley TM. Carotid cavernous fistula: ophthalmological implications. *Middle East Afr J Ophthalmol* 2009; 16: 57-63.
2. Jayanta KD, Jnanankar M, Pankaj P, Nilutpol B, Kasturi B, Ganesh CK, Hemlata D, Biraj JG. Clinical spectrum of spontaneous carotid-cavernous fistula. *Indian J Ophthalmol* 2007; 55: 310-2.
3. Yu JS, Lei T, C Jin-cao, HE Yue, C Jian, LI Ling: Diagnosis and endovascular treatment of spontaneous direct carotid -cavernous fistula. *Chin Med J* 2008; 121: 1558-62.
4. Subramanian PS, Williams ZR. Arteriovenous malformation and carotid-cavernous fistulae. *Int Ophthalmol Clin* 2009; 49: 81-102.
5. Hamani C, Andrade AF, Figueiredo EG, Ciquini O Jr, Marino R Jr. Spontaneous subarachnoid hemorrhage as the primary manifestation of carotid cavernous fistulas: case report. *Arq Neuropsiquiatr* 2001; 59:593-5.

6. Murata H, Kubota T, Murai M, Kanno H, Fujii S, Yamamoto I. Brainstem congestion caused by direct carotid-cavernous fistula--case report. *Neurol Med Chir* 2003; 43: 255-8.
 7. Gemmete JJ, Chaudhary N, Pandey A, Ansari S. Treatment of carotid cavernous fistulas. *Curr Treat Option N* 2010; 12: 43-53.
 8. Lee KY, Kim SM, Kim DI. Isolated bilateral abducens nerve palsy due to carotid cavernous dural arteriovenous fistula. *Yonsei Med J* 1998; 39: 283-6.
 9. Ameridou I, Spilioti M, Amoiridis G. Bithalamic infarcts: embolism of the top of basilar artery or deep cerebral venous thrombosis? *Clin Neurol Neurosur* 2004; 106: 345-7.
 10. Cabellora PEJ. Bilateral Paramedian Thalamic Artery Infarcts: Report of 10 Cases. *J Stroke Cerebrovasc* 2010; 19: 283-9.
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