

Carotid-Cavernous Fistula Arising and Regressing Spontaneously: A Case Report

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ABSTRACT

Carotico-Cavernous fistulas (CCF) are abnormal communications between carotid arterial system and venous cavernous sinus that may develop spontaneously or after trauma. Spontaneous CCFs are rare and even if they tend to regress spontaneously, permanent neurological deficits are observed up to 20-30% among untreated patients. A 74 year-old female patient admitted to neurology department with a complaining of headache, and blurred vision in her left eye. Her medical history revealed hypertension for ten years. There was no history of trauma. Her neurological examination revealed redness in retina, blurred vision and restricted left eye movements. Her laboratory investigations were unremarkable. Cranial magnetic resonance imaging and angiography was performed and a spontaneous dural CCF was observed. Age, postmenopausal period, and hypertension were suggested as risk factors for CCF. After one week DSA angiography was repeated to perform embolisation however, it was noticed that the fistula was regressed spontaneously. The patient is now being followed for 18 months and has no complaint. In that regard sudden onset headache among middle aged and elderly patients should be regarded as an alarming complaint and further investigations should be performed. In patients with temporal headache, conjunctival edema and / or ophthalmoplegia, CCFs should be considered.

Keywords: carotico-carotis fistulas, spontaneous regression, headache

ÖZET

Karotiko-kavernöz fistüller (KKF) travma sonrası veya spontan olarak, karotis arteriyel sistemi ile kavernöz sinüs venöz yapılar arasındaki anormâl bağlantılar sonucu ortaya çıkmaktadır. Spontan KKF'ler nadirdir. Her ne kadar bu fistüller gerileme eğiliminde olsalar da, tedavi edilmeyen olgularda %20-30 oranında kalıcı nörolojik sorunlara neden olabilmektedir. 74 yaşında kadın hasta baş ağrısı, sol gözde bulanık görme şikâyeti ile nöroloji birimine başvurdu. Özgeçmişinde hipertansiyon öyküsü mevcuttu, fakat herhangi bir travma öyküsü yoktu. Nörolojik muayenede retinada kızarıklık, bulanık görme ve sol göz küresi hareketlerinde kısıtlılık gözlemlendi. Laboratuvar tetkikleri normaldi. Çekilen Kraniyal MR ve ardından yapılan kraniyal anjiyografi sonucunda spontan meydana gelen dural KKF saptandı. Olgunun yaşının postmenapozal evrenin ve özgeçmişinde hipertansiyon öyküsü varlığının KKF gelişiminde risk faktörü olabileceği düşünüldü. Olgunun tedavisinde nöroradyoloji biriminde endovasküler embolizasyon planlandı. İlk anjiyografiden yaklaşık 1 hafta sonra tekrarlanan anjiyografide KKF'nin yine spontan olarak tromboze olduğu ve gerilediği gözlemlendi. Klinik takipte olgunun baş ağrıları ve göz bulguları geriledi. Bu olgu bağlamında ileri yaşlarda temporal bölgede baş ağrısı ile gelen olgularda sekonder baş ağrıları düşünülmeli ve menopoz sonrası kadın olgularda sıklığının fazla olması, görme kaybının görülebilmesi nedeniyle temporal arterit yanı sıra KKF'ler de akılda tutulmalıdır.

Anahtar Kelimeler: karotikokavernöz fistüller, spontan regresyon, baş ağrısı

INTRODUCTION

Carotico-cavernous fistulas (CCF) are abnormal communications between carotid arterial system and venous cavernous sinus. They may develop following trauma or in occasional cases spontaneously and comprise about 10-15% of all intracranial vascular malformations (Quinones et al. 1997). Although they

tend to regress spontaneously, permanent neurological deficits are observed up to 20-30% among untreated patients (Goldberg et al. 1996).

Our aim is to present a case with spontaneous CCF and review the literature from the standpoint of clinical and radiological features as well as treatment modalities.

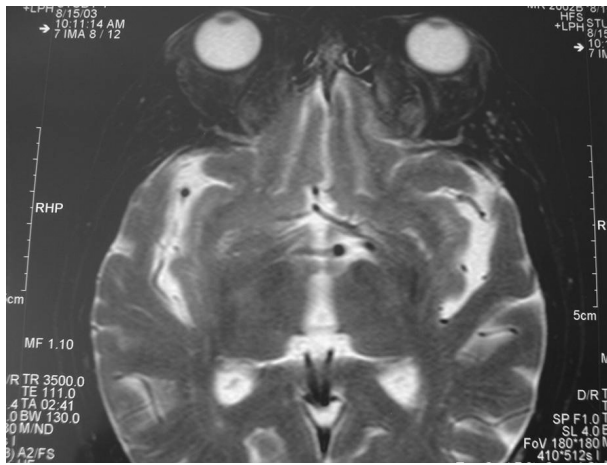


Figure 1 A: Cranial MRI and angiography examinations detecting fistula

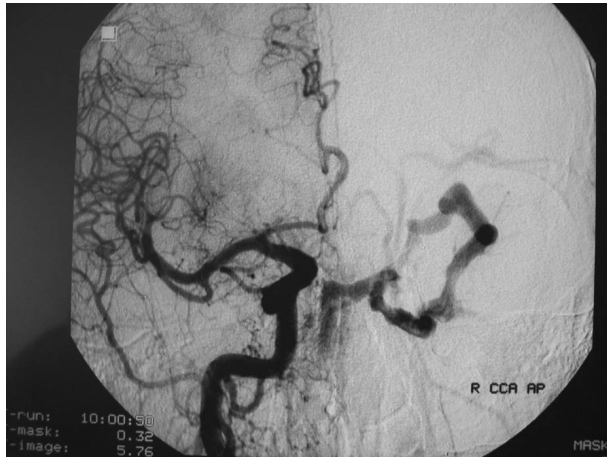


Figure 1 B: Cranial MRI and angiography examinations detecting fistula a

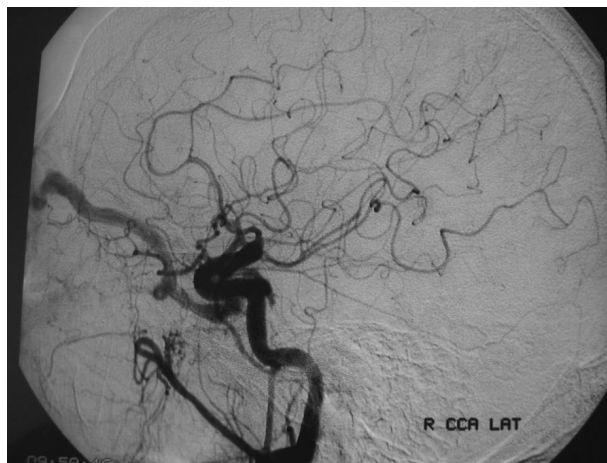


Figure 1 C: Cranial MRI and angiography examinations detecting fistula

CASE PRESENTATION

A 74 year old woman admitted with complaints of headache and blurred vision. Headache was blunt in character, located in left temporal region and persistent for 3 months. Although it was mild initially and responded to analgesic medications, intensity of symptoms increased gradually and blurred vision in the left eye accompanied for 10 days. Her medical history revealed hypertension and depression for 10 years and she was using verapamil for hypertension during this period. Her family history was unremarkable.

On admission she was awake and oriented. Neurological examination was normal except minimal blurred vision in her left eye. Laboratory findings and cranial MRI were normal except increased erythrocyte sedimentation rate and C - reactive protein. Although clinical findings of patient did not meet diagnostic criteria of temporal arteritis precisely, regarding her age, character of symptoms and laboratory findings, low dose corticosteroid therapy was initiated, and significant improvement was achieved. However 4 months later she admitted to our clinic again with complaints of severe headache, visual loss and diplopia in left eye. Upon neurological examination chemosis and restricted movements in her left eye was noted.

Especially left abducens palsy was prominent (Picture I). Repeated Cranial MRI (Figure 1A) demonstrated fistula between carotid artery and cavernous sinus which was proved later by digital subtraction angiography (DSA) (Figure IB, C). After one week DSA angiography was repeated to perform embolisation however, fistula was shown to regress in the second DSA (Figure II). After spontan regression of fistula the patient is now being followed in our out-patient clinic for 18 months and has no complaint. (Picture II).

DISCUSSION

The clinical and radiological findings of our patient were indicating CCF which arose and regressed spontaneously. Spontaneous CCFs evolve from the rupture of carotid aneurysms but CCFs are usually congenital arteriovenous connections formed in the collagen vascular disease, atherosclerosis and hypertension (Djindjian and Merland 1978). Also, in postmenopausal women CCFs are reported much more frequently. In our patient, hypertension and postmenopausal period were regarded as risk factors.

The fistulas are angiographically classified by Barrow et al, Type A fistulas are direct communications between internal carotid artery and cavernous sinus (1985). Type B, C and D are indirect (dural) shunts

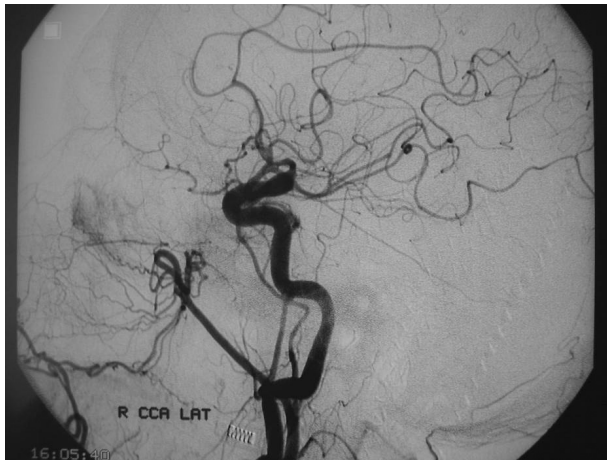


Figure II: Regression of fistula in cranial angiography



Picture 1: Abducens palsy in left eye



Picture II: Normal eye movements

which cavernous sinus could not open directly to the internal carotid artery but via the small meningeal divisions. In our patient dural (indirect) fistula was detected.

Signs and symptoms in CCFs are related to specific anatomy of the cavernous sinus. Since it is directly connected to the ophthalmic vein, abnormal shunt between the sinus and the carotid artery reflects the pressure to the venous system. Thus clinical findings in indirect fistulae are mild compared to direct ones and they progress slowly. Triad of chemosis, pulsatile exophthalmus and ocular signs that are observed in direct fistulae are not seen in indirect forms (Chaudhary et al. 1980, Gioulekas et al. 1997) Chronic red eye due to tortuous conjunctival arteries is frequently encountered in indirect fistulae (De Kiezer 1982).

Cranial MR findings are enlargement of ipsilateral cavernous sinus, torsion and dilatation of the superior ophthalmic vein, enlargement of the extraocular muscles, proptosis, abnormal flow in the affected cavernous sinus, dilatation of intercavernous sinuses and intercavernous veins, lateral wall convexity of the cavernous sinus, ipsilateral or contralateral dilatation of the superior ophthalmic vein and orbital edema. Cerebral angiography is considered as the "gold standard" method in diagnosis and to determine the type of the CCF (Ishida et al. 2003).

Indirect fistulae improve spontaneously in 20-50% of the cases. Interestingly, they may regress spontaneously after the angiography. Unfortunately in 20-30% of the cases visual loss is observed if they are left untreated. On the other hand symptomatic direct fistulae almost always require urgent treatment. The aim of the therapy is to prevent fistulized flow and at the same time to protect the uniformity of the internal carotid artery.

In the clinical practice, indications for treatment are proptosis, visual loss, abducens paresis, severe pain, angiographically increased cortical venous filling, and increased intracranial pressure. In our patient, treatment was indicated since she had severe pain and abducens paresis. During angiography it was shown that the CCF was spontaneously thrombosed. Her complaints were completely disappeared one month after the spontaneous closure of the CCF.

In conclusion sudden onset especially temporal headache among middle aged and elderly patients should be regarded as an alarming complaint and further investigations should be performed. However, clinical, laboratory and radiological findings sometimes could not be satisfactory for accurate diagnosis. In patients with temporal headache, conjunctival edema and / or ophthalmoplegia, CCFs should be considered in the etiology besides temporal arteritis. Detailed history is a must and without objective findings, clinical diagnosis should not be made. Radiological examina-

tions must be considered and ordered logically with expectations that could be problem solving.

REFERENCES

- Barrow DL, Spector RH, Braun IF, Landman JA, Tindall SC, Tindall GT. (1985) Classification and treatment of spontaneous carotid-cavernous sinus fistulas. *J Neurosurg*; 62: 248-256.
- Chaudhary MY, Sachdev VP, Cho SH (1982) Dural arteriovenous malformation of the major venous sinus: an acquired lesion. *AJNR*; 3: 13-19.
- De Keizer RJ (1982) The "red eye"; diagnosis of spontaneous carotidocavernous fistula. *Ned Tijdschr Geneesk*; 126: 144-1450.
- Djindjian R, Merland JJ (1978) Superselective arteriography of the external carotid artery. New York: Springer; 34-35, 405-412.
- Gioulekas J, Mitchell P, Tress B, McNab AA (1997). Embolization of carotid cavernous fistulas via the superior ophthalmic vein. *Aust N Z J Ophthalmol*; 25: 47-53.
- Goldberg RA, Goldey SH, Duckwiler G, Vinuela F. (1996) Management of cavernous sinus-dural fistulas. Indications and techniques for primary embolization via the superior ophthalmic vein. *Arch Ophthalmol*; 114: 707-714.
- Ishida F, Kojima T, Kawaguchi K, Hoshino T, Murao K, Taki W.(2003) Traumatic carotid-cavernous fistula identified by three-dimensional digital subtraction angiography—technical note. *Neurol Med Chir (Tokyo)*; 43: 369-372.
- Quinones D, Duckwiler G, Gobin PY, Goldberg RA, Vinuela F (1997). Embolization of dural cavernous fistulas via superior ophthalmic vein approach. *AJNR Am J Neuroradiol*; 18: 921-928.