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Carotid-cavernous fistula: A case report

Karotiko-kavernöz fistül: Olgu sunumu

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Abstract
Carotid-cavernous fistula is an abnormal arteriovenous communication between the carotid arteries and the cavernous sinus. Imaging studies are the most important step of diagnosis. We herein report a 50-year-old woman with a carotid cavernous fistula, who presented with double vision, right ptosis and ipsilateral pain. All imaging studies were normal except digital subtraction angiography, which revealed a carotid cavernous fistula arising from the meningohypophyseal trunk of right internal carotid artery, internal maxillary artery and middle meningeal artery. The patient underwent endovascular coiling of the fistula. She had an excellent recovery after intervention. In rare situations, carotid cavernous fistula cannot be revealed by computerized brain tomography or magnetic resonance imaging. If there is a strong suspicion of carotid cavernous fistula, digital subtraction angiography must be obtained for diagnosis and treatment.
Keywords: Carotid artery, Fistula, Angiography

Öz
Karotiko-kavernöz fistül, karotis sistemi ile kavernöz sinüs arasındaki anormal arteriyovenöz bağlantıdır. Görüntüleme yöntemleri tanıda en önemli basamağı oluşturmaktadır. Karotiko-kavernöz fistülü olan 50 yaşındaki kadın hastayı sunmaktayız. Hastanın çift görmesi, sağda göz kapağı düşüklüğü ve sağ göz çevresinde ağrısı mevcuttu. Yapılan diğer görüntüleme yöntemleri normal saptanan hastanın dijital subtraksiyon anjiografisinde sağ internal karotis arterin meningohipofisyal arterinden, internal maksiller arterden ve orta meningeal arterden köken alan karotiko-kavernöz fistülü tespit edildi. Hastanın fistülünün koillenmesinin ardından kişi eski sağlığına kavuştu. Nadir durumlarda, karotiko-kavernöz fistül manyetik rezonans görüntüleme ya da bilgisayarlı tomografi ile tespit edilemeyebilmektedir. Eğer hastada fistül olduğuna dair kuvvetli şüphe var ise, dijital subtraksiyon anjiografi yapılması konusunda ısrarcı olunmalıdır.
Anahtar kelimeler: Karotis arteri, Fistül, Anjiografi

Introduction

Carotid cavernous fistula (CCF) is a type of abnormal shunt from the carotid artery to the cavernous sinus [1]. Its symptoms are mainly due to important structures residing in the cavernous sinus [2]. CCFs are classified based on hemodynamics (high flow or low flow), pathogenesis (spontaneous or traumatic), and angiographic anatomy (direct or indirect) [3]. Its main treatment is ligation. We herein report a case of CCF with radiologic findings.

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Case presentation

A 50-year-old woman with a history of falling down the stairs and suspected trauma to the head one month earlier presented with double vision, right ptosis and ipsilateral retro-orbital pain. She had bilateral cataract surgery fifteen days before. Examination of right eye revealed ptosis, near complete ophtalmoplegia, poor light response and midriasis (6mm). She had hypoesthesia on cranial nerve V_1(ophthalmic nerve). There was no oscultation finding over the globe. Fundoscopic examination was also normal. Further general physical examination showed no abnormalities. Her blood tests including infectious disease testing and vasculitis panel were normal. Computed brain tomography (CT), contrast-enhanced cranial magnetic resonance images (C-MRI), cranial magnetic resonance angiography (CMRA), constructive interference in steady state (CISS) sequence cranial MR were all normal. Sterile lumbar puncture was performed with an opening pressure of 17 cm CSF, analyses of which were normal. Due to neurological examination findings pointing out cavernous sinus pathologies, cavernous sinus focused cranial MRI was performed, which revealed nothing. Because vascular pathology was suspected, digital subtraction angiography was planned, which revealed a carotid cavernous fistula arising from meningohypophyseal trunk of right internal carotid artery, internal maxillary artery and middle meningeal artery (Figure 1). The patient underwent endovascular coiling of the fistula under general anesthesia (Figure 2). On the visit a month after surgery, her examination was all normal.

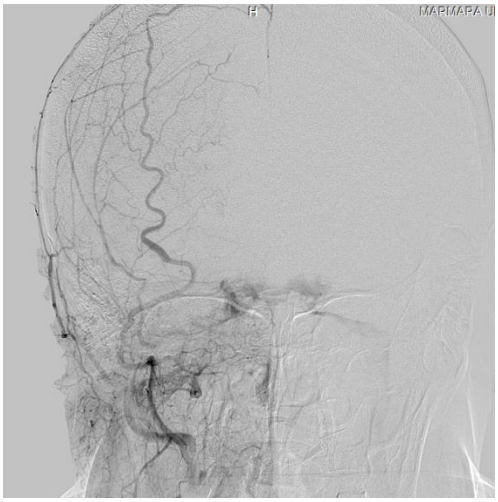

Figure 1: Carotid cavernous fistula arising from the meningohypophyseal trunk of right internal carotid artery, internal maxillary artery and middle meningeal artery

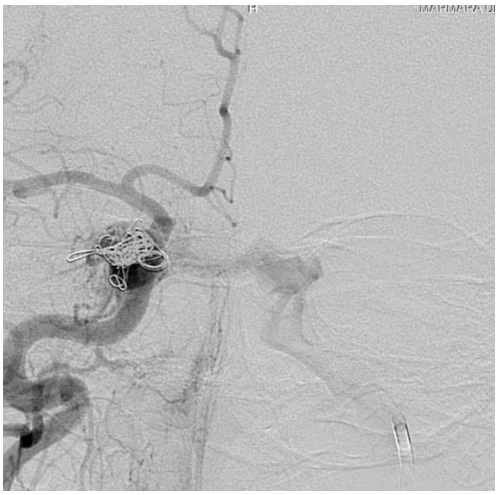

Figure 2: Endovascular coiling of the fistula

Discussion

Carotid cavernous fistula is an abnormal arteriovenous communication between the carotid arteries and the cavernous sinus [1]. It can arise spontaneously or from secondary causes (e.g. head trauma). If there is a direct connection between intracavernous carotid artery and sinus, it is termed “a direct fistula” and has high flow. Indirect or dural fistula implies that a fistula is located between the internal or external carotid artery branches and the sinus, and it has low flow. As arterial blood enters the cavernous sinus under high pressure, main pathology is arterialization of orbital veins. The symptoms of direct CCFs are proptosis, chemosis, painful ophthalmoplegia, and conjunctival injection. The onset of symptoms is almost always acute and rapidly progressive [3]. On the other hand, indirect CCFs have slower progression than direct CCFs. Symptoms depend on the venous drainage type. If it drains anteriorly by the superior ophthalmic vein, orbital symptoms like chemosis, proptosis can be observed, but if the drainage is posterior through the inferior or superior petrosal sinus, orbital symptoms and signs are usually absent [1,4]. Patients with low flow CCFs usually have mild signs and symptoms and may be misdiagnosed [5].

The disorders causing painful ophthalmoplegia may be considered for differential diagnosis, which include trauma, infection, neoplasms, vascular problems or miscellaneous. A detailed history, examination findings and imaging studies help the clinician to rule out these conditions.

Imaging studies include computed tomography (CT), CT angiography (CTA), magnetic resonance imaging (MRI), MR angiography (MRA) and sometimes orbital ultrasonography. Both CT and MRI show proptosis, extraocular muscle enlargement, and dilation of the cavernous sinus and superior ophthalmic vein. Although CTA has better sensitivity than MRA in detecting CCFs, especially in cases where the fistula lies in the proximal portion of the cavernous sinus, 3-D time of flight (3-D TOF) MRA has a 100% specificity in revealing a fistula[6]. Digital subtraction angiography should be used if the diagnosis cannot be made through other imaging techniques. It is both a diagnostic and a therapeutic test.

The goals of therapy are treatment of ophthalmologic complications and closure of CCF. The mainstay of therapy is endovascular occlusion of the affected cavernous sinus with coils, glue, or balloons. Transarterial embolization is the main method for direct CCFs and transvenous embolization is preferred for indirect CCFs [7]. Surgery and other interventions (e.g stereotactic radiosurgery) can be considered when the endovascular treatment is not possible or is unsuccessful. In one series, 9% of patients had a recurrence [8], but in general the success of treatment is pretty high.

Conclusion

CCFs are a rare but treatable cause of orbital injury and vision loss. Obtaining an accurate history of the onset of symptoms is important as they can explain the etiology of the CCFs. Some options are available for the management of the shunt and the goal is to achieve complete occlusion of the fistula while preserving normal carotid flow. Since most CCFs are not life-threatening, treatment as quickly as possible is necessary to prevent permanent injury to the involved eye.

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