Radiology Case Reports 18 (2023) 3876–3880

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Case Report

Carotid cavernous fistula (CCF) with coil embolization in interventional radiology: A case report ☆

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ARTICLE INFO

Article history:
Received 19 June 2023
Revised 2 August 2023
Accepted 3 August 2023
Available online 26 August 2023

Keywords:
Carotid cavernous fistule
CT-angiography
Digital subtraction angiography

ABSTRACT

Carotid cavernous fistula (CCF) is a condition with abnormal flow from the carotid artery to the cavernous sinus. The characteristic clinical findings are ophthalmic monoplegia, ptosis, exophthalmos, chemosis, Horner's syndrome, facial pain, loss of sensation in the distribution of the trigeminal nerve, and headache. Digital subtraction angiography (DSA) is the gold standard modality for evaluating CCF. An 18-year-old patient came with decreased bilateral visual acuity after a traffic accident. CT angiography was performed on the patient and found a dilated right superior ophthalmic vein. CCF is present in the patient. The DSA procedure was performed on the patient, and embolization was performed on the right ophthalmic vein. On post-embolization angiography, the tubular structure is no longer visible, which is bilateral superior ophthalmic vein dilatation.

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Introduction

Carotid cavernous fistula (CCF) is where there is abnormal flow from the carotid artery into the cavernous sinus either directly or indirectly [1]. CCF is currently classified based on 3 conditions, hemodynamic status (high flow & low flow), etiology (spontaneous & traumatic), and fistula anatomy (direct & indirect). Another classification for the diagnosis of

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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https://doi.org/10.1016/j.radcr.2023.08.018

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(a) (b)

Fig. 1 – Local status of the bilateral orbital region: precoiling (a) and postcoiling (b).

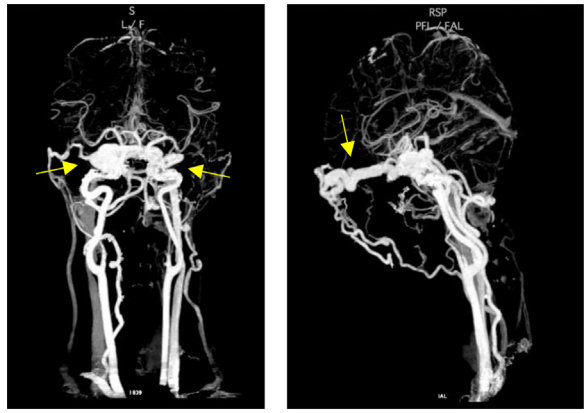
CCF is Barrow's criteria which divides CCF into 4 types: type A, type B, type C, and type D [2].

Direct fistulas (type A in Barrow's classification) are usually caused by penetrating or blunt trauma. In contrast, indirect fistulas (types B, C, and D in Barrow's classification) are generally caused by nontraumatic conditions such as hypertension, fibromuscular dysplasia, type IV Ehlers-Danlos and internal carotid artery dissection [3]. The clinical presentation in patients with CCF is often associated with ophthalmic disturbances, such as ophthalmic monoplegia, ptosis, loss of sensation in the distribution area of the trigeminal nerve, exophthalmos, facial pain, chemosis, Horner's syndrome, and headache [4].

Digital subtraction angiography (DSA) is a method for determining the condition of blood vessels by assessing whether there are abnormal conditions [3]. The main goal in treating CCFs is maintaining the integrity of the flow in the internal carotid artery. The therapy itself is mainly done by closing the fistula [5]. Clinical indications for emergency management of CCF are progressive ptosis, decreased vision, bleeding (intracranial or external), and increased intracranial pressure [5]. The cure rate in CCF patients after endovascular treatment is approximately 80%. On the other hand, patients with indirect CCF may recover spontaneously [6].

Case report

An 18-year-old presented to our institution with decreased bilateral visual acuity. The reduced bilateral visual acuity began to appear about 4 months before coming to the hospital. The vision on the right side is more blurry than the left side. Another complaint is swelling in both eyes area (Fig. 1a). In the past, patients fell onto the asphalt while riding a motorbike because they collided with another motorbike from the opposite direction. The patient was not wearing a helmet at the time of the incident and was unconscious for 30 minutes after the accident. After the fall, the patient experienced complaints of vomiting twice, nose bleeding, and ringing in the right ear. There were no seizures or mouth and ear bleeding. The patient has normal hearing and olfactory function. History of diabetes mellitus, hypertension, tuberculosis, allergies, and malignancy in the family was denied. On local physical examination of the right orbital region: there is a superior & inferior palpebral hematoma and conjunctival injection. No lacerations, hyperemia, ptosis, and corpus alienum were found.



(a) (b)

Fig. 2 – Results of 3-dimensional CT-angiography (a) anterior and (b) lateral dextra. There were findings of bilateral superior ophthalmic vein dilatation (yellow arrow) associated with the left and right internal carotid arteries.

Radiological finding

CT-angiography

The patient underwent MSCT cerebral angiography to reconstruct the head and neck vessels, MPR, MIP, vascular volume rendering, and MIP angulation setting on November 8, 2022 (Fig. 2). Scanning was performed using ultravist intravenous contrast 370 mg, 50 mL with a flow of 17 mL/s bolus tracking. The size, shape, and position of the right internal carotid artery, right and left middle cerebral artery, and right and left anterior cerebral artery were normal. The size, shape, and position of the anterior and posterior communicating arteries and the pericallosal arteries appear normal. A tubular structure is seen in the right retrobulbar space dilated by the right superior ophthalmic vein. This structure is related to the right internal carotid artery with a diameter of 0.89 cm and a length of 1.83 cm, which causes exophthalmos in the right bulbus oculi. A tubular structure is seen in the left retrobulbar space, which appears to dilate the left superior ophthalmic region, causing exophthalmos of bulbus oculi. The distance between the anterior margin bulbus oculi from the right inter zygomatic line is ± 2.14 cm and 2.4 cm to the left. The distance of the posterior margin bulbus oculi from the right interzygomatic line is ± 0.39 cm and ± 0.26 cm to the left. After the CT Scan examination, the patient underwent ORIF with indications of a parasymphysis et orbita dextra and mandibular fracture. The patient is also suspected of having a CCF.

DSA procedure

The patient underwent the DSA procedure.

The DSA procedure begins with aseptic and antiseptic actions on the right and left inguinal areas. Then the patient has performed angiography of bilateral internal carotid arteries and their branches with an approach through the bilateral femoral arteries. A tubular structure is seen, which is

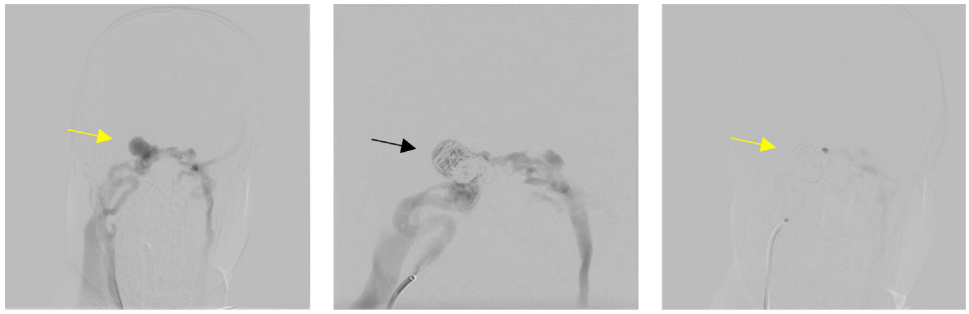


Fig. 3 – (a) Pre-DSA, (b) DSA with coiling (black arrow) (c) No longer visible tubular structure, which is a dilatation of superior ophthalmic veins (yellow arrow).

a dilatation of the right superior ophthalmic vein that seems to be connected to the left superior ophthalmic vein through the anterior intercavernosus (Fig. 3a).

Superselective catheterization was performed with a microcatheter on the right external carotid artery branch, then embolization was performed using a detachable coil on the right superior ophthalmic vein (Fig. 3b). On postembolization angiography, the tubular structure, which is bilateral superior ophthalmic vein dilatation no longer visible (Fig. 3c). CCF diagnosis obtained postembolization. The patient's complaints of swelling in both eyes area slightly decreased after embolization (Fig. 1b).

Discussion

Definition

CCF is a case where there is an abnormal vascular shunt that allows blood to flow directly or indirectly from the carotid artery into the cavernous sinus [1].

Classification

CCF is classified based on 3 types, namely hemodynamic (high flow & low flow), etiology (spontaneous & traumatic), and anatomy of the fistula (direct & indirect). In addition, Barrow's criteria are also used in the classification of CCF. These criteria divide into 4 types, type A, type B, type C, and type D [2].

Barrow's classification type A is a direct fistula, while Barrow's classification types B, C, and D are indirect types [2]. A direct fistula is usually a high-low fistula characterized by a direct connection between the internal carotid artery (ICA) and the cavernous sinus. Barrow type B fistula affects the meningeal branches of the ICA, Barrow type C involves the external carotid branches, and Barrow type D fistula affects the meningeal branches of both the internal and external carotid arteries. Barrow fistula type D is usually a spontaneous fistula [2].

Pathophysiology, etiology, and patient presentation

Until recently, the cause of direct fistulas (type A Barrow's classification) was usually penetrating or blunt trauma. Other causes include rupture of the ICA aneurysm in the cavernous sinus, Ehlers-Danlos syndrome type IV, and iatrogenic intervention. Causes of iatrogenic interventions include transarterial endovascular interventions, internal carotid endarterectomy, percutaneous treatment of trigeminal neuralgia, resection of trans-sphenoidal tumors, pituitary, and maxillofacial surgery [2]. Barrow's classification of CCF type A is so far known to be the most common type of CCF [5]. The most common cause of type A CCF is a traumatic injury, with a higher prevalence in young men. This traumatic CCF is usually unilateral but may occur bilaterally in 2% of patients [5].

In Barrow's classification, indirect causes of fistulas (types B, C, and D) are usually nontraumatic causes, including hypertension, fibromuscular dysplasia, Ehlers-Danlos type IV, and ICA dissection. Postmenopausal women are also reported to frequently experience indirect CCF [2]. Indirect CCF processes in the postmenopausal group often occur typically and spontaneously [3]. However, this condition can also occur in other age groups due to pregnancy, sinusitis, or cavernous sinus thrombosis [3]. The exact mechanism by which CCF spontaneously causes is unknown. However, it may be related to several events, such as venous thrombosis in the cavernous sinus and weakening of the arterial wall from predisposing factors such as hypertension, atherosclerosis, or connective tissue disease [5]. Indirect CCF is a low-flow lesion with a high probability of spontaneous resolution [6].

Clinical manifestations

Clinical manifestations of indirect CCF usually have a lower severity compared to clinical manifestations of direct CCF [6]. Vascular malformations in CCF cases are known to interfere with the cavernous sinus, the anatomical site traversed by the ICA and cranial nerves III, IV, V1, V2, and VI [5]. Thus, neurological abnormalities of these nerves may occur.

The classic manifestations usually present in cases of CCF are ophthalmic monoplegia, ptosis, exophthalmos, chemosis,

Horner's syndrome (characterized by the presence of partial ptosis and miosis, with or without anhidrosis), facial pain or loss of sensation in the distribution of the trigeminal nerve, and headache [4]. Based on the percentage of occurrence, these clinical findings were found in patients in the form of proptosis as much as 72%-98%, chemosis 55%-100%, orbital bruits 71%-80%, and headaches 25%-84% [1]. Most patients also complain of visual disturbances, including diplopia in approximately 88% of patients, blurred vision, and orbital pain. These visual complaints are thought to occur due to retinal ischemic conditions. Other disorders of the organs of vision, such as ophthalmoplegia, were reported to occur in approximately 23%-63% of patient cases. Clinical findings of cranial nerve deficits have been reported in 17%-44% of patients [1]. Another rare clinical finding in CCF patients is intracerebral and subarachnoid bleeding, which occurs in 5% of patients [1].

Treatment

DSA is the procedure of choice to accurately define vascular architecture to assess the presence of an injured process and pattern of vascular drainage [3]. Angiographic evaluation of DSA is carried out in 2 phases, diagnostic catheterization and super selective catheterization.

The main goal in therapy for CCFs is to maintain flow in the ICA while closing the fistula [5]. Clinical indications for emergency management of CCF are if there are findings of progressive ptosis, decreased vision, bleeding (intracranial or external), and increased intracranial pressure. Indications for the need for angiography itself for emergency management are the presence of cavernous sinus varices, pseudoaneurysms, cortical venous drainage, and thrombosis of the distal venous pathway [5].

Endovascular management is the modality of choice for CCF [5]. Transarterial embolization is the method of choice for direct CCFs, and transvenous embolization is preferred for CCFs. In transarterial endovascular management, a microcatheter is passed through the fistula into the cavernous sinus. Then, a coil and/or embolic agent, such as Onyx (Covidien, Ireland), is placed in the cavernous sinus. In the transvenous approach, the cavernous sinus is most often accessed via the inferior petrosal sinus. The cure rate after endovascular treatment of CCF is approximately about 80% [5].

In this case, the patient had an ophthalmological complaint of decreased bilateral visual acuity since 4 months ago. Complaints of the right eye are felt to be heavier than the left. Several weeks earlier, the patient had a traumatic condition due to a traffic accident. The pathological condition in the patient was a fracture of the parasymphysis of the dextra mandible and the dextra orbit. The patient has been treated for trauma. The ophthalmological pathological condition in the patient is thought to be the result of a previous traumatic process. The underlying mechanism is an anatomical abnormality that affects the location through which the eye nerve passes. DSA action was performed to confirm the presence of CCF. Based on the type, the patient has CCF type A in Barrow's classification. This is consistent with data where type A is closely related to traumatic events.

Conclusion

CCF occurs when an abnormal shunt allows blood to flow from the carotid artery into the cavernous sinus. CCF can occur as a result of traumatic conditions or as a result of spontaneous events. CCF due to trauma, which is included as a direct CCF according to Barrow's classification, requires DSA action as a gold standard. The goal of treatment is to optimize internal carotid artery flow.

Patient consent

Written informed consent for publication of their case was obtained from our patient.

Statement of informed consent

Purpose of the Study: The purpose of this study is to add insight about diagnosing Carotid Cavernous Fistula early and handling it, where Carotid Cavernous Fistula are rare cases so that they can be used as learning material.

Confidentiality: All personal data obtained is confidential which is guaranteed by the author. The author will not publish personal information of subjects that is not related to research interests.

Participation: Subject participation in this study is voluntary. Subject will not be compensated for the participation in this study. Subject may withdraw his participation at any time. Subject may also choose not to participate in this study.

Risks: Subjects will not be compensated for their participation. However, the subject's family can get an explanation about the disease and its treatment. If the family wishes, the author will provide an initial report on my findings to the subject's family.

Informed Consent: Subjects voluntarily participate in this study. Subject agree that Subject have given the opportunity to ask questions and have them answered to Subject satisfaction. Subject have received a copy of this consent form signed by the researcher.

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