CASE REPORT
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Ophthalmological presentation of carotid cavernous fistulas: case series and a paradigmatic case report

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ABSTRACT

The purpose of this paper is to describe the clinical presentation of carotid cavernous fistulas (CCFs) managed in Hospital Egas Moniz between January 2010 and December 2020. Patients presenting with orbital congestive signs or symptoms with an ophthalmological evaluation and a diagnosis of CCF confirmed by digital subtraction angiography were included retrospectively. Carotid cavernous fistulas are rare and difficult to diagnose. Direct CCFs tend to have a more severe, acute onset presentation, while indirect CCFs are associated with a more insidious onset and chronic course. Anterior and inferior drainages are the most common in both direct and indirect CCF. Clinical presentation can be varied and nonspecific; however, patients commonly present with ophthalmological manifestations. All of our case series patients presented with ophthalmological complaints. The most frequently seen signs or symptoms were red eye (n = 8), increased intraocular pressure (IOP) (n = 8), and ophthalmoparesis with diplopia (n = 7).

KEY WORDS: ophthalmology; carotid cavernous fistulas; clinical presentation

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INTRODUCTION

Carotid-cavernous fistulas (CCFs) result from an anomalous connection between the internal and/or external carotid arteries and the cavernous sinus.

The cavernous sinus comprises a network of venous channels through which the cavernous portion of the internal carotid artery (ICA), the internal carotid sympathetic plexus, and cranial nerve (CN) VI course. The CNs III, IV, and V (first and second divisions) run within the dura of the lateral wall of the cavernous sinus [1].

Carotid-cavernous fistulas can be classified by:

- etiology: traumatic or spontaneous;
- flow volume and speed: high or low;
- angiographic architecture: direct or indirect.

The most commonly used classification scheme for the latter was established by Barrow et al. He divided CCFs into four types, depending on the arterial feeders. Type A fistulas represent direct communications between the ICA and the cavernous sinus, usually associated with high flow rates. Indirect fistulas (types B, C, and D) are dural CCFs fed by the

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meningeal branches of the ICA, the external carotid artery, or both [1, 2]. Direct CCFs occur most commonly after trauma or surgical procedures, predominantly in young men. On the other hand, indirect CCFs are usually seen in elderly postmenopausal women or during pregnancy and are associated with hypertension, atherosclerosis, neurofibromatosis, or collagen vascular disorders [3, 4].

Carotid-cavernous fistulas drainage pathways may affect the clinical presentation and treatment approach. They may drain anteriorly via ophthalmic veins, inferiorly via pterygoid plexus and inferior petrosal sinus, contralaterally via intercavernous connections, posteriorly via deep venous system, superior petrosal sinus, and cerebellar veins, and superiorly via superficial middle cerebral vein [4].

Orbital doppler ultrasonography (US), computed tomography (CT), magnetic resonance imaging (MRI), and digital subtraction angiography are valuable imaging diagnostic methods, the latter being the gold standard for diagnosis while allowing for planning and execution of therapeutic procedures [5, 6].

This paper aims to describe a paradigmatic clinical case and the clinical presentation of CCFs managed in Hospital Egas Moniz between January 2010 and December 2020.

MATERIAL AND METHODS

Patients presenting with orbital congestive signs or symptoms between January 2010 and December 2020 with an ophthalmological evaluation and a diagnosis of CCF confirmed by digital subtraction angiography were included retrospectively. Demographic, clinical, and angiographic data were collected. Special care was given to individual information such as age, sex, history of previous brain injury, ophthalmological signs and symptoms at presentation, and angiographical findings that allowed classification of the fistulas, according to Barrow et al. [2].

A descriptive study of our case series was performed, and a paradigmatic case was depicted to illustrate the clinical picture.

RESULTS

Ten patients were identified, 7 of which female, with an average age of 62.7 years old (Tab. 1). All of our patients presented with ophthalmological complaints. The most frequently seen signs or symptoms were red eye (n = 8), increased intraocular pressure (IOP) (n = 8), and ophthalmoparesis with diplopia (n = 7). Regarding the latter, five patients had paresis of the abducens nerve, while one had a third nerve palsy and another had both. Proptosis and chemosis were observed in 5 and 4 patients, respectively. Three patients reported tinnitus and decreased visual acuity, fundus abnormalities were detected also in three patients. One patient presented with a headache. With respect to the angiographic classification, two CCFs were direct and six indirect. One case was not classified due to images not being available in our archiving system. The other case referred to an intra-clival Cognard type IV arteriovenous fistula that drains to the cavernous sinus and, therefore, resembled a CCF. Two patients were managed conservatively, while the remainder were submitted to endovascular procedures. The most common persistent signs or symptoms, one week after treatment, were increased IOP (n = 7) and ophthalmoparesis with diplopia (n = 5) (Tab. 2).

Table 1. Descriptive statistics

Whole sample (n=10)	
Mean age at diagnosis (years)	62.7
Sex (female)	7
Fistula classification	
Direct	2
Indirect	6
Cognard type IV	1
Treatment modality	
Conservative	2
Endovascular	8
Reintervention	1

Clinical case

A seventy-five-year-old woman was admitted to the emergency department complaining of diplopia and red eye for a week. Besides a recent arterial hypertension diagnosis, she was healthy and had no relevant ophthalmological clinical history.

The patient's best-corrected visual acuity was 1.0 in both eyes. There was normal pupillary response bilaterally, with normal color vision and visual field examination. Eye movements assessment showed a dextroversion limitation with diplopia. IOP was 32 mm Hg in the right eye (RE) and 16 mm Hg in the left eye (LE). Clinical examination revealed a RE ptosis and proptosis with conjunctival and episcleral injection (Fig. 1), with no

Table 2. Presentation signs and symptoms (T0) and after treatment (T1)

Whole sample (n = 10)	T0	T1
Increased IOP	8	7
Red eye	8	3
Diplopia	7	5
Ophthalmoparesis	7	5
Proptosis	5	2
Chemosis	4	0
Tinnitus	3	1
Decreased visual acuity	3	0
Fundus abnormalities	3	0
Headache	1	0
Relative afferent pupillary defect	0	0
Retrobulbar pain	0	0

IOP — intraocular pressure

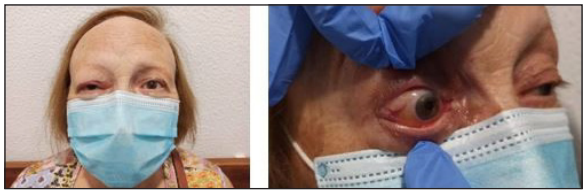

FIGURE 1. Presentation on admission. Note the right eye (RE) ptosis and episcleral veins dilatation

other findings in the anterior segment and fundus slit-lamp examination. Gonioscopy exposed the presence of blood in the Schlemm canal.

The patient underwent a cranial CT angiogram that showed indirect signs of a CCF, with asymmetric superior ophthalmic vein caliber, showing engorgement and arterialization on the right (Fig. 2). A digital subtraction angiography confirmed the presence of a dural arteriovenous fistula involving the posterior aspect of the right cavernous sinus (Fig. 3AB). The patient was submitted to endovascular treatment with the placement of coils in the cavernous sinus and posterior portion of the right superior ophthalmic vein via the facial vein. Digital subtraction angiography at three months demonstrated resolution of the fistula (Fig. 3CD). One week later, there was a remarkable improvement (Fig. 4) with no diplopia complaints.

DISCUSSION

Carotid cavernous fistulas are rare and difficult to diagnose. In both direct and indirect CCFs, anterior and inferior drainages are the most common. CCFs that drain anteriorly usually produce visual symptoms and signs. Direct CCFs tend to have a more severe, acute onset presentation, while indirect CCFs are associated with a more gradual onset and chronic course with milder symptoms. This may lead to a misdiagnosis of eye surface or unspecific orbital diseases. A diagnostic clue is given by the arterialization of scleral and episcleral veins, resulting in the elevation of the episcleral venous pressure and consequently the IOP. Ocular motility impairment can be present in about 60% of

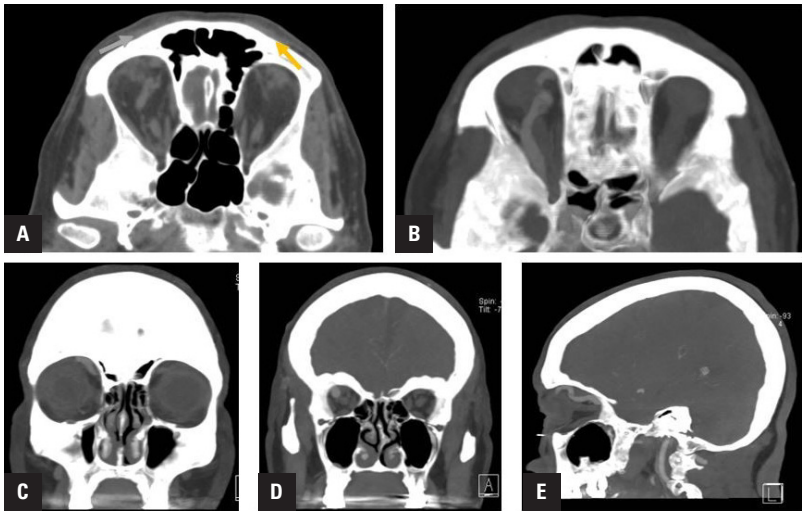

FIGURE 2. Non-contrast computed tomography (CT) on admission (**A**) showed right superior ophthalmic vein engorgement (green arrow) compared to the contralateral one (purple arrow). CT angiography (**B–E**) showed opacification of the right superior ophthalmic vein, suggesting arterialization of this structure, a characteristic often seen in arteriovenous fistulas

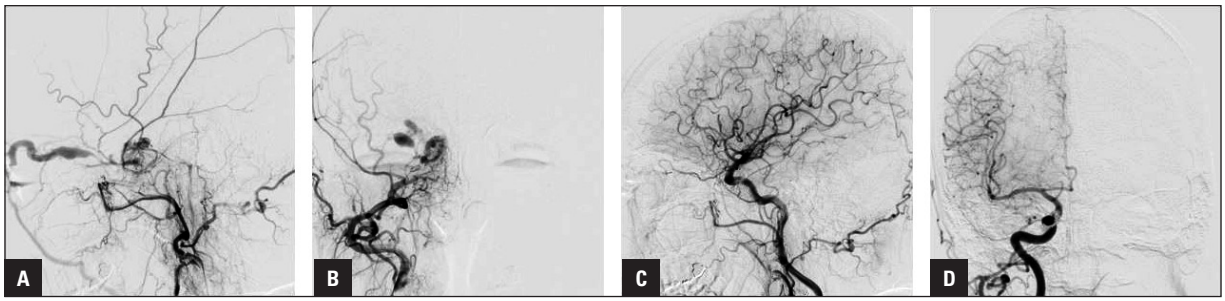

FIGURE 3. Digital subtraction angiography before (**A** — in lateral view, **B** — in anteroposterior view) showing dural arteriovenous fistula with concomitant enlargement of the right superior ophthalmic vein, and after (**C** — a in lateral view, **D** — in anteroposterior view) endovascular treatment with complete resolution of abnormal imaging findings

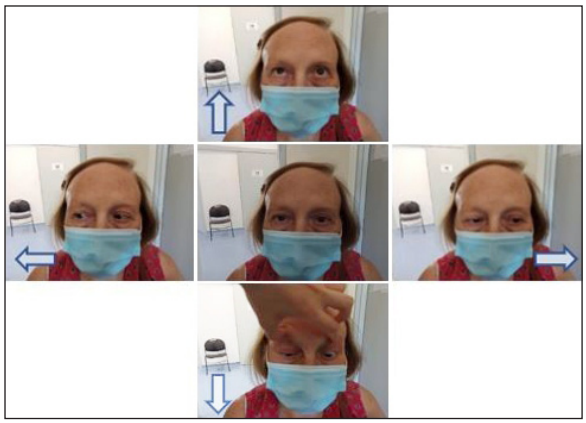

FIGURE 4. One week after endovascular treatment

patients and may be due to nerve compression, vascular steal, or thrombosis [6–8].

Clinical signs in our case series match those described in the literature, including proptosis, red eye arterialization of the conjunctival and episcleral vessels, chemosis, neurogenic strabismus most commonly due to a sixth nerve palsy, orbital congestion, ocular bruit, increased IOP, stasis retinopathy, central retinal vein occlusion, and optic neuropathy [2, 9–11].

When a CCF is suspected, a combination of exams can be performed, such as CT scanning, CT angiography, MRI, MR angiography, orbital ultrasonography, transorbital, and transcranial color Doppler imaging. However, the gold standard remains a digital subtraction angiography [6, 12], and it was performed in all our patients.

Most indirect CFFs close spontaneously, and often all clinical manifestations will resolve without vascular intervention. Several different endovascular treatment options for direct CCFs are currently available. The method chosen in a given patient depends on the anatomy of the fistula and the doctor or institutional preference. The goal of treatment in direct CFFs is to occlude the site of communication between the ICA and the cavernous sinus while preserving the patency of the ICA. The reported cure rate rounds 80% being much lower in post-traumatic CCFs [12].

CONCLUSION

Carotid cavernous fistulas presentation can be varied and nonspecific; however, patients commonly present with ophthalmological manifestations. A careful clinical history collection and ophthalmological examination allied to a high index of suspicion are essential to an accurate diagnosis.

The group of signs/symptoms identified in our study matches the clinical picture described in the literature, which ophthalmologists should be aware of, as they may be the first clinician examining these patients. Close collaboration between Ophthalmology and Neuroradiology is required for an early diagnosis and appropriate management to prevent vision loss and life-threatening complications.

Conflict of interest

The authors declare no conflicts of interest in association with the present paper.

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