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SPONTANEOUS HIGH FLOW CAROTID-CAVERNOUS SINUS FISTULA CONFIRMED FROM DIGITAL SUBTRACTION ANGIOGRAPHY: A VERY RARE CASE

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ABSTRACT

Background: Carotid cavernous fistula (CCFs) is characterized by an aberrant channel linking the carotid artery directly to the cavernous sinus. Most of the CCFs are classified as traumatic CCFs, which is occurred following trauma. Meanwhile spontaneous CCFs, which is occurred spontaneously, are very rare. Spontaneous CCFs typically occur in elderly women and are usually classified as low-flow, indirect fistulas of Barrow type D. This study aimed to present a rare case of a spontaneous high-flow CCFs classified as Barrow type A, due to its uncommon occurrence. **Case presentation:** A 56-years-old woman came fully alert presented with swelling and redness on the right eye since 1-month prior admission. Palpebral edema, episcleral injection, conjunctival chemosis, proptosis, and bruit were found from her right eye. CT-angiography found engorgement on the right cavernous sinus with superior ophthalmic vein (SOV) dilatation. The RICA DSA revealed that the contrast filled the cavernous sinus, SOV, facial vein, and internal jugular vein on the early artery phase, which indicates a high flow CCFs at the C4 with Barrow classification type A. The patient was planned for fistula embolization with coil. **Conclusion:** Spontaneous high flow CCFs is an extremely rare condition. Accurate history taking and thorough physical examination are essential to identify the classic triad of CCFs symptoms: proptosis, chemosis, and ocular bruit. The definitive diagnostic tool is a cerebral angiogram, which can reveal the presence of a fistula by demonstrating contrast filling of the cavernous sinus.

INTRODUCTION

Carotid cavernous fistula (CCFs) is characterized by an aberrant channel linking the carotid artery directly to the cavernous sinus.[1] The clinical presentation of a CCFs varies depending on which critical neural and vascular components within the cavernous sinus are affected. These include cranial nerves III (oculomotor), IV (trochlear), V1 (ophthalmic branch of the trigeminal nerve), V2 (maxillary branch), and VI (abducens). CCFs can be categorized according to their hemodynamic characteristics, underlying cause, or the anatomical configuration of the shunt. Treatment decisions are influenced by the fistula's flow rate, the venous drainage pattern, and the progression of the patient's symptoms.[2]

Etiologically, CCFs are classified as traumatic CCFs if occurred following trauma, and spontaneous CCFs if occurred spontaneously.[2] Traumatic events, which includes penetrating or sharp-force injuries, basilar skull fractures, or medical procedures, are responsible for approximately 75% of all CCFs cases. These cases typically occur in young men and are usually characterized as direct high-flow fistulas.[3] Spontaneous CCFs are uncommon and typically arise from ruptured aneurysms or inherited connective tissue disorders that increase susceptibility to vascular damage, such as fibromuscular dysplasia or Ehlers-Danlos syndrome. They are most frequently observed

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in elderly women and are usually classified as Barrow type D, indirect low-flow fistulas.[1,3]

Spontaneous CCFs of the Barrow Type A, which is characterized by direct, high-flow connections between the internal carotid artery (ICA) and the cavernous sinus, are exceedingly rare. The majority of Type A CCFs are associated with trauma, such as head injuries, and spontaneous occurrences are uncommon. In a study analyzing 16 cases of spontaneous CCFs, only one was identified as Barrow Type A, while the remaining cases were classified as Types B, C, or D, which are indirect fistulas.[4] Furthermore, recent study found that spontaneous CCFs of Barrow Type A are exceedingly rare, as all spontaneous cases in the analyzed cohort were classified as Types B, C, or D, with Type A exclusively associated with traumatic origin.[5] This suggests that spontaneous direct (Type A) CCFs constitute a small fraction of all spontaneous CCF cases.

Since 2023, our hospital has started routinely to perform brain DSA examinations, and we recently identified our first case of a spontaneous high-flow CCFs classified as Barrow Type A. Given its extremely rare occurrence, this study aims to report and highlight this unusual presentation.

CASE PRESENTATION

A 56-years-old woman came fully alert presented with swelling and redness on the right eye since 1-month prior admission. The complaint was started with redness on her right eye, and then followed with swelling and protruded in one week. The chief complaint also accompanied by hissing sound that can be heard by her right ear. The complaint was not accompanied by pain on her right eye, blurry vision, and headache. The patient had no history of trauma, seizure, vomit, and fever. The patient had history of chronic vein insufficiency on her right leg 2-months prior admission. Other chronic disease such as hypertension and diabetes mellitus was denied.

Figure 1. The patient presented with palpebral edema (star) and episcleral injection with chemosis (arrow) on her right eye, while her left eye was within normal limit. Abnormal forward displacement or bulging of the right eye compared to the left eye was found which suggest right eye proptosis. Ocular bruit was found from the auscultation of the right eye.

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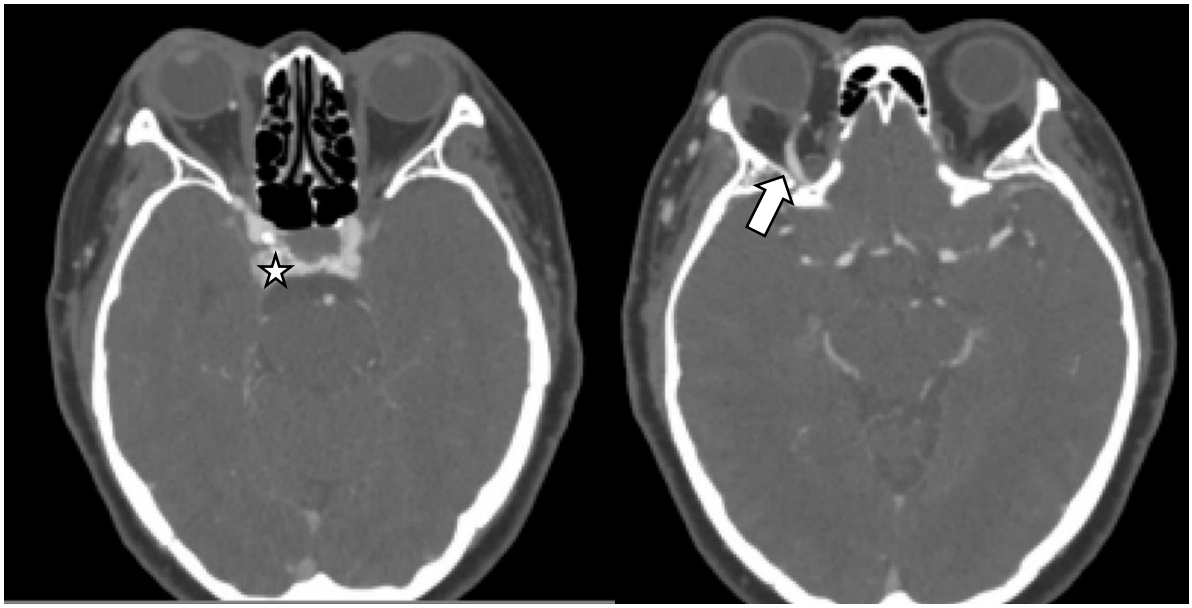

Figure 2. CT-angiography revealed engorgement on the right cavernous sinus (star) and superior ophthalmic vein (SOV) dilatation (arrow) that suggest carotid-cavernous fistula (CCF).

From the physical examination the patient had good general condition and stable vital sign. Palpebral edema, episcleral injection, conjunctival chemosis, proptosis, and bruit were found from her right eye, while her left eye was within normal limit (Figure 1). From the neurologic status, the patient was fully alert with GCS score 15 and had round equal pupil with diameter 3 mm on both eyes. Her visual acuity was 6/60 on both eyes. There was no limitation in eye movement on both eyes. The patient already had a CT-angiography prior to admission to our hospital which found engorgement on the right cavernous sinus with superior ophthalmic vein (SOV) dilatation that suggest carotid-cavernous fistula (CCF), slight right bulbus oculi proptosis, without brain parenchymal lesion (Figure 2).

The patient was assessed with right ocular proptosis suspected due to CCF. We performed diagnostic digital subtraction angiography (DSA) to confirm the diagnosis. The patient was supine positioned with local anesthesia on the right femoral region. DSA was accessed through the right femoral artery with 5 French femoral sheath. A 5 French diagnostic catheter and a 0.035" guide wire were inserted through the femoral sheath until the right internal carotid artery (RICA) was reached. Contrast was injected through the diagnostic catheter and images were captured radiographically to show the vascularization.

The RICA DSA revealed that the contrast filled the cavernous sinus, SOV, facial vein, and internal jugular vein on the early artery phase (Figure 3), which indicates a high flow CCF at the C4 with Barrow classification type A. The contrast also filled the right anterior cerebral artery (ACA) and middle cerebral artery (MCA) until its distal branches and drained through the veins. From the 3D imaging it was found the fistula diameter was 2.2 mm. There were no other pathologies found from the other artery branches, such as right external carotid artery (RECA), left common carotid artery (LCCA), and both vertebral artery (VA). The diagnosis of right CCF Barrow classification type A were confirmed and the patient was planned for fistula embolization with coil.

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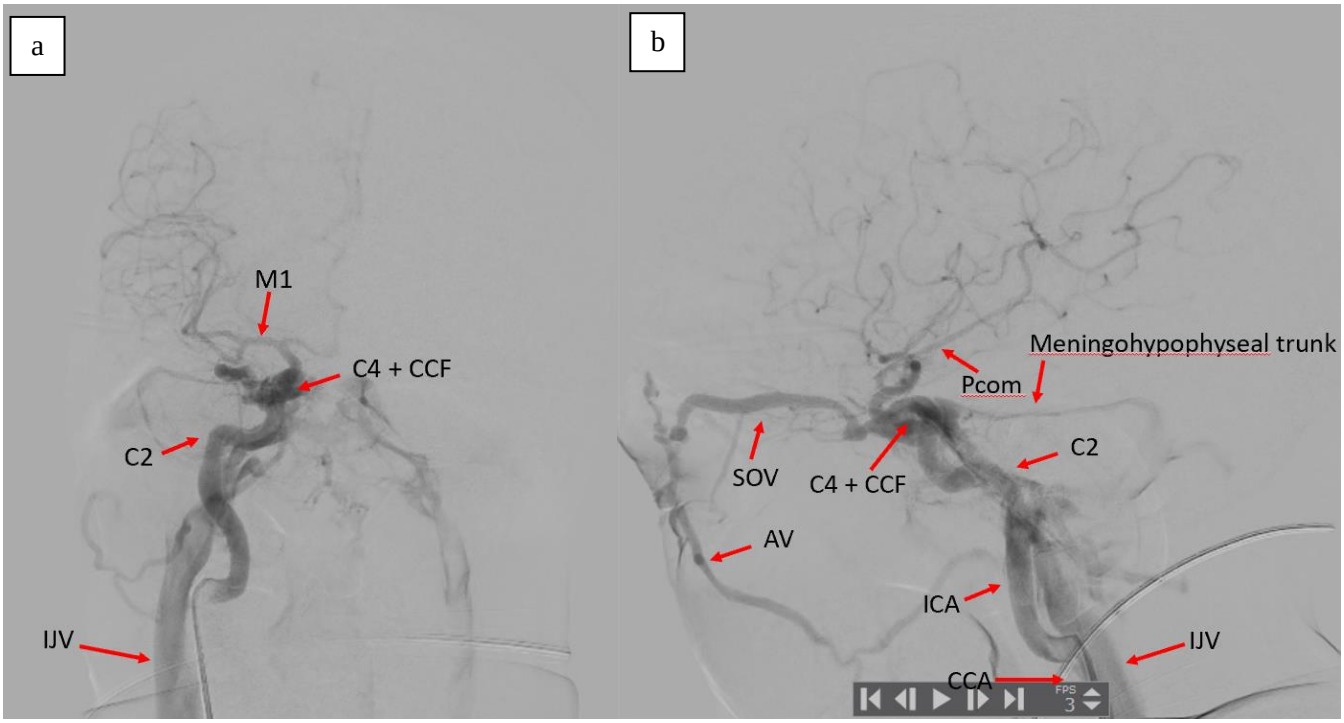

Figure 3. DSA RICA anteroposterior projection (a) and lateral projection (b) revealed the contrast filled the cavernous sinus, SOV, facial vein, and internal jugular vein on the early artery phase, which indicates a high flow CCF at the C4 with Barrow classification type A. IJV – internal jugular vein; CCA – common carotid artery; ICA – internal carotid artery; C2 – ICA pars petrous; C4 – ICA pars cavernous; M1 – middle cerebral artery; AV – angular vein

DISCUSSION

Carotid cavernous fistula (CCFs) is characterized by an aberrant channel linking the carotid artery directly to the cavernous sinus.[1] Taking a detailed history regarding symptom onset is crucial, as it can help identify the underlying cause of the CCFs. While the classic triad of CCFs such as proptosis, chemosis, and ocular bruit is frequently observed, other symptoms like orbital discomfort, visual impairment, and cranial nerve dysfunction may also occur.[6, 7] The patient presented in this case also exhibited the classic triad of CCFs (proptosis, chemosis, and ocular bruit) on her right eye, making the initial suspicion of a right-sided CCFs a reasonable diagnosis. Moreover, the patient presented in this case also had history of chronic vein insufficiency (CVI) on her right leg. Although there is still no direct or well-established association between CVI and spontaneous CCFs in literature, both conditions can be linked through shared vascular risk factors and underlying systemic conditions such as Ehlers-Danlos syndrome and fibromuscular dysplasia which can predispose individuals to vascular abnormalities, potentially contributing to both CVI and spontaneous CCFs.

The choice of diagnostic tests and imaging for CCFs is guided by the patient's symptoms and whether they present in an emergency or outpatient setting. Tonometry or pneumotonometry can reveal a higher ocular pulse amplitude on the side affected by the fistula compared to the opposite eye.[3, 8] B-scan ultrasound or color Doppler imaging can reveal signs of orbital congestion or dilated superior ophthalmic vein (SOV). Patients suspected of having a CCFs are typically evaluated with noninvasive imaging techniques such as computed tomography (CT)-scan or magnetic resonance imaging (MRI), which may demonstrate a dilated SOV, orbital congestion, or enlarged extraocular muscles. Additionally, CT-angiography or magnetic resonance angiography (MRA) can be employed, as both are highly sensitive in identifying CCFs associated with visual disturbances.[1] The patient reported in this case also had a CT-angiography prior to admission which revealed engorgement on the right cavernous sinus

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with SOV dilatation that suggest CCFs. Mild proptosis of the right bulbus oculi was also noted, with no abnormalities in the brain parenchyma. Thus, diagnosis of suspected right CCFs presented in this case was also appropriate based on the clinical findings and additional CT-angiography imaging.

DSA is considered the gold standard for diagnosing a CCFs, as it can visualize the filling of the cavernous sinus through the fistula, the venous drainage pattern, and any reflux into cortical veins after contrast injection into the common carotid artery (CCA), internal carotid artery (ICA), or external carotid artery (ECA).[3] The patient in this case also underwent DSA to confirm the diagnosis of CCFs. The RICA DSA of the patient revealed that the contrast filled the cavernous sinus, SOV, facial vein, and internal jugular vein on the early artery phase, which indicates a high flow CCFs at the C4 with Barrow classification type A. Thus, it was confirmed that the patient had right CCFs Barrow classification type A from the DSA.

Management strategies for CCFs vary based on the fistula's flow rate, with the primary aim being complete closure of the fistula while maintaining normal blood flow through the ICA. For low-flow fistulas, compression therapy is a recommended noninvasive approach. This involves applying pressure to the cervical carotid artery several times daily over a period of 4-6 weeks to encourage spontaneous thrombosis of the fistula. The compression is typically performed using the opposite hand, as any resulting cortical ischemia would naturally cause the hand to release pressure. The main advantage of compression therapy is because it is a simple, low-cost intervention that can be performed by patients themselves, reducing the need for surgical or endovascular procedures.[9] When appropriately selected, patients often tolerate compression therapy well, with minimal side effects. However, successful closure using this conservative method occurs in only about 30% of cases.[10] Other main disadvantage of compression therapy is that it is generally ineffective for direct, high-flow fistulas, which typically require endovascular intervention.[9] This is the reason compression therapy is not appropriate for the present case.

Although surgical intervention is the most invasive treatment option, it can provide a definitive solution. Surgical approaches such as suturing or clipping the fistula, packing the cavernous sinus, or ligating the ICA have reported success rates ranging from 31%-79%.[1] The advantages of surgical intervention are able to get direct access to the fistula and become the definitive treatment when endovascular approaches fail.[3, 11] The disadvantages of surgical intervention are invasive with higher risk of morbidity, longer recovery time, and technically challenging.[3] Radiosurgery is another viable option, particularly for low-flow indirect fistulas, achieving obliteration rates between 75% -100%. The main advantage is its non-invasive which no incisions or catheterizations are required, making it safer for elderly or high-risk patients. It is also useful for inaccessible lesions.[12, 13] However, due to its delayed effect which often taking months to years for full closure, radiosurgery is not suitable for acute or emergency cases.[13]

Endovascular surgery is the primary treatment approach for CCFs, with a success rate exceeding 80%.[14-17] The advantages of endovascular surgery in CCFs includes high success rates, minimally invasive approach, preservation of parent artery, and versatility in access routes.[18-21] One of the main disadvantage is require prolonged antiplatelet therapy to prevent in-stent thrombosis, which may not be suitable for all patients.[18] For direct, high-flow CCFs, a trans-arterial approach is typically preferred. Once the ICA is accessed, the fistula can be treated using coil embolization or liquid embolic agents. Alternative methods include placing a covered stent graft in the ICA or performing arterial sacrifice via endovascular techniques.[3] In this presented case, the patient was also scheduled for coil embolization to treat the CCFs, making the proposed management appropriate.

For indirect CCFs, a transvenous approach is generally favored due to the elevated risk of embolic stroke when accessing arterial feeders via a trans-arterial route. The cavernous sinus is commonly reached through the inferior petrosal sinus or via the facial vein leading to the SOV.[3, 7] In more complex cases, such as those involving venous thrombosis or highly tortuous vessels, direct cannulation of the superior ophthalmic vein after surgical exposure may be necessary to access the cavernous sinus.[22]

CONCLUSION

Spontaneous high-flow CCFs are exceptionally rare. Thorough history-taking and physical

examination are essential to identify the classic triad of symptoms: proptosis, chemosis, and ocular bruit. Imaging studies such as CT angiography can help support the diagnosis by revealing orbital congestion, SOV dilation, or enlarged extraocular muscles signs. The definitive diagnostic tool remains cerebral angiography, which allows visualization of the fistula and contrast filling of the cavernous sinus. Endovascular therapy is preferred as the first-line treatment, offering a high success rate.

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DECLARATION OF PATIENT CONSENT

The authors certify that all appropriate patient consent forms have been obtained.

FINANCIAL DISCLOSURE

The authors declare that no financial support was received from any organization for the submitted work.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

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