

Case Report

Carotid Cavernous Sinus Fistula with Contralateral Feed Not Diagnosed by Virtual Assessment or by Non-Invasive Vascular Imaging

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Keywords

Carotid cavernous fistula · Dural fistula · Digital subtraction angiography · Contralateral feed

Abstract

A 67-year-old woman had delayed initial diagnosis of her right low flow carotid cavernous fistula (CCF) during the coronavirus disease (COVID-19) pandemic due to difficulty detecting ocular signs via online virtual examinations. Her right eye conjunctival erythema and proptosis with medial rectus enlargement on computed tomography scan was initially misdiagnosed as euthyroid thyroid-associated orbitopathy without lid retraction. She developed vision loss, and increasing episcleral venous congestion and CCF was suspected. Computed tomographic angiography did not show an obvious fistula. Digital subtraction angiography revealed the right-sided low flow CCF, which was fed from vessels from the contralateral side.

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Introduction

Low flow carotid cavernous fistulas (CCF), also known as dural CCF, are predominantly spontaneous, anomalous communications between the arteries and veins within the cavernous sinus or their branches. It is not uncommon for low flow CCF to initially be misdiagnosed. Non-invasive angiography may not disclose the CCF, and digital subtraction angiography may be required.



Fig. 1. There are prominent episcleral vessels in the right eye that extend to the limbus 360° circumferentially. There was no upper or lower lid retraction, and the lids are being retracted manually.

Case Presentation

This 67-year-old woman had a 1-year history of progressive right proptosis with conjunctival injection and a left frontal headache. Due to coronavirus disease (COVID-19), several assessments were conducted online. Her proptosis and conjunctival injection were initially diagnosed as thyroid-associated orbitopathy (TAO). She was biochemically euthyroid and a nonsmoker. Other ocular history included mild hyperopia with narrow angles, treated with bilateral laser iridotomies.

After several months, she was referred to our orbital service due to vision loss in the right eye, persistent conjunctival erythema, and diplopia on eccentric gaze. On direct questioning, she endorsed an intracranial bruit. There was no history of hypertension or head trauma.

Best-corrected acuities were 20/140 in the right eye and 20/40 in the left eye. She could not see colour plates with the right eye but read them fully with the left eye. Pupils were equal, but there was a grade 2/4 right relative afferent pupillary defect. There was no lid retraction. There was 2-mm relative right proptosis with 25% loss of right abduction. There were prominent dilated 360° episcleral veins in the right eye that extended to the limbus (shown in Fig. 1). The intraocular pressures were 20 in the right eye and 13 in the left eye with the suggestion of pulsatility in the tonometry mires in the right eye. There was mild right retinal venous tortuosity and subtle disc edema. On optical coherence tomography, there was an average retinal nerve fibre layer height of 153 microns in the right optic nerve and 113 microns on the left. The cup-to-disc ratio was 0.4 on the right and 0.6 on the left.

CT scan and CT angiogram (CTA) showed an enlarged right medial rectus muscle with mild proptosis. The right superior orbital vein was only slightly prominent. There was query subtle increased vascularity at the right cavernous region around the internal carotid artery (shown in Fig. 2). The neuroradiologic differential diagnosis included TAO, orbital myositis, and IgG4-related disease. The findings were not sufficiently conclusive for low flow CCF also known as dural CCF. Because of a high clinical suspicion of dural CCF, intravenous digital subtraction angiogram (DSA) was performed. This showed a dural CCF at the level of the right cavernous sinus, mainly supplied by the left external carotid artery and, to a lesser extent, by the left internal carotid artery meningeal branch of the left meningohypophyseal trunk. The actual fistulous connection appeared to be intercavernous with shunting into the right cavernous sinus and thence forward into the right superior orbital

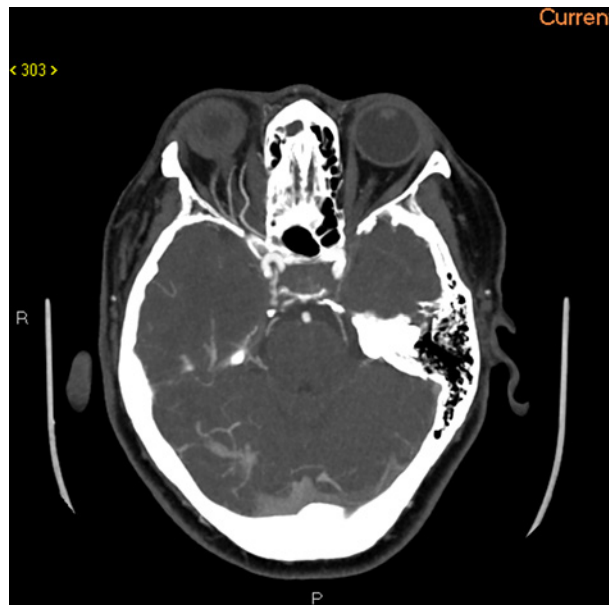


Fig. 2. CTA showed an enlarged right medial rectus muscle and slight prominence of the right superior orbital vein. There was subtle increased vascularity at the right cavernous region around the internal carotid artery. CTA, CT angiogram.

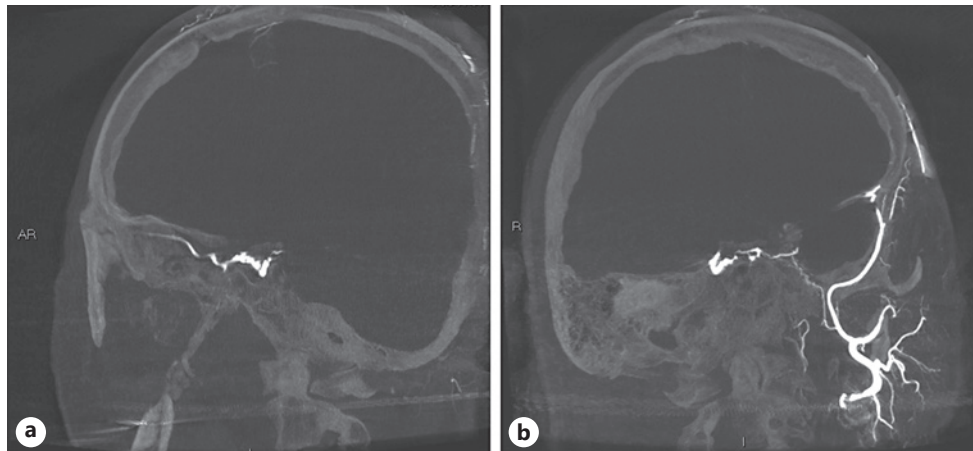


Fig. 3. A DSA showed a dural arteriovenous fistula of the right cavernous sinus with no contribution from the right internal or external carotid arteries. **B** The right dural arteriovenous fistula is mainly supplied by the left external carotid artery and, to a lesser extent, by the left internal carotid artery meningeal branch of the left meningohypophyseal trunk. The actual fistulous connection appeared to be intercavernous with shunting into the right cavernous sinus and thence forward into the right superior orbital vein. DSA, digital subtraction angiogram.

vein. There was no contribution by the right internal or external carotid arteries to the dural CCF. (See Fig. 3) As indicated earlier, there was forward drainage into the right superior ophthalmic vein and no evidence of cortical venous reflux. As many low flow dural arteriovenous fistulas/CCF spontaneously close after DSA, the patient was observed with gentle intermittent orbital massage. At 5-week follow-up, there was a marked decrease in the episcleral venous congestion, her vision improved to 20/60 in the affected eye with an intraocular pressure of 13, and clinical regression of the disc edema with an optical coherence tomography average nerve fibre layer 129 microns. Her right eye abduction improved, and she had much less diplopia on right gaze.

Discussion

CCFs can be direct or indirect (dural) and high flow or low flow. Direct CCF arises from abnormal communication between the internal carotid artery and the cavernous sinus, usually after trauma. Indirect CCF is from abnormal communications between the dural branches of the internal or external carotid arteries with the cavernous sinus. Most low flow CCFs are spontaneous and occur in older women [1]. Dural arteriovenous fistula can cause vision loss, abducens paresis and be potentially life-threatening if there is cortical venous reflux [2]. Treatment options for CCF include observation, intermittent massage of the ipsilateral internal carotid artery or orbit, endovascular intervention, or stereotactic radiosurgery. Up to 70% of dural CCF may close spontaneously and can be managed conservatively, or with gentle, intermittent massage of the ipsilateral carotid artery or superior ophthalmic vein [3]. If intervention is required for dural CCF, endovascular procedures are usually performed.

This patient's case is instructive for 5 reasons. (i) Although CCF can masquerade as TAO [4], upper eyelid retraction occurs in at least 75% of patients with TAO [5]. Eight percent of patients with TAO are euthyroid [6]. The lack of lid retraction and the euthyroidism should widen the clinical differential diagnosis beyond TAO to include dural CCF. In one series, 39% of patients with dural CCF were initially misdiagnosed as having other ocular diseases [7], including drop medicamentosa and central venous occlusion. (ii) Although online eye examinations are more frequent during COVID [8], virtual assessments may fail to distinguish TAO from dural CCF. Without marked magnification and eversion of the lids, ocular signs such as 360° episcleral venous distension extending to the limbus, a characteristic of dural CCF, may not be discernible with virtual examinations. (iii) CTA and MR angiography may not disclose the dural CCF. The sensitivity of DSA versus CTA versus MR angiography to detect the fistulous tracts of CCF was 94%, 87%, and 80%, respectively [9]. (iv) The site of the fistula was contralateral to the eye with venous congestion. In one series of 27 patients with CCF, 2 patients had a unilateral orbital presentation contralateral to the fistula, and several patients had bilateral orbital involvement with a unilateral CCF [10]. (v) Some low flow CCF may improve after DSA or gentle orbital or carotid massage.

In summary, online examinations of the eye may not show sufficient detail and may cause delay in diagnosis. In patients with apparent TAO, CCF should be suspected if the episcleral vessels extend to the limbus and if there is no lid retraction. Digital subtraction angiography remains the gold standard to confirm CCF and may occasionally show that the CCF is fed from the contralateral side. Low flow fistulas can close spontaneously.

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Statement of Ethics

The research was conducted in accordance with the Declaration of Helsinki. The patient provided written informed consent to publish her case and for the publication of images. Study approval statement: the case report was deemed exempt by the Michael Garron Hospital Research Ethics Board.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Each of the authors performed all 4 of the ICMJE criteria. Edsel Ing contributed to the conception, design, data acquisition, data analysis/interpretation, drafting and critical revision of the article, and final approval and is accountable for all aspects of the work. Felix Tyndel contributed to the conception, design, data acquisition, data analysis/interpretation, drafting and critical revision of the article, and final approval and is accountable for all aspects of the work. Joyce Tang contributed to the data acquisition, data analysis/interpretation, drafting and critical revision of the article, and final approval and is accountable for all aspects of the work. Tom Marotta contributed to the data acquisition, data analysis/interpretation, drafting and critical revision of the article, and final approval and is accountable for all aspects of the work.

Data Availability Statement

There is no database associated with this case report. The details of the patient's history and radiology are documented on the patient's private office and hospital medical charts.

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