Spontaneous near-complete resolution of direct carotid-cavernous fistula resulting from ruptured cavernous internal carotid artery aneurysm

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DESCRIPTION

This 57-year-old woman with a history of migraine, hypertension and pulmonary embolism presented to an outside facility with spontaneous left ptosis, ophthalmoplaegia and headache for 4 days. Her symptoms began abruptly with retro-orbital pain, nausea, vomiting and left eye proptosis. There was no antecedent trauma. MRI was negative for acute pathology. A lumbar puncture opening pressure was 50 cm H_2O, prompting transfer to our facility for presumed idiopathic intracranial hypertension. On arrival, she had proptosis and ptosis of the left eye with cranial nerve 3, 4 and 6 palsies. Facial sensation was normal. Visual acuity was 20/20 and 20/40 in the right and left eyes, respectively. Intraocular pressure was elevated in left eye at 22 mm H_2O. There was no bruit on auscultation of the left eye. Her differential diagnosis localised to the left cavernous sinus and included carotid-cavernous fistula (CCF), cavernous sinus thrombosis, cavernous sinus syndrome and Tolosa-Hunt syndrome. A CT angiogram demonstrated bilateral cavernous sinus contrast opacification and a small anterior projecting irregularity of the proximal left cavernous internal carotid artery (ICA) concerning for cavernous aneurysm and direct CCF. Digital subtraction angiography (DSA) confirmed the diagnosis of left cavernous ICA aneurysm and direct CCF (figure 1A,B). The patient was started on dual antiplatelets and returned 2 weeks later for flow-diverter placement; however, there was near-complete spontaneous resolution of her fistulous connection on DSA with minimal shunting of contrast into the cavernous sinus (figure 1C). Given the persistence of the cavernous aneurysm, a Pipeline Flex embolization device (PED; Medtronic) was deployed within the cavernous ICA to assist with fistula obliteration (figure 2). She tolerated the procedure well without complication and reported immediate improvement in her ptosis and subjective visual acuity improvement.

CCFs are classified as either direct or indirect, depending on whether the arterial fistulous connection arises directly from the cavernous ICA

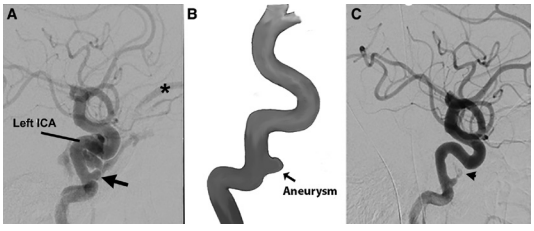

Figure 1 (A) Lateral DSA of left ICA shows direct CCF from ruptured cavernous ICA aneurysm (arrow) with engorged cavernous sinus and dilated superior ophthalmic vein (asterisk). (B) Line drawing depicting the anatomy of the left ICA and the ruptured cavernous ICA aneurysm. (C) Lateral view DSA of left ICA 2 weeks later demonstrates near-complete resolution of direct CCF with minimal shunting of contrast (arrowhead) into the cavernous sinus. CCF, carotid-cavernous fistula; DSA, digital subtraction angiography; ICA, internal carotid artery.

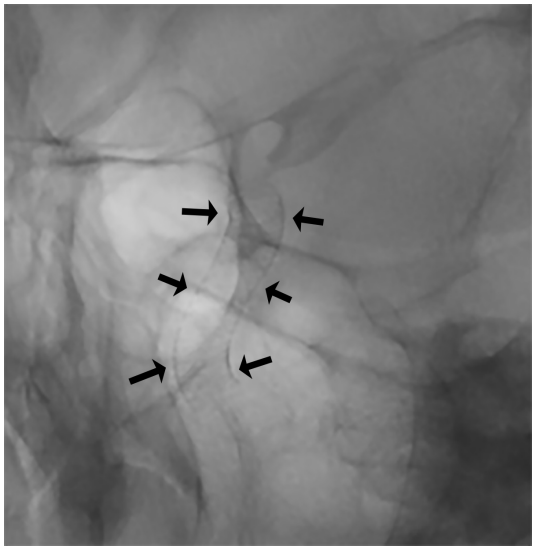

Figure 2 Left anterior oblique view after embolisation shows the PED (arrows) deployed within cavernous ICA. ICA, internal carotid artery; PED, Pipeline Flex embolization device.

Learning points

- Direct carotid-cavernous fistulas (CCFs) are high-flow connections that manifest with devastating ocular and cranial nerve symptoms.
- Spontaneous resolution of direct CCF is rare and is associated with low-flow connections, hypotension, severe ocular symptoms, carotid artery dissection or spasm and elevated intracranial pressure.
- Endovascular treatment with flow-diverting stents is a mainstay treatment that is both safe and efficacious.

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or indirectly from either intracranial or extracranial branches of the ICA or external carotid artery, respectively.[1] Direct CCFs are high-flow connections most commonly resulting from skull base trauma; although they may also arise spontaneously from rupture of an intracavernous carotid artery aneurysm as in our patient. High-flow fistulas rarely resolve without treatment, and these lesions clinically manifest with abrupt ocular and cranial nerve features including rapid vision deterioration, proptosis, retro-orbital pain, ophthalmoplaegia, ocular injection and chemosis. Spontaneous closure of direct CCFs is rare, with rates estimated to be 1.2%–4%.[2] A 2019 literature review identified 37 patients with 43 direct CCFs (bilateral fistulas in 6) that resolved spontaneously.[2] Factors associated with spontaneous resolution included low-flow fistulous connection, profound hypotensive episodes, severe ocular symptoms, carotid dissection or spasm and elevated intracranial pressure, consistent with the patient's presentation.[2] Aggressive treatment of direct CCFs is recommended, even in the setting of near-complete spontaneous resolution, because of the potential for devastating ocular effects if left untreated. Endovascular treatment has become a mainstay treatment with improved efficacy and decreased surgical morbidity. In particular, use of flow-diverting stents placed within the cavernous ICA across the fistulous connection is both safe and efficacious.[3]

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