CASE REPORT

Carotid-Cavernous Fistula Manifesting as Symptomatic Bradycardia: A Unique Presentation

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ABSTRACT

A 54-year-old female with vertigo and sarcoidosis presented to the emergency department with a 4-day history of generalised malaise, headache, fever, and near syncope. She was noted to have severe bradycardia and was admitted for possible pacemaker implantation. During the admission, the patient voiced left eye complaints, the workup of which revealed a left internal carotid aneurysm that ruptured, causing a direct carotid-cavernous fistula. The patient underwent multiple procedures to tamponade the fistula, and the bradycardia resolved, avoiding pacemaker implantation.

Keywords: Bradycardia, carotid-cavernous fistula, internal carotid aneurysm

INTRODUCTION

A carotid-cavernous fistula (CCF) is an abnormal connection between a high-pressure arterial vessel and a lower pressure venous structure within the cavernous sinus. The more common presenting symptoms of a carotid-cavernous fistula are unilateral exophthalmos (90%), conjunctival chemosis (90%), diplopia (50%), and orbital or cephalic bruit (25%). These are the result of an increase in ipsilateral orbital venous pressure.^{1,2} Presenting signs and symptoms can vary significantly, ranging from very mild complaints to complete visual loss (up to 50%).^{1–4} A CCF is typically caused by head trauma, but can occur without precipitating injury. A delay between inciting event and presentation of symptoms is common. An interval of up to 2 years has been reported in the literature.^{2,3} We report a patient with an atypical presentation of symptoms and an atraumatic history. To our knowledge, this is the first case of symptomatic bradycardia induced by a CCF.

CASE REPORT

A 54-year-old female with a medical history of vertigo and sarcoidosis presented to the emergency department complaining of generalised malaise, headache, fever, and near syncope for 4 days. Electrocardiogram (EKG) showed sinus bradycardia at 40 beats per minute with no ST segment changes. The patient was admitted to the hospital and cardiology service recommended a pacemaker to treat the bradycardia. The patient also reported left eye discomfort shortly after admission and was found to have left upper eyelid swelling.

Ophthalmology consultation revealed trace left temporal upper eyelid oedema with no induration or erythema. The patient had no visual complaints. Her near visual acuity measured J1+ and J1 in the right and left eyes, respectively, with intact colour vision. There was neither proptosis nor afferent pupillary defect. Intraocular pressures were 11 mm Hg right eye and 18 mm Hg left eye. The left lacrimal

FIGURE 1 Reconstructed axial (LEFT) and coronal (RIGHT) post-contrast CT images of the orbits reveals a dilated left superior ophthalmic vein and enlarged left cavernous sinus.

gland was nodular to palpation. There was trace conjunctival injection of the left eye. The rest of the anterior and posterior segment examinations were within normal limits. The patient was thought to have dacryoadenitis related to her diagnosis of sarcoid. Oral corticosteroids had already been initiated by the pulmonology service for the treatment of underlying pulmonary sarcoidosis. Therefore, no additional treatment was initiated at the time of the initial ophthalmology consultation.

The following day, the patient was examined and pacemaker implantation was deferred until her eye condition resolved. The left eyelid oedema had progressed to 2+, with development of diffuse erythema around the left eye. Ocular motility examination revealed –1 abduction deficit of the left eye. The eye remained painful, and now had progressed to have trace injection and dilation of the conjunctival vasculature. Computerised tomography of the orbits demonstrated minor swelling of the left lacrimal gland, aneurysmal dilation of the left cavernous sinus, and expansion of the left orbital vein (Figure 1). Neurovascular radiology was consulted. Cerebral angiography revealed a 16 × 14 mm left internal carotid artery aneurysm that had ruptured and formed a carotid-cavernous fistula (Figure 2). Balloon-assisted embolisation of the aneurysm with pipeline deployment across the neck of the aneurysm within the left internal carotid artery was performed the next morning (Figure 3).

The patient stabilised and reported improvement in symptoms over postoperative days 1 and 2. On postoperative day 3, the patient had increased left eye pain with diplopia. Ocular examination revealed left eye visual acuity of 20/50, left eye 2+ chemosis, proptosis, and palsy of cranial nerves III, IV, and VI. Intraocular pressure (IOP) of the left eye was 29 mm Hg. Interventional radiology emergently performed embolisation of the left cavernous sinus via a transvenous approach to close the venous access of the fistula.

FIGURE 2 Lateral angiogram after left internal carotid artery injection in the arterial phase demonstrates a left cavernous internal carotid artery aneurysm, left cavernous sinus, and dilated superior ophthalmic vein consistent with carotid-cavernous fistula.

On subsequent postoperative day 1, the patient reported significant improvement in left eye pain but persistent diplopia. IOP of the left eye was 14 mm Hg, and cranial nerve palsies persisted with limited improvement. Right eye visual acuity was J1 at presentation and stable throughout hospital stay. The initial systemic complaints upon presentation had largely resolved with treatment of the CCF. The patient remained bradycardic in the 40–50 beats per minute range but was asymptomatic. Mean arterial pressure ranged from 68 to 110 mm Hg during treatment course with no discernable relationship between blood pressures and interventions. The patient was discharged home on theophylline 300 mg twice a day (BID) for management of bradycardia.

FIGURE 3 (LEFT) Lateral view during initial treatment shows coils in aneurysm placed with balloon assistance. (MIDDLE) Lateral view during pipeline placement post coiling. (RIGHT) Lateral angiogram after left internal carotid artery injection in the arterial phase demonstrates residual fistula.

FIGURE 4 (LEFT) Lateral angiogram after left internal carotid artery injection in the arterial phase demonstrates after transvenous embolisation of the left cavernous sinus demonstrates a very small residual fistula. (MIDDLE) Lateral view post onyx embolisation of the residual fistula. (RIGHT) Lateral angiogram after left internal carotid artery injection in the arterial phase shows complete obliteration of the fistula.

Five days after discharge, the patient presented again to the emergency department with headache, decreased vision, and left eye pain of 1 day's duration. The patient continued to have asymptomatic bradycardia. Ophthalmology examination revealed left eye visual acuity of hand motion, non-reactive left pupil, 2+ left afferent pupillary defect, and complete left upper lid ptosis. Cranial nerve III, IV, and VI palsies persisted. Left eye IOP was 11 mm Hg. Cerebral angiogram revealed the coils and pipeline to be in place and intact as well as resolution of left superior ophthalmic vein dilation. Due to presenting symptoms, there was concern that her CCF had reopened. Repeat angiography did reveal residual CCF flow, and the patient underwent complete Onyx embolisation of the left cavernous sinus (Figure 4). Over postoperative days 1 and 2, the patient had complete resolution of most of her presenting ocular symptoms. Diminished extraocular motility, 2+ afferent pupillary defect, and diplopia persisted in the left eye. On the day of discharge, left

eye visual acuity was 20/25 and heart rate ranged from 73 to 99 beats per minute.

Three months after discharge, the patient's bradycardia had resolved, and she was no longer taking theophylline. Her left eye visual acuity was 20/20. Her ocular motility had recovered except for a trace limitation in abduction of her left eye. She denied diplopia. The patient's pupil examination was normal with no residual afferent pupillary defect.

COMMENTS

Carotid-cavernous fistulas can be classified as either direct or indirect. Direct is the most common type, accounting for up to 80% of cases.⁴ A direct CCF involves the cavernous portion of the internal carotid artery (ICA), whereas an indirect CCF involves branches of the ICA or the external carotid artery. Roughly 20% of direct cases are atraumatic and likely result from rupture of an ICA aneurysm or an

atherosclerotic, structurally weak ICA.² In the case presented, imaging revealed this CCF to be the result of a ruptured ICA aneurysm with connection made to cavernous sinus vasculature, making this a direct, atraumatic CCF.

When working up a patient with a suspected CCF, many non-invasive image modalities are available, with the decision being guided by patient factors, available resources, and symptomatology. Computed tomography (CT), magnetic resonance imaging (MRI), CT angiography, and MR angiography are common study choices. The current gold standard for accurate diagnosis is digital subtraction angiography (DSA).^{2,4} A CT of the orbits was the initial study in our case, with the decision driven by the patient's initial ocular complaints.

Various methods are available for the management and treatment of a CCF, including conservative measures, open surgical correction, and endovascular methods. Currently, endovascular intervention is the preferred route of treatment.⁴ This is accomplished with balloons, coils, stents, or liquid embolic agents. Our case involved the use of coils placed during two separate surgeries. A pipeline stent was initially unsuccessful, requiring eventual Onyx embolisation during a third procedure to gain complete control of the fistula tract. This combination is common and effective, as the use of coils, balloons, and stents alone have limitations described in the literature.^{2,4} Onyx is an injectable liquid embolic agent used in the treatment of CCFs to obliterate the fistula tract. It is very effective in gaining control of fistula flow, but it is not without complications. Transient and permanent cranial nerve (CN) deficits have been observed post Onyx therapy. Proposed mechanisms of these deficits include infarction of nerve fibers or nerve compression due to post-embolisation swelling.⁵ However, Zanaty *et al.* reported an increased risk of cranial nerve deficits post coiling when compared with liquid embolic agents.⁴ In our case, CN deficits developed after coiling but prior to the use of Onyx.

This case is reported due to the unique presentation of the CCF. The relationship between the CCF and the bradycardia is not fully understood. One plausible explanation could be related to a sustained

oculocardiac reflex. This well-described reflex involves transmission of stimuli along an afferent pathway involving the ophthalmic branch of the trigeminal nerve and an efferent pathway involving increased vagal stimulation of the myocardium, resulting in bradycardia. This reflex has been described related to ophthalmic surgical procedures or ophthalmic trauma.^{6,7} We postulate that the relationship of the ophthalmic branch of the trigeminal nerve to the cavernous sinus was the cause of bradycardia. To our knowledge, this is the first time that a CCF initially presented with symptomatic bradycardia. Ocular complaints after the patient had been admitted prompted a workup that revealed the CCF. The planned implantation of a pacemaker was unnecessary after the CCF was discovered and definitively treated.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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