

2014

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Recommended Citation

Eswar A, Pomeranz HD, Vishnubhakat SM, Oller-Cramsie M. Carotid-cavernous fistula as a mimicker of myasthenia gravis. . 2014 Jan 01; 5():Article 1302 [p.]. Available from: <https://academicworks.medicine.hofstra.edu/articles/1302>. Free full text article.

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Surg Neurol Int. 2014; 5: 140.

PMCID: PMC4192903

Published online 2014 Sep 26. doi: [10.4103/2152-7806.141888](https://doi.org/10.4103/2152-7806.141888)

Carotid-cavernous fistula as a mimicker of myasthenia gravis

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Received 2014 Jun 26; Accepted 2014 Aug 7.

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Abstract

Background:

A carotid-cavernous fistula (CCF) is an abnormal communication between the carotid arterial system and the cavernous sinus. Common symptoms of CCFs include proptosis and ophthalmoplegia, but fluctuating diplopia and presence of ptosis are not typical.

Case Description:

We present an unusual case of CCF with fluctuating binocular diplopia and ptosis, mimicking myasthenia gravis. Electrodiagnostic testing, which included repetitive nerve stimulation and single-fiber electromyography, was normal. Magnetic resonance imaging of the brain and orbits was initially normal, but later magnetic resonance angiography revealed enlargement of the left superior ophthalmic vein along with a left CCF. Patient underwent a successful left cavernous sinus embolization.

Conclusion:

Fluctuating ophthalmic symptoms are a typical presentation of myasthenia gravis; however, there may be an association of these symptoms with a CCF. Repetitive nerve stimulation and single-fiber electromyography played a key role in diagnosis of this case, as the normal result led to further investigations revealing a CCF.

Keywords: Carotid-cavernous fistula, fluctuating ophthalmic symptoms, myasthenia gravis, repetitive nerve stimulation, single-fiber electromyography

INTRODUCTION

A carotid-cavernous fistula (CCF) is an abnormal communication between the carotid arterial system and the cavernous sinus. Common symptoms of CCFs are conjunctival injection, proptosis, ophthalmoplegia, and decreased visual acuity.[7] However, fluctuating diplopia and ptosis are not commonly seen and are more characteristic of neuromuscular junction diseases such as myasthenia gravis. We present an unusual case of CCF mimicking myasthenia gravis.

CASE REPORT

A 69-year-old man with past medical history of diverticulitis and irritable bowel syndrome presented with 1 month history of binocular vertical diplopia that was worse in right gaze and downward gaze. He also noted mild ptosis of his left upper lid, some increased tearing in the left eye, and discomfort behind the left eye, which he attributed to preexisting seasonal allergies. On examination, visual acuity with correction was 20/30 in the right eye and 20/20 in the left eye. Pupils were normal and visual fields were full. Ocular motility examination showed grossly full ductions and versions. Maddox rod testing revealed a small left hyperdeviation that was the same in all directions of gaze except in downward gaze, where the left hyperdeviation increased. He was able to fuse with two prism-diopters base-down prism in front of the left eye. He had a mild left upper lid ptosis, which did not appear fatigable with prolonged upgaze. Cogan's lid twitch was absent. Globes were normal to retropulsion. Orbicularis function, anterior segment examination, and fundoscopic examination were normal bilaterally. Intraocular pressure was 18 mm Hg in both eyes. The patient was provided with prism lenses which led to slight resolution of symptoms over 2 weeks. Acetylcholine receptor antibodies were undetectable and thyroid stimulating hormone levels were normal. Patient returned a week later with the complaint that diplopia returned and was now almost constant. The diplopia was more horizontal than vertical. On examination, he had an 8–10 prism-diopter esotropia, as well as a small left vertical deviation. Repeat acetylcholine receptor antibodies were negative. Patient was referred for magnetic resonance imaging which demonstrated an incidental planum sphenoidale meningioma. On follow-up 3 weeks later, the patient had improvement in double vision, with symptoms worsening toward evening. On examination, he had small left hyperdeviation. However, the esodeviation from prior examination had disappeared and he had a small exodeviation. MuSK myasthenia antibody was negative. One month later, diplopia worsened, with exam demonstrating left sixth nerve palsy. Electrodiagnostic testing revealed normal nerve conduction and electromyographic studies, as well as normal repetitive nerve stimulation and single-fiber electromyography. Magnetic resonance imaging and magnetic resonance angiography were performed at this time. Magnetic resonance angiography revealed enlargement of left superior ophthalmic vein along with a left CCF. The patient underwent a left cavernous sinus embolization by transvenous approach [Figures 1 and 2]. On 3-month follow-up, he had a left sixth nerve palsy with a large amplitude esotropia. On 1 year follow-up, he had complete resolution of sixth nerve palsy and prior symptoms.

DISCUSSION

CCFs can be classified into four types based on angiographic characteristics. Type A fistulas are high-flow direct communications between the internal carotid artery and the sinus. Types B, C, and D are of low flow. [2] Acute onset of symptoms is characteristic of direct high-flow CCF, due to high-pressure arterial blood transmission directly into the cavernous sinus and ophthalmic veins. [1] However, indirect symptoms of CCFs usually present more gradually and with milder ophthalmic signs, such as chronic red eye, elevated intraocular pressure, Proptosis, or ocular palsy. [5] Since these symptoms are mild, usually developing slowly, they are often misdiagnosed. [4] Tears of the internal carotid artery, typically due to trauma, cause direct fistulas, whereas spontaneous fistulas are typically of low flow. In our case, the patient had the latter, with no history of trauma and presence of mild and progressive symptoms.

Dural CCFs can be further classified as posteriorly and anteriorly draining, with differences in clinical presentation. CCFs draining posteriorly into the superior and inferior petrosal sinuses are typically asymptomatic, but can present with cranial neuropathies. Trigeminal neuropathy, facial nerve paresis, and ocular motor nerve paresis have been described. [5] However, CCFs draining anteriorly present with more apparent visual symptoms and signs. In mild cases, there may be eye redness, conjunctival chemosis, proptosis, or a combination of all. In more severe cases, orbital congestion, proptosis, chemosis, and dilation of the conjunctival vessels are obvious and severe. [6]

It is important to remember that patients may regard diplopia as fluctuating when noted only with specific direction of gaze. This can be resolved with careful neurological examination and repeated testing to establish reproducibility.[3] Our patient underwent a careful neuro-ophthalmologic evaluation; however, it was the worsening of the symptoms that prompted further investigation. Repetitive nerve stimulation and single-fiber electromyography played a key role in diagnosis of this case, as the normal result led to further investigations ultimately revealing a CCF. This was further supported by negative serology and imaging findings and led to appropriate treatment.

Fluctuating ophthalmic symptoms are a typical presentation of myasthenia gravis; however, there may be an association of these symptoms with a CCF. It is important to consider this when the workup for neuromuscular junction disorders is negative, in order to facilitate early diagnosis and treatment.

Footnotes

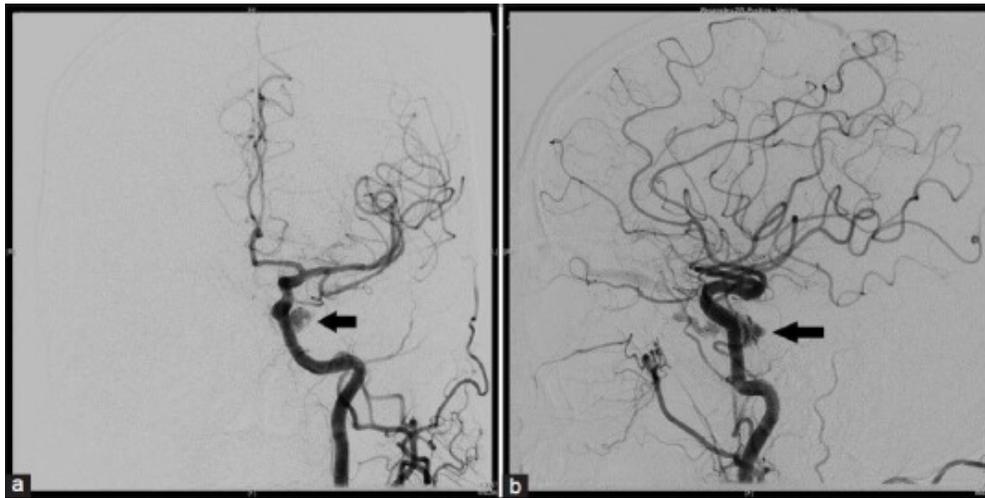
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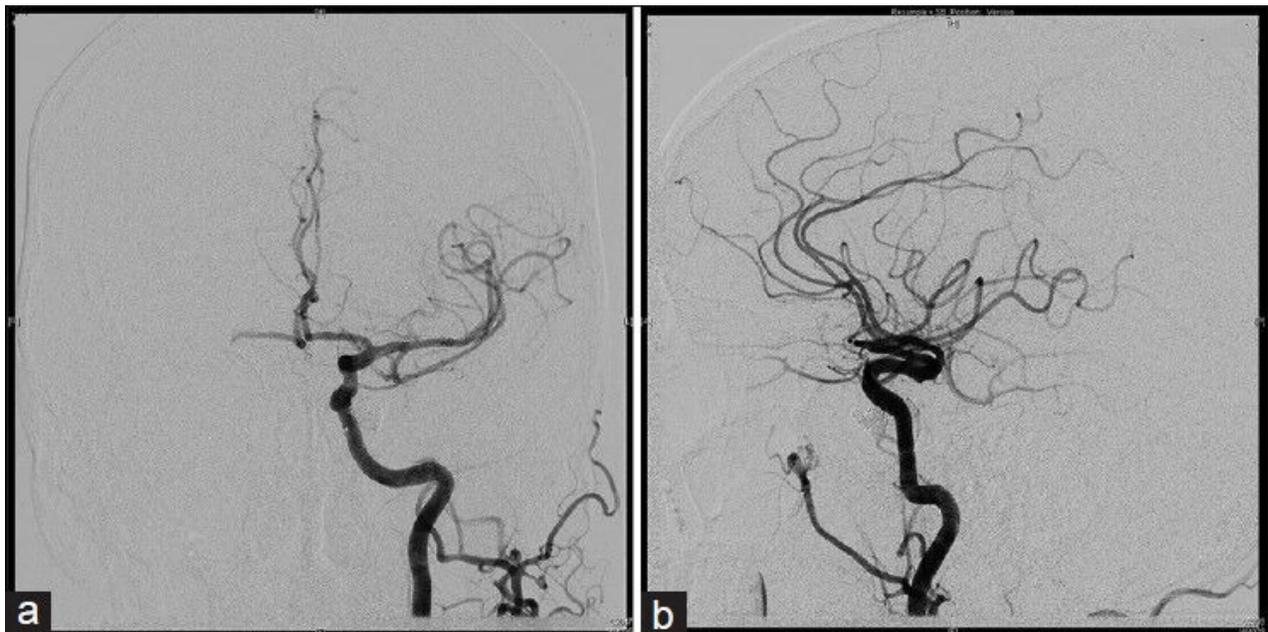
Figures and Tables

Figure 1



(a) Left carotid cavernous dural arteriovenous fistula draining into the left cavernous sinus (arrow) in AP digital subtraction angiography of the left carotid artery. (b) Left carotid cavernous dural arteriovenous fistula draining into the left cavernous sinus (arrow) in lateral digital subtraction angiography of the left carotid artery

Figure 2



(a) Arterial phase of the left internal carotid artery post embolization demonstrating complete occlusion of the left carotid cavernous dural arteriovenous fistula in AP view. (b) Arterial phase of the left internal carotid artery post embolization demonstrating complete occlusion of the left carotid cavernous dural arteriovenous fistula in lateral view

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