Grand Rounds
A 76-year-old woman with progressive right-sided proptosis, blepharoptosis, vision loss, and ophthalmoplegia

Grayson W. Armstrong, MD, MPH, **Karen Jeng-Miller, MD, MPH**, **Patrick Oellers, MD**, and **Michael K. Yoon, MD**

Author affiliations: Department of Ophthalmology, Massachusetts Eye and Ear Infirmary; Harvard Medical School, Boston, Massachusetts

History

A 76-year-old woman who had emigrated from Haiti 6 days prior presented emergently at the Massachusetts Eye and Ear for evaluation of slowly progressive right-sided proptosis and blepharoptosis, vision loss, and ophthalmoplegia, as well as seizurelike activity over a 2-year period. Early that morning, the patient awoke and experienced an episode of shortness of breath and involuntary shaking of her entire body, followed by generalized weakness and no recollection of the event. The patient had no known past medical history or history of trauma. She had never received an eye examination.

Examination

Visual acuity in the right eye was hand motions. The right pupil was nonreactive, with an afferent pupillary defect and a normal intraocular pressure. The right eye exhibited complete ptosis, complete ophthalmoplegia, and 3 mm of proptosis by exophthalmometry (Figure 1); the left eye exhibited no ptosis or ocular motility deficits. Anterior segment evaluation of the right eye demonstrated central corneal opacification, with mild thinning and diffuse neovascularization as well as a dense cataract. There was no view posteriorly. B-scan ultrasonography revealed an attached retina and vitreous debris with excavation of the optic nerve head. The left eye examination was normal, with a visual acuity of 20/30, reactive pupil, and normal intraocular pressure. Notably, the patient exhibited jaw thrust to the right while at rest and right-sided temporalis muscle wasting.

Ancillary Testing

Computed tomography (CT) of the brain with thin orbital cuts identified a right giant cavernous carotid aneurysm with osseous remodeling (Figure 2). Magnetic resonance imaging (MRI) and magnetic resonance angiogram (MRA) of the brain demonstrated a 4.9 cm partially thrombosed, peripherally calcified, saccular aneurysm arising from the paraclinoid segment of the right internal carotid artery with associated mass effect on surrounding brain parenchyma, the optic canal and superior orbital fissure, the prechiasmatic right anterior optic nerve, and the cisternal segment of the right trigeminal nerve (Figure 3). The right side of the optic chiasm was displaced superiorly. Atrophy of the ipsilateral extraocular, masticator, pterygoid, and temporalis muscles were noted (Figure 4). The left superior ophthalmic vein was dilated. Notably, there was no evidence of aneurysmal rupture. Electroencephalography was negative for epileptiform discharges.

Treatment

Neurosurgical evaluation was obtained and levetiracetam was initiated. Outpatient intervention was planned given the chronicity and presumed stability of the aneurysm. The patient underwent aneurysmal flow diversion at an outside institution without complication.

Differential Diagnosis

Our patient had a chronic and slowly progressive set of clinical signs and symptoms, including unilateral ophthalmoplegia, complete ptosis, optic neuropathy, right-sided jaw thrust and temporalis wasting, and seizurelike episodes. This collection of clinical findings localizes any neurologic lesion to the right-sided cavernous sinus and Meckel's cave. Any space-occupying lesion in this area could result in an identical presentation, although optic neuropathy from a lesion in the cavernous sinus

Figure 1. Clinical photographs of nine cardinal positions of gaze in patient with right eye exhibiting complete ophthalmoplegia in all directions of gaze. Due to complete right-sided ptosis, the upper eyelid was manually supported during photography.

and Meckel's cave would only result from a lesion extending anteriorly to the orbital apex or superiorly to the optic nerve or chiasm. Unilateral complete ophthalmoplegia can result from multiple causes. Rapid-onset of symptoms can result from infectious etiologies (mucormycosis, orbital cellulitis, cavernous sinus thrombosis), vascular etiologies (traumatic carotid cavernous fistula, retrobulbar hemorrhage, pituitary apoplexy), traumatic etiologies (orbital floor fracture, orbital foreign body), or inflammatory etiologies (granulomatosis with polyangiitis, orbital pseudotumor, thyroid eye disease, sarcoidosis, Tolosa-Hunt syndrome). More chronic presentations with insidious onset can result from neoplastic etiologies (orbital apex tumors, cavernous sinus mass, paranasal sinus tumors, skull base tumors, metastases) and vascular etiologies (arteriovenous fistula, carotid aneurysm). Complete unilateral external ophthalmoplegia can result from neuromuscular etiologies, such as myasthenia gravis, although pupil involvement would not be expected.

Diagnosis and Discussion

Cavernous carotid aneurysms (CCAs) are lesions of the intracavernous portion of the internal carotid artery that remain separated from the subarachnoid space by the venous dural lining of the cavernous sinus and account for 2%–6% of all intracranial aneurysms.[1,2] CCAs are categorized according to their maximal diameter: small aneurysms, <1 cm; large aneurysms, 1–2.5 cm; giant aneurysms, >2.5 cm. Corresponding risk of aneurysmal rupture over 5 years is 0%, 3%, and 6.4%, respectively.[2,3] Fortunately, the risk of subarachnoid hemorrhage following CCA rupture is low, and although these entities are generally considered benign, focal neurologic deficits such as diplopia, pain, or facial paresthesias may warrant intervention in select patients.[1,4]

Epidemiologically, diplopia and orbital pain are the most common clinical symptoms, although partial ophthalmo-

plegia (with the sixth cranial nerve being most commonly affected because of to its location adjacent to the internal carotid artery within the cavernous sinus) and ptosis are the most common clinical signs.[1,4] Our patient exhibited additional ocular signs attributed to the aneurysm, including complete ophthalmoplegia, proptosis, and optic neuropathy. These signs are relatively rare presentations of a giant CCA. The largest case series of 206 CCAs of all sizes found that 18% of patients with CCAs exhibited a cavernous sinus syndrome involving cranial nerves 3, 4, and 6 in varying degrees, whereas a second series of giant CCAs found that only 16% exhibited complete ophthalmoplegia.[1,4] Studies have shown that only 7% of CCAs exhibit proptosis, which is thought to be due to both laxity of the atrophied extraocular muscles allowing forward motion of the globe and also increased dural sinus congestion and venous pressure.[4] Notably, complete third nerve palsy, which our patient had, can also result in up to three millimeters of proptosis due to resultant extraocular muscle laxity. In our case, wasting of the extraocular muscles was evident on imaging studies as a result of the long-standing lesion. Although decreased vision in these cases can result from varying etiologies, such as corneal hypesthesia and keratopathy, compressive optic neuropathy is a rare cause of vision loss, as was the case in our patient.[1]

The patient also exhibited evidence of unilateral trigeminal neuropathy. The fifth cranial nerve supplies both sensory and motor innervation to facial structures. The first and second branch travel through the cavernous sinus, and the third branch exits the skull base through the foramen ovale prior to entering the cavernous sinus. Large cavernous sinus lesions, such as our patient's aneurysm, exert sufficient mass effect to impinge on the third branch of the fifth cranial nerve prior to exiting the skull base. Our patient experienced corneal thinning and scarring suggestive of neurotrophic cornea as well as a right jaw thrust from unilateral pterygoid muscle atrophy

and temporalis muscle wasting, indicating compressive neuropathy of the motor branch of the fifth cranial nerve. Muscle paralysis of the pterygoid and temporalis muscles from compressive CCA is a rare clinical sign, present in only 4% of cases and resulting from compression of the trigeminal ganglion in Meckel's cave.[4]

In our case, the patient initially sought medical care as a result of seizurelike activity. While intracranial aneurysms rarely cause seizurelike activity, case reports of internal carotid aneurysms causing epileptiform seizures do exist.[5,6] To date, existing literature suggests that intracranial aneurysms are more likely to be epileptogenic if they are in direct contact with anterior temporomedial brain structures.[6] Indeed, CT and MRI imaging in our case showed displacement of temporal lobe parenchyma by the aneurysm, which lends support to the belief that the aneurysm may have led to the patient's seizurelike symptoms.

Prior studies have found that giant CCA are more common in women and in advanced age.[1] While many CCA are idiopathic, these lesions may arise as a result of connective tissue disease, neoplasm, or vasculitis.[7] Specific subtypes of CCA result from trauma or infection, although these distinct entities each warrant unique evaluation and treatment strategies.[8,9] Given the non-life-threatening nature of these lesions, many warrant clinical observation initially. Studies have shown that untreated lesions may spontaneously regress, and that mild symptoms can spontaneously improve.[1] However, orbital pain or symptomatic diplopia may persuade some to consider neurosurgical intervention, as studies have shown that intervention is associated with a higher rate of resolution of CCA-related pain.[1] Notably, treatment has not been found to reliably forecast improvement of diplopia, although shorter duration of ophthalmoplegia is typically associated with a better chance of recovery.[1] Current treatment options for unruptured aneurysms include aneurysmal sac clamping, ligation, balloon occlusion of the proximal internal carotid artery, and most recently, flow diversion therapy.[2,10,11]

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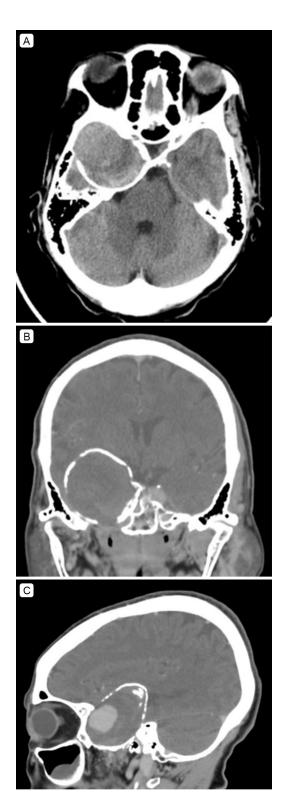
Figure 2. Computed tomography (CT) imaging demonstrating a giant cavernous carotid aneurysm. Axial CT (A) shows a calcified aneurysmal lining with displacement of the right temporal lobe; coronal (B) and sagittal (C) CT images demonstrate the large size of the giant aneurysm.
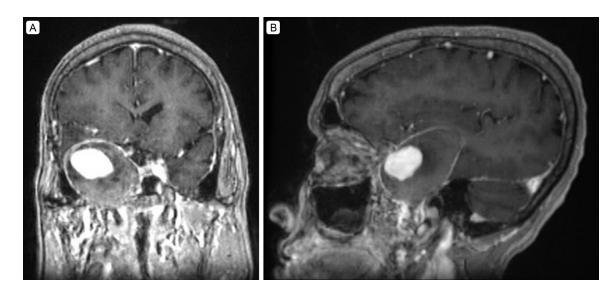
Figure 3. T1-weighted coronal (A) and sagittal (B) post-contrast magnetic resonance imaging (MRI) demonstrating a giant cavernous carotid aneurysm with partial thrombosis of the peripheral aneurysmal sac.

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