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Expert Opinion

Headache as the Only Symptom of a Spontaneous Dural Carotid-Cavernous Fistula

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When the cause of a headache is not certain or the headache is nonresponsive to treatment, a multitude of health care providers are often consulted. An astute physician may reinterpret a "normal" study and make the correct diagnosis.

CASE HISTORY

A 59-year-old female with a history of type II diabetes and hypothyroidism presented with a new onset constant daily right-sided throbbing temporal headache. For the first 2 months, the headache had an intensity on a visual analog scale ranging from a 4–5/10 but then increased to a 8–10/10. The headache was less often an occipital aching or sharp pain that radiated from the roof of her mouth to the vertex of her head. Initially, there were no associated symptoms including sinus congestion, eye redness, blurred vision, photophobia, ptosis, nasal congestion or drainage, proptosis, diplopia, nausea, or tinnitus. There was no prior history of headaches. She denied visual aura, jaw claudication, amaurosis fugax, myalgia, or scalp tenderness. She had lost some weight due to loss of appetite because of the headache.

A month after the headache started, she noted a loud roaring constant pulsatile tinnitus in the right ear. She denied any hearing loss. She consulted an ENT physician for the tinnitus and his examination of her was reportedly normal. Over the next 3 months, she saw numerous health care providers. She was then seen at an ER and advised to see a neurologist who obtained a noncontrast head CT that was reported negative. An MRI of the brain with and without contrast was also reported as normal. She then had an occipital nerve block that did not help. She was placed on Neurontin and Elavil for a few weeks without benefit. She then saw another neurologist who tried Indocin and oral prednisone for 3 days without relief. An orthodontist found normal temporomandibular joints. An orthopedist then administered three more occipital steroid injections without help. The patient's gynecologist tried Valtrex for possible herpetic neuralgia but it did not help. An optometrist reported a normal eye examination. Treatment by a chiropractor for 3 weeks was of mild benefit.

Two months after headache onset, her husband noted that the right eye seemed to be slightly puffy and more prominent as compared to the left. Soon thereafter she also began noting right eye pain with eye movement; however, she denied eye redness, diplopia, facial numbness, or blurred vision.

Three months after the headache onset, a third neurologist, started her on Ultracet, and this did not provide relief. Her CBC was normal, ESR was 10, and

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CRP was negative. An MRI of the brain and orbits with and without contrast, MRA of head and neck, MRV of brain, and CT angiography of the brain were all interpreted as normal by a neuroradiologist. She was sent to a neuro-ophthalmologist.

Three-and-a-half months after headache onset, while vacationing in Mexico, she was hospitalized for dehydration. She was started on Alparazolam and Haldol, in addition to Maxalt, which she was already taking. After being on these medications for a week, she noted decreased severity of the headache to 3–4/10. For a week, she had also noted binocular diplopia at both far and near.

Neuro-ophthalmological examination 4 months after headache onset revealed best corrected visual acuity of 20/15 in each eye with a moderate hyperopic correction. Color vision and amsler grid were normal, Humphrey 24-2 size III, SITA STD showed some nonspecific changes. She had bilateral ptosis relatively symmetric. Examination of the pupils showed 0.25 mm anisocoria in both light and dark and no dilation lag and normal reaction and no afferent pupillary defect. Exophthalmometry measured 17 mm OD and 15 mm OS, with no abnormal retropulsion. Motility examination showed the ductions and versions to appear normal; however, with cover testing it was apparent that she had a small esotropia in right gaze (8 to 10 prism diopters) with a right hypertropia of 6 prism, she had in left gaze an exophoria of 6 prism diopters, in up gaze she had 4 to 6 prism of a right hypotropia and in down gaze a 4 prism diopter right hypertropia. These measurements supported a partial right VIth nerve paresis and a subtle partial right III nerve paresis. Slit lamp examination showed subtle conjunctival chemosis on the right eye, but no evidence of arterialization of the conjunctival vessels. The of slit lamp examination is unremarkable. Intraocular pressure (IOP) was 19 OU. Gonioscopy showed narrow, nonoccludable anterior chamber angles OU without blood in Schlemm's canal. Fundus examination revealed a solitary cotton wool spot along the left inferior temporal arcade and tiny spot of retinal pigmentary epithelium (RPE) atrophy above the fovea OD. There was no other evidence of diabetic retinopathy disc, hemorrhages, or neovascularization. Cranial nerves V, VII, IX, X, and XII were intact. A bruit could not be heard over the cranium or eye.

The MRI was reinterpreted by the neuro-ophthalmologist who noted a vessel suggestive of a posterior draining vein from the right cavernous sinus. The superior ophthalmic vein was not enlarged.

Subsequently, the patient had a catheter cerebral arteriogram that documented a dural fistula supplied by both internal and external branches of the carotid for which she underwent endovascular treatment.

Postoperatively the pulsatile tinnitus resolved immediately, although she experienced severe orbital and head pain for a period of about 2 weeks. She also developed a near total right VI nerve palsy. After 2 weeks, the orbital and head pain totally disappeared and after several months the right VI nerve paresis totally resolved. Ten months later, she continues to be pain-free and diplopia-free and is doing well. A follow-up arteriogram 9 months after the endovascular treatment shows no evidence of recurrent or residual fistula.

Question 1.—What are carotid-cavernous fistulas? How often are headaches associated?

EXPERT OPINION

Carotid-cavernous sinus fistulas (CCF) are acquired pathological climinata shunts from the cavernous portion of the internal carotid artery (ICA) into the enveloping cavernous sinus. The overwhelming majority (80%) are the result of a traumatic injury to the ICA or a branch artery, while the rest can develop spontaneously.[3] Spontaneous CCFs arise from a variety of etiologies that predispose to a weakness in the wall of a cavernous branch artery or the ICA.[3] About 60% of spontaneous CCF cases are secondary to a specific disorder. These predisposing disorders include Ehlers-Danlos syndrome, pseudoxanthoma elasticum, fibromuscular dysplasia, aneurysm of the cavernous ICA, a persistent embryological trigeminal artery, and other nonspecific angiodysplasia. In general, CCFs are classified as either direct or indirect (dural).[4]

Both direct and indirect fistulas occur. A direct fistula comes directly from the trunk of the ICA while indirect fistulas arise from the small branches of the ICA. Both direct and indirect fistulas can be caused

traumatically or spontaneously. However, direct fistulas are much more likely to be posttraumatic, while indirect fistulas are more likely to have a spontaneous occurrence. Direct fistulas usually have faster flow characteristics and therefore have more proptosis, chemosis, and are more often associated with an audible bruit than the indirect fistulas. The indirect fistulas as described in this case, are more insidious in nature and more commonly affect both sides with the inciting side due to the fistula itself and the other side affected because of recruitment.

Fistulas may drain anteriorly, posteriorly, or both. When the fistula drains anteriorly, it drains toward the orbit in which case there is usually a large superior ophthalmic vein, under high pressure leading to decreased venous return from the orbit, that may cause the orbital muscles to increase in size thus creating proptosis. When the fistula drains posteriorly, it drains away from the orbit and therefore a large superior ophthalmic vein or orbital congestion may not occur. It is understood that when a fistula drains anteriorly, there is little risk of an intracranial or subarachnoid hemorrhage as the venous hypertension is well maintained in the cavernous sinus and orbit. However, when a fistula drains posteriorly, the problem that arises is that the venous hypertension involves the intracranial cortical veins outside of the protective coat of the cavernous sinus. Venous cortical hypertension, if high enough, could lead to rupture of such veins and cause subarachnoid and/or intraparenchymal hemorrhage. Therefore, posterior draining fistulas are not as "benign" as anterior draining fistulas for life. However, anterior draining fistulas may cause devastating problems to one or both eyes. Problems to the eye in anterior draining fistulas include ocular ischemia with vision loss, glaucoma (due to raised episcleral venous pressure), oculomotor disorders, and marked exophthalmos with corneal exposure.

Ocular pain (16%) and headache (40%) are usually present in CCFs; however, they rarely manifest as the only clinical symptoms.[1,4,5] Herein, we describe 1 patient in whom ocular and head pain were the only manifestations initially and remained the main manifestations even when there were other signs that developed indicating a cavernous sinus syndrome.

The clinical presentation of anterior draining dural CCF usually presents with conjunctival injection, chemosis, proptosis, diplopia, reduced visual acuity, elevated IOP, ophthalmoplegia with or without associated cranial nerve palsies (III, IV, and VI) and periorbital bruit. Ocular and head pain is more often a minor component of the presentation, and when present, it is not typically the only symptom as was the case initially with our patient.[4] The most likely cause of the ocular and head pain is stretching of the richly innervated dural membrane in dural CCF include of the cavernous sinus, venous thrombosis, venous hemorrhage, or pathologic involvement of the V cranial nerve, either by way of direct pressure in the cavernous sinus or pulsatile pressure from abnormal arterial flow into the sinus.[2] Severe or focal head pain is an ominous complaint that warrants urgent attention and evaluation because it may signify stretching of cerebral veins from posterior cortical venous drainage. As previously stated, posterior drainage from CCF, leading to venous hypertension, exposes the patient to the risk of intracranial hemorrhage which can be fatal.

Although there were a paucity of findings on imaging in this patient, her examination findings later in her presentation, pointed to a cavernous sinus process and combined with her history of pulsatile tinnitus (also later in her presentation), this was consistent with a right dural CCF. The prominence of her headache, the mildness of proptosis and congestion (and normal IOP), and lack of a large superior ophthalmic vein suggested that her fistula was draining mostly posteriorly and therefore putting her at some risk for intracranial hemorrhage. As to why she had so much facial pain one may speculate that the fistula was extensively involving a long segment of the wall of the cavernous sinus. The supply was quite extensive with multiple small dural branches from multiple sites. In this situation, it is very conceivable that the cranial nerves can be irritated just from an extensive arterial network. In addition, the venous drainage from the fistula was to cortical veins and it may have been a combination of extensive dural arterial meshwork and deep venous drainage that accounted for her pain.

Question 2.—What other vascular causes of headaches does a neuro-ophthalmologist see?

There are various causes of headaches attributable to vascular lesions. Often, the pain characteristics are really not diagnostic and the headaches can mimic primary headaches.[5] The key in the history is clearly the onset of a new headache and other symptoms and signs that accompany the pain in the patient. This discussion therefore pertains to vascular lesions that are not obvious on standard imaging and may require MRA/MRV and/or angiography. We are excluding headaches due to strokes or vasculitis here. The vascular lesions that can present with headache/eye pain and are not be obvious on MRI include: CCF as in our case, cervical or vertebral basilar dissection, venous sinus thrombosis, and rarely small cerebral AVMs that are small and only obvious on catheter angiogram.

In the case of a CCF headache the presence of tinnitus, proptosis, diplopia, etc. are helpful features.

In the case of a cervical carotid dissection, in addition to ocular and head pain, nuchal pain may be an additional feature. A history of cervical trauma or chiropractic manipulation may or may not be present. A history of a transient ischemic attack involving the carotid circulation (eye or brain) may be recalled and the finding of a Horner's syndrome ipsilateral to the carotid circulation involved is key. The Horner's syndrome may only be diagnosed in the dark and only obvious in the first 5 seconds of the darkened examination room. It is crucial to see this "dilation lag" in the first 5 seconds because the side of the Horner syndrome can catch up with the normal side after 15 seconds. Similarly, a vertebral artery dissection may be associated with a Horner's syndrome as well as vertebral basilar TIAs.

Cerebral venous thrombosis in its most benign form, may be difficult to see if it is not involving the sagittal sinus and may present with symptoms of raised intracranial pressure and simulate the presentation of pseudotumor cerebri with papilledema and transient obsurations of vision. There may also be pulsatile tinnitus. Similarly, small arteriovenous cerebral malformations may present with raised intracranial pressure and a syndrome mimicking pseudotumor cerebri with papilledema and tinnitus.

This case illustrates that ocular and head pain as a prominent symptom can occur as the main manifestation of CCF. Therefore, clinicians should consider the diagnosis of CCF in patients with this type of pain of unknown etiology. A neuro-ophthalmic examination may uncover subtle CN paresis that is not obvious grossly as well as find clues of raised IOP. An MRI may be negative or may show subtle signs that may be missed by a radiologist. An MRA may be negative and the diagnosis can only be confirmed by an arteriogram.

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