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New-Onset Headache in a 71-Year-Old Man

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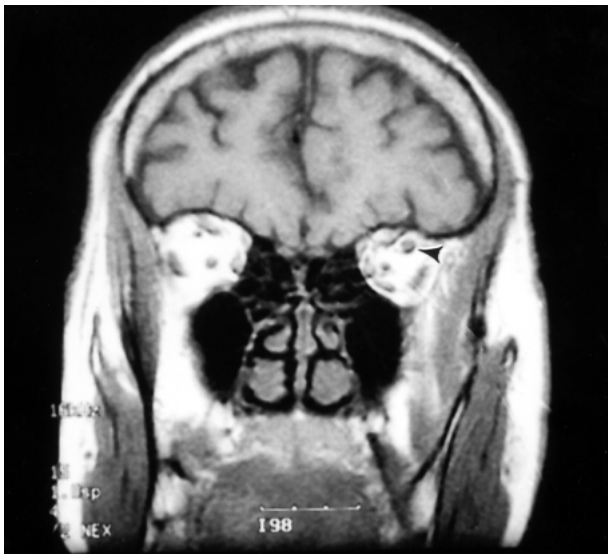

Figure 1. Coronal T1-weighted magnetic resonance image of the brain and orbits. Note the aberrant structure on the left side (*arrowhead*).

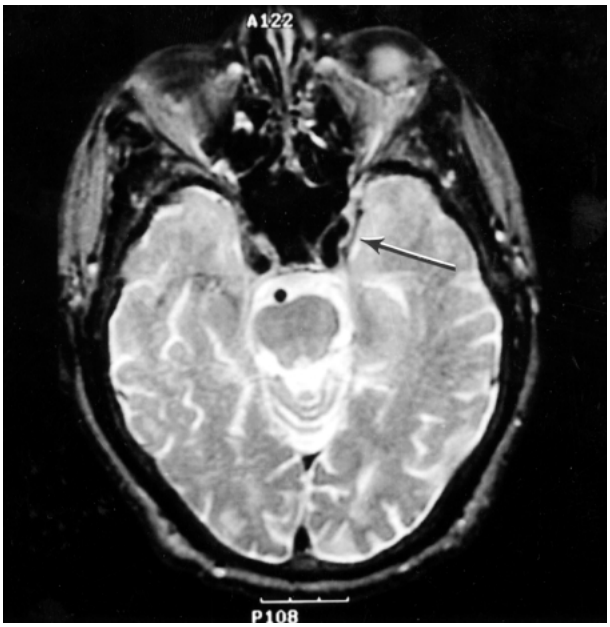

Figure 2. Axial T2-weighted magnetic resonance image of the brain at the level of the cavernous sinus. An aberrant flow void is visible (*arrow*).

CASE PRESENTATION

A 71-year-old man developed left hemicranial headaches associated with pulsatile tinnitus 6 months before admission. He denied jaw claudication or generalized myalgia. Three weeks before evaluation, the tinnitus changed abruptly, resolving completely except during Valsalva maneuver. The head pain persisted. At approximately the same time, the patient began experiencing horizontal binocular diplopia that was most prominent when he looked toward the left.

Physical examination revealed subtle proptosis, mild periorbital edema, and marked conjunctival injection on the left side. No ocular pulsation or auscultatory abnormality was detected around the globe or mastoid process. Funduscopic examination revealed mild dilatation of retinal veins in the left eye, but no papilledema or retinal hemorrhage was observed; otoscopy likewise was unremarkable. The temporal arterial pulse was normal bilaterally, and firm palpation in this region failed to elicit focal tenderness. Other than a moderate abduction deficit of the left eye, results of the neurologic examination were normal.

Laboratory studies (including complete blood count with differential, sedimentation rate, C-reactive protein level, syphilis serology, thyroid function tests,

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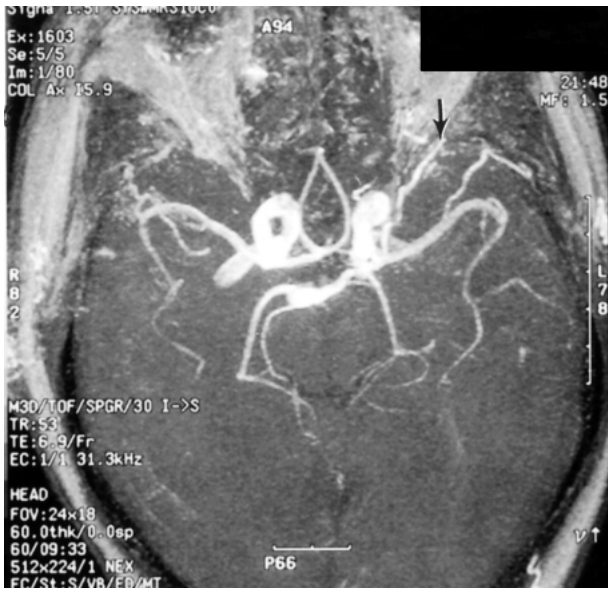

Figure 3. Magnetic resonance angiography further demonstrates the aberrant flow void (*arrow*) exhibited in Figure 2.

and a full chemistry panel including hemoglobin A_{1C}) were nondiagnostic. Magnetic resonance imaging (MRI) of the brain demonstrated an anomalous structure, possibly corresponding to a distended superior ophthalmic vein in the left orbit (**Figure 1**). Axial MRI sections demonstrated an aberrant flow void medial to the left medial temporal lobe and lateral to the left cavernous sinus (**Figure 2**), a finding more conspicuous on 3-dimensional time-of-flight MR angiography at the level of the circle of Willis (**Figure 3**).

WHAT IS YOUR DIAGNOSIS?

A) Benign intracranial hypertension
B) Carotid-cavernous (CC) sinus fistula
C) Cavernous sinus thrombosis
D) Temporal arteritis

ANSWER

The correct answer is CC sinus fistula (B).

DISCUSSION

New-onset headache in an older adult raises a number of differential possibilities, many of which demand prompt intervention to prevent complications. Important historical and physical examination features in the present case include pulsatile tinnitus and unilateral lateral rectus palsy.

Pulsatile tinnitus refers to an auditory percept, synchronous with the pulse, in one or both ears that occurs without an external stimulus. The phenomenon occurs when auditory signals originate from vibration transmitted to the cochlea through routes other than tympanic membrane displacement by airborne sound waves. For example, pulsatile tinnitus can result from turbulent flow in vascular structures adjacent to the inner ear. Turbulence may be caused by systemic factors producing a hyperdynamic state (eg, hyperthyroidism, anemia). Alternatively, turbulent flow can develop in the setting of various vascular lesions. Differential diagnosis of pulsatile tinnitus includes glomus jugulare tumor (paraganglioma), carotid artery dissection, intracranial atherosclerosis, dehiscence of the carotid artery or internal jugular vein into the middle ear, and dural arteriovenous fistula involving the cerebral sinuses.[1–4] Other causes include congenital anatomic variants such as a superiorly displaced jugular bulb, persistence of the stapedial artery, and an aberrant mastoid emissary vein.[5–7] When caused by intracranial arteriovenous fistulae or intracranial arterial stenosis, compression of the ipsilateral carotid artery can reduce or eliminate the tinnitus, a diagnostic maneuver not available in the case described.

Benign intracranial hypertension (BIH) or pseudotumor cerebri primarily presents with headache. Pulsatile tinnitus and extraocular palsies also have been attributed to BIH.[8,9] Pulsatile tinnitus in BIH may arise from the effect of increased intracranial pressure on flow through the jugular bulb. In contrast to the case described above, however, headache typically does not localize to one side. More importantly, the vast majority of cases of BIH develop in young female patients. Clinical suspicion increases if funduscopy demonstrates papilledema without explanation on neuroimaging, neither of which pertained to the patient discussed here.

Cavernous sinus thrombosis (CST) causes many of the symptoms present in the case patient, including retro-orbital pain, conjunctival injection and chemosis, and ophthalmoparesis. Almost universally, CST develops in the setting of uncontrolled infection situated in the medial face, orbit, paranasal sinuses, or temporal bone.[10] Less commonly, CST may complicate hypercoagulable states or vascular anomalies such as aneurysm. Accordingly, constitutional symptoms and signs, such as fever, chills, and malaise accompany CST in most instances. The oculomotor, trochlear, and abducens nerves as well as the V1 and V2 divisions of the fifth cranial nerves traverse the cavernous sinus. Therefore, the complex ophthalmoplegia associated with CST is often accompanied by facial sensory symptoms rather than isolated lateral rectus dysfunction as observed in our patient.[11] High-resolution contrast-enhanced cranial computed tomography (CT) confirms the diagnosis if

it reveals confluent areas of non-opacification that correspond to thrombus within the venous sinus. On brain MRI, CST manifests as abnormal T1-signal intensity arising from the thrombus expanding the cavernous sinus. Gadolinium-enhanced MRI exposes filling defects in the corresponding region.[12,13]

Temporal (cranial) arteritis usually occurs during or after the sixth decade of life. It is characterized by headache and is often accompanied by the systemic symptoms of polymyalgia rheumatica.[14,15] Besides unilateral head pain, temporal arteritis may produce ophthalmoparesis. However, to our knowledge, pulsatile tinnitus has not been described in association with temporal arteritis, and vascular engorgement of orbital contents would not be expected. Furthermore, in the present case, an elevated erythrocyte sedimentation rate or C-reactive protein level was not detected, making the diagnosis of temporal arteritis less tenable.

The physical manifestations of a fistula between the intracavernous segment of the carotid artery and the cavernous sinus depend on several factors. First, the mechanism of injury influences presentation, with abrupt onset of symptoms occurring in the 75% of patients who present after penetrating orbital injury or basilar skull fracture, and a more insidious or delayed onset after spontaneous arterial rupture.[16] Secondly, variations in venous sinus anatomy permit egress of blood in multiple directions; anterior fistulae transmit flow forward via the ophthalmic veins to the external jugular system, whereas posterior fistulae drain predominantly via the inferior and superior petrosal sinuses into the internal jugular vein.[17] Because of the importance of the cavernous sinus in orbital venous drainage, CC fistulae produce dramatic alterations in ocular and adnexal appearance and function, with severe chemosis, conjunctival injection, and protrusion of the globe.[18] As in the patient described here, isolated abducens or sixth cranial nerve palsy represents the most common oculomotor deficit caused by CC fistulae. Restrictive ophthalmoparesis also may develop simply through congestion of orbital contents. In addition, CC fistulae can produce pulsatile tinnitus, presumably due to rhythmic vibration of venous structures in contact with the temporal bone (ie, petrosal sinus). Auscultation of the affected orbit over a closed eyelid may disclose a bruit that permits examiners to share their patient's auditory experience.

Prior to the availability of MRI technology, radiographic confirmation of a CC fistula necessitated contrast angiography, an invasive and potentially morbid procedure. Beam hardening produced by the adjacent apex of the petrous temporal bone and the clinoid process pose particular difficulty in the interpretation of CT images of this region. In contrast, standard MRI sequences readily identify aberrant flow voids resulting from "arterialization" of venous channels that ordinarily flow toward the cavernous sinus.[19] Associated findings include thrombus formation within the sinus, similar to CST. MR angiography provides noninvasive images of large- and medium-sized arteries as well as abnormal venous channels that carry blood at arterial pressure.

Monocular visual failure due to venous stasis represents the most common serious complication of CC fistulae. In contrast, symptomatic intracranial hemorrhage rarely occurs, presumably due to the capacity of the venous sinus to adapt to increased pressure and flow. Arterial pressure raises venous pressure inside orbital structures, which can lead to ischemia from venous stasis and secondary glaucoma from elevated intraocular pressure. To salvage vision, surgical interventions in the past entailed risky procedures such as carotid artery ligation. More modern approaches to CC fistula repair, not as widely available, include endovascular correction, wherein the anomolous vascular passage is repaired through embolization or stenting.[20] For less rapidly progressive cases with milder manifestations, the fistula may obliterate with repetitive, mild compression of the cervical common carotid artery, an external maneuver that can be performed by the patient. This maneuver was used successfully in the case described here. **HP**

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