Multifocal Dural Enhancement Associated With Temporal Arteritis

Ealon Joelson, MD; Breanna Ruthrauff, MD; Feraz Ali, MD; Neil Lindeman, MD; Frank R. Sharp, MD

Objective: To report the association of temporal arteritis and enhancement of the dura mater and temporalis muscle.

Design and Methods: A single patient with a complaint of headache and diplopia was studied.

Setting: Academic medical center.

Patient: A 69-year-old man presented with lateral rectus weakness, temporal artery tenderness, and an erythrocyte sedimentation rate of 65 mm/h.

Intervention: Biopsy of temporal artery and dura mater.

Main Outcome Measures: Brain magnetic resonance imaging and pathological findings.

Results: Magnetic resonance imaging of the brain showed multifocal dural enhancement and enhancement of the temporalis muscles. The temporal artery showed a necrotizing vasculitis and the dura showed perivascular inflammatory cells.

Conclusion: It is proposed that the temporal arteritis caused the multifocal dural enhancement and temporalis muscle enhancement on magnetic resonance imaging.

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WITH THE advent of magnetic resonance imaging (MRI), several entities have been particularly well seen by MRI. Isolated diffuse dural enhancement or multifocal dural enhancement can be visualized by MRI and spare the leptomeninges. Causes of diffuse dural enhancement include intracranial hypotension and causes of multifocal dural enhancement include cranial operations and dural carcinomatosis.\textsuperscript{1-5} Enhancement of the dura follows the contour of the skull whereas leptomeningeal enhancement is adjacent to cortical gyri.\textsuperscript{1-5}

Temporal arteritis is a well-recognized rheumatological disorder not known to involve the dura. The involved arteries, including the external carotid and ophthalmic, typically have a prominent external elastic membrane.\textsuperscript{6,7} As cranial blood vessels approach the dura they lose the external elastic membrane within a few millimeters of entering the dura.\textsuperscript{6,7} Because the clinically relevant vasculitis due to temporal arteritis often occurs in the distribution of the external carotid artery, it is possible that the dura, which is supplied by that vessel, might also be affected in some cases. However, there have been no well-documented reports of this occurring. We present clinical, imaging, and pathological support for the premise that the dura can be affected by temporal arteritis.

REPORT OF A CASE

The patient is a 69-year-old white, diabetic man who complained of headache and double vision starting approximately 1 month prior to admission. He was treated as an outpatient with antibiotics for a presumed sinus headache. He was noted to have right lateral rectus weakness that was initially attributed to diabetes. He was admitted because of an associated 9-kg weight loss, myalgias, and fatigue. During hospitalization, his body temperature ranged from 37.2°C to 37.6°C. He had right temporal tenderness and a complete right lateral rectus palsy. He also had decreased downward and medial movement of the right eye with diplopia in those respective directions of gaze. Upgaze and left eye movements were normal. The pupils, the retinas, and the optic nerves were normal. He complained of bilateral tongue numbness. However, touch and pain tongue sensation and taste sensation were normal on direct testing. He had decreased reflexes and a distal sensory loss in the legs consistent with a diabetic polyneuropathy.

From the Departments of Neurology (Drs Joelson, Ruthrauff, and Sharp), Neuroradiology (Dr Ali), and Neuropathology (Dr Lindeman), University of California, San Francisco. Dr Sharp is now with the Department of Neurology, University of Cincinnati, Cincinnati, Ohio.
Laboratory values for complete blood cell count, total eosinophils, electrolytes, serum urea nitrogen, creatinine, rapid plasma reagin, and liver function tests were normal. Urinalysis revealed the following: protein, 0.3 g/L; red blood cell count, 1 to 5 × 10⁶/L; white blood cell count, 0 to 3 × 10⁶/L; and cultures positive for enterococcus. Blood glucose level ranged from 6.7 to 10.0 mmol/L (120-180 mg/dL). No abnormalities were seen on chest x-rays and abdominal and pelvic computed tomographic scans. Magnetic resonance imaging of the head performed with and without gadolinium showed thickening of the dura, multifocal dural enhancement (Figure 1, A, arrows), right temporalis muscle enhancement (Figure 1, B), and equivocal right lateral rectus muscle enhancement (not shown). There was no leptomeningeal enhancement and there were no intracranial brain abnormalities. Lumbar puncture revealed the following values: opening pressure, 145 mm H₂O; mononuclear cells, 6 cells/µL (>0.98 lymphocytes); glucose, 7.1 mmol/L (128 mg/dL) (serum glucose, 8.6 mmol/L [155 mg/dL]); and protein, 78 mg/100 mL. Results of cytologic studies and cultures (including fungal and acid-fast bacillus) were negative. Test results for human immunodeficiency virus and hepatitis B were negative. Urine histoplasma antigen was negative. Cryptococcal antigen and coccidiomycosis titers were negative in the serum and cerebrospinal fluid. The C3, C4, angiotensin-converting enzyme, antinuclear antibody, antineutrophil cytoplasmic antibody, double-stranded DNA, and CH₅₀ values were normal, but the erythrocyte sedimentation rate was 65 mm/h and the rheumatoid factor was 1:80. The patient was treated with 70 mg/d of prednisone. His headache resolved the next day. Three months later, when his lateral rectus weakness had resolved and his sedimentation rate was 15 mm/h, no abnormalities were noted on a gadolinium-enhanced brain MRI (not shown).

**PATHOLOGICAL FINDINGS**

The temporal artery biopsy specimen showed an inflammatory infiltrate within and around the temporal artery that included neutrophils, lymphocytes, and macrophages (Figure 2, A). There was fibrinoid necrosis of the arterial wall (Figure 2, B). There were no giant cells as occurs in half the patients with temporal arteritis.⁶,⁷ Analysis of the dural biopsy specimen demonstrated several foci of perivascular inflammation (Figure 2, C), although there was no definitive evidence of a vasculitis of the dural vessels. The inflammatory infiltrate around the dural vessels, however, was similar to that seen in and around the temporal artery (Figure 2, D). Gram stain and silver stains of the temporal artery and dura showed no bacteria or fungi. Biopsy findings of the temporal artery are consistent with a diagnosis of giant cell arteritis/temporal arteritis.

**COMMENT**

The patient was diagnosed as having temporal arteritis based on the headache, lateral rectus weakness, elevated sedimentation rate,⁶ and a temporal artery biopsy-proved vasculitis.⁶ As many as half the cases of temporal arteritis do not have giant cells on biopsy.⁷ Fibrinoid necrosis can be seen in the acute phase,⁷ as was true in this case. The rapid resolution of the headache and the elevated sedimentation rate following glucocorticoid therapy are also consistent with the diagnosis.³ The absence of systemic organ involvement and negative test results for systemic collagen vascular diseases makes other types of vasculitis unlikely.⁷

Temporal arteritis affects elderly patients with a preponderance in whites and females.⁸,⁹ The headache associated with temporal arteritis is often described as a continuous pain over the involved artery, occasionally with scalp tenderness,¹⁰ as occurred in our patient. Tongue ischemia may explain our patient’s complaint of tongue numbness, since temporalis, tongue and masseter claudication,⁶ scalp necrosis, gangrene of the tongue,¹¹ and bilateral tongue numbness have been reported in patients with temporal arteritis.¹² Constitutional symptoms, including weight loss, malaise, anorexia, muscle pain, and fever, as were seen in this patient, are common.⁸,¹⁰,¹²

Ophthalmologic symptoms include visual field defects—scotomata, sector cuts, and blindness—that occur sec-
ondary to anterior ischemic optic neuropathy, central retinal artery occlusion, or anterior segment ischemia. Transient and permanent ophthalmoplegias occur due to involvement of the vasonervorum to cranial nerves III, IV, and VI or to ischemia of the nutrient arteries to the affected eye muscle(s). The latter may be the cause of the lateral rectus weakness in our patient since there was equivocal contrast enhancement of that eye muscle.

Temporal arteritis is an inflammatory disease of arteries occurring primarily in the branches of the external carotid artery. The cause is presumed to be autoimmune because CD4 cell infiltrates and giant cells containing internal elastic membrane fragments and immune complexes are frequently demonstrable in the walls of affected vessels.

Dural enhancement and dural abnormalities due to temporal arteritis have not been previously reported. Dural enhancement is recognized to be a consequence of intracranial operations, neoplasms, intracranial hypertension from a persistent cerebrospinal fluid leak, and chronic hypertrophic pachymeningitis. Single cases of dural enhancement have been reported due to tuberculosis, aspergillus, syphilis, pseudotumor cerebri, and venous sinus thrombosis. Patients with sarcoidosis can have dural thickening with enhancement but without parenchymal disease in a small proportion of those with central nervous system involvement. Noncaseating granulomas are found on dural biopsy evaluation.

Rheumatological diseases other than temporal arteritis have been reported with dural enhancement. A patient with Wegener granulomatosis had dural enhancement on MRI and dural biopsy examination that showed necrotizing vasculitis. A patient with mixed connective tissue disease had dural enhancement by MRI, although there was no biopsy performed. A patient with rheumatoid arthritis had dural enhancement by MRI, but this patient also had intracranial hypotension that could have caused the dural enhancement. A patient with dural enhancement ascribed to “chronic hypertrophic pachymeningitis” had myocarditis and a positive antinuclear antibody and anti–double-stranded DNA, suggesting that a vasculitis caused the dural abnormality and dural enhancement on MRI.

The multifocal dural enhancement in the present case almost certainly was related to the temporal arteritis. The clinical picture was consistent with this diagnosis, results of the temporal artery biopsy showed arteritis, and the clinical and imaging findings resolved with glucocorticoid treatment. Since the dura is supplied mainly by the middle meningeal artery, which is a branch of the external carotid artery, the dura can be added to the list of structures that can be affected by temporal arteritis, including the scalp, eyes, extraocular muscles, temporalis muscles, jaw muscles, and tongue muscles. The dural enhancement presumably reflects “leaky” dural vessels.

Figure 2. Biopsy specimen of the temporal artery shows inflammatory cells (lymphocytes, leukocytes, and some macrophages) in the temporal artery (A and B) and its branches (A) with fibrinoid necrosis of the temporal artery wall (B). A similar inflammatory infiltrate occurred around smaller arteries in the dura (C and D).
Exactly how the temporal arteritis affected the dural vessels in this case is uncertain. Since temporal arteritis primarily affects vessels with a prominent external elastic membrane\textsuperscript{24,25} and may be an autoimmune disease directed at the external elastic membrane,\textsuperscript{14} there may not be a vasculitis of the dural vessels except for the few millimeters where the external carotid arteries retain their external elastic membrane as they penetrate the dura.\textsuperscript{24,25} This is consistent with the pathological features of the dural vessels in this case, since they do not show a definite vasculitis. It is possible that vasculitis of the external carotid proximal to the dura produced secondary changes in the dural vessels resulting in perivascular inflammatory cells and enhancement of the dura on MRI. This could have occurred due to temporal arteritis induced ischemia of dural vessels or embolism to dural vessels. Alternatively, vasculitis directly involving dural vessels could also explain the contrast enhancement. This would be consistent with rare reports of arteritis of intracranial vessels, that have no external elastic membrane, in patients with temporal arteritis and cerebral infarction.\textsuperscript{8,10,12,23}

The multifocal dural enhancement seen in this case can be differentiated from the true diffuse dural enhancement of all of the dura that occurs following intracranial hypotension associated with a “sagging brain.” This case also highlights the need to be aware of enhancement of cranial structures in the distribution of the external carotid artery as a sign of ischemia due to temporal arteritis and other types of vasculitides that should aid in proper diagnosis and treatment.

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Reprints: Frank R. Sharp, MD, Department of Neurology, University of Cincinnati Medical Center, PO Box 670525, 231 Bethesda Ave, ML 0525, Cincinnati, OH 45267-0525.

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