Hypertrophic Pachymeningitis as a Potential Cause of Headache Associated with Temporal Arteritis

Shin-ichi Tokushige¹, Hiroyoshi Matsuura¹, Takuto Hideyama¹, Koichi Tamura², Risa Maekawa¹ and Yasushi Shiio¹

Abstract

We herein describe a rare case of temporal arteritis associated with hypertrophic pachymeningitis. An 81-year-old man presented with a right temporal headache that had persisted for one month. A right superficial temporal artery biopsy revealed intimal hypertrophy with increased elastic fibers, consistent with temporal arteritis. Brain MRI using gadolinium enhancement showed thickened dura mater on the right frontal and temporal lobes, which led to the diagnosis of hypertrophic pachymeningitis. Intravenous methylprednisolone and oral prednisolone improved the patient’s symptoms. According to our findings, hypertrophic pachymeningitis may be a potential cause of an ipsilateral temporal headache associated with temporal arteritis.

Key words: temporal arteritis, hypertrophic pachymeningitis, headache, dura mater


Introduction

Temporal arteritis is an inflammatory giant cell vasculitis that tends to affect elderly patients and is characterized by a temporal headache and a fever (1). A variety of symptoms, such as vision loss, tongue ischemia, masseter claudication and scalp necrosis, has been reported in patients with temporal arteritis, possibly due to the involvement of branches of the external carotid artery.

Hypertrophic pachymeningitis is a clinical entity that is characterized by inflammation, fibrosis and thickening of the dura mater (2). It sometimes occurs concurrently with disorders such as infections, autoimmune diseases and neoplasms. However, few case reports have described hypertrophic pachymeningitis associated with temporal arteritis (3-7).

We herein present a rare case of temporal arteritis associated with hypertrophic pachymeningitis and propose the hypothesis that dural inflammation can cause an ipsilateral temporal headache associated with temporal arteritis.

Case Report

An 81-year-old man presented to our department following a low-grade fever and right temporal headache which persisted for one month. His blood pressure was 102/74 mmHg, his pulse was 112 beats/min, and his body temperature was 36.5°C. Neurological examinations revealed right ptosis and the loss of lower visual field in the right eye without ophthalmoptosis. A fundoscopic examination showed ischemic optic neuritis in the right eye. No other neurological symptoms were noted.

Routine laboratory test results showed a normal blood cell count and elevated C-reactive protein (CRP) (11.1 mg/dL), erythrocyte sedimentation rate (ESR) (120 mm/h) and serum IgG (1,822 mg/dL) levels; however, the serum IgG4 level was not elevated. Serological tests showed that anti-nuclear antibodies (ANA), anti-neutrophil cytoplasmic antibodies (ANCA), anti-Smith antibody, anti-single stranded and anti-double stranded DNA antibodies and anti-Sjögren’s syndrome A/B antibodies were negative. Angiotensin-converting enzyme levels were not elevated.

A cerebrospinal fluid (CSF) examination showed slight pleocytosis (9 cells/μL, all mononuclear leukocytes), ele-
Figure 1. Brain MRI. A post-gadolinium T1-weighted image on the second day (A, B) and the 16th day (C, D) after admission. The dura mater was partially thickened in the right frontal and temporal lobes using gadolinium enhancement (A, B, arrow), and this thickening improved after the initiation of corticosteroid therapy (C, D).

Elevated protein (64 mg/dL), normal glucose (CSF glucose 49 mg/dL, blood glucose 100 mg/dL) and a normal IgG index (0.47). CSF bacteriological examinations were negative, and CSF cytology showed no malignant cells.

Brain MRI revealed partial thickening of the dura mater with enhancement on the right frontal and temporal lobes in a T1-weighted image using gadolinium enhancement (Fig. 1A, B). T2-weighted, fluid-attenuated inversion recovery (FLAIR) and diffusion-weighted images showed no abnormal signal intensities in the brain. These MRI findings were consistent with partial hypertrophic pachymeningitis. The serum inflammatory markers and the elevated CSF cell count and protein also supported the diagnosis of hypertrophic pachymeningitis.

The patient fulfilled the American College of Rheumatology diagnostic criteria for temporal arteritis (8) for age (over 50 years), temporal headache and elevated ESR. A biopsy of the right superficial temporal artery showed intimal thickening and an increase in elastic fibers (Fig. 2) that was consistent with temporal arteritis, although giant cells were not observed.

He received a 3-day course of intravenous methylprednisolone (1,000 mg/day) followed by oral prednisolone (1 mg/kg daily). After the second day of treatment, his headache and other symptoms had substantially decreased. On the fourth day of treatment, the patient’s serum CRP and ESR were reduced to 1.96 mg/dL and 25 mm/h, respectively. Brain MRI on the 16th hospital day showed that the enhancement and dura mater thickening were improved (Fig. 1C, D). At discharge, he remained on oral prednisolone (0.5 mg/kg daily).

Discussion

We herein describe an 81-year-old man who was diagnosed with temporal arteritis associated with hypertrophic pachymeningitis. He presented with a right temporal headache, right ptosis and vision loss in the right eye, and a thickened dura mater that was identified using gadolinium enhancement; these were all resolved by intravenous and oral steroid therapy.

The association between temporal arteritis and hypertrophic pachymeningitis is not well recognized, and only five cases have been previously reported (Table). The mechanism of how these disorders could coexist is currently unknown. Because the dura mater is supplied by the middle meningeal artery, which is a branch of the external carotid artery, it is hypothesized that vasculitis of the external carotid artery proximal to the dura mater might spread distally to the dural vessels, resulting in dural inflammation (3). This hypothesis is supported by the dural biopsy findings in two previous cases of temporal arteritis with hypertrophic
Headache associated with temporal arteritis has been mainly ascribed to inflammation in the affected superficial temporal artery itself, because arteries are naturally pain-sensitive structures (9). In some patients whose headache is pachymeningitis (3, 6), which showed inflammation involving the small arteries in the dura mater. In addition, the laterality of temporal arteritis and thickened dura mater were identical in five of the six cases (Table), which also supports the concept that dural inflammation is caused by external carotid artery vasculitis proximal to the dura mater.

Table. Previous Cases of Temporal Arteritis Associated with Hypertrophic Pachymeningitis.

<table>
<thead>
<tr>
<th>Age/Sex</th>
<th>Symptom</th>
<th>Laterality of temporal arteritis</th>
<th>Laterality of dural inflammation</th>
<th>Treatment</th>
<th>Outcome</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>69/M</td>
<td>Right temporal headache, right lateral rectus palsy, tongue numbness</td>
<td>Right</td>
<td>Bilateral (Left dominant)</td>
<td>Oral steroid</td>
<td>Improved</td>
<td>(3)</td>
</tr>
<tr>
<td>65/F</td>
<td>Right temporal headache, photophobia, phonophobia</td>
<td>Right</td>
<td>Right</td>
<td>Oral steroid</td>
<td>Improved</td>
<td>(4)</td>
</tr>
<tr>
<td>80/F</td>
<td>Right-sided retro-orbital and temporal headache, malaise, pain on mastication</td>
<td>Right</td>
<td>Right</td>
<td>Intravenous and oral steroids</td>
<td>Clinically improved, but radiologically unchanged</td>
<td>(5)</td>
</tr>
<tr>
<td>59/M</td>
<td>Temporal and frontal headache, jaw claudication, a fever</td>
<td>Bilateral</td>
<td>Bilateral</td>
<td>Oral steroid</td>
<td>Improved</td>
<td>(6)</td>
</tr>
<tr>
<td>72/M</td>
<td>Left temporal headache</td>
<td>Left</td>
<td>Left</td>
<td>Oral steroid</td>
<td>Improved</td>
<td>(7)</td>
</tr>
<tr>
<td>81/M</td>
<td>Right temporal headache, right ptosis, visual field deficit in the right eye</td>
<td>Right</td>
<td>Right</td>
<td>Intravenous and oral steroids</td>
<td>Improved</td>
<td>This case</td>
</tr>
</tbody>
</table>

Figure 2. Histological findings of the right superficial temporal artery. (A) Hematoxylin and Eosin staining. (B) Elastica-Masson Goldner staining. Intimal thickening (white arrow) and an increase in elastic fibers (black arrow) are shown. Giant cells were not observed. Scale bar: 100 μm.
not simply a reflection of an inflamed superficial temporal artery, cytokines and other inflammatory mediators participate in the propagation of the inflammatory processes from the artery (9). However, given that temporal arteritis may result in ipsilateral hypertrophic pachymeningitis, we propose that dural inflammation may be the cause of an ipsilateral headache associated with temporal arteritis. The present case provides additional evidence in support of this hypothesis. Scalp tenderness, which possibly reflects inflammation in the affected temporal arteries or subcutaneous tissue, is known to arise in approximately half of the patients with this disease (10). Conversely, approximately half of the patients do not have scalp tenderness, and inflammation in the dura mater, rather than in the extracranial structures, might explain the headache in some of these patients.

Our patient presented with ptosis, a relatively rare symptom in temporal arteritis. Temporal arteritis can involve the extraocular muscles including the levator palpebrae, leading to ptosis and diplopia, and its etiology is considered to involve the vasa nervorum to cranial nerves III, IV and VI or ocular muscle ischemia (2, 11-13). Although the ophthalmic artery supplying these cranial nerves and extraocular muscles is usually a branch of the internal carotid artery, the ophthalmic artery anastomoses with branches of the external carotid system (14), and in some patients, it arises from the middle meningeal artery (MMA), a branch of the external carotid artery (15). These vascular anatomical structures could explain why temporal arteritis, which involves the external carotid system, could result in diplopia and ptosis.

As discussed above, there appears to be two kinds of headache in temporal arteritis: vascular pain from temporal arteritis itself and meningeal pain from secondary hypertrophic pachymeningitis. This difference in headache origin results in different clinical characteristics. A vascular headache is usually characterized by throbbing and pulsating pain in the scalp, especially in the swollen superficial temporal artery (1, 9), while a meningeal headache has no tenderness in the temporal artery itself (4, 6). Thus, clinicians should note that the absence of tenderness in the temporal arteries does not necessarily rule out the diagnosis of temporal arteritis, and in such cases, the headache might result from hypertrophic pachymeningitis.

The coexistence of temporal arteritis and hypertrophic pachymeningitis has rarely been described, possibly because gadolinium-enhanced brain MRI is not performed in most patients with temporal arteritis. More detailed examinations including gadolinium-enhanced MRI for temporal arteritis would enable us to determine more cases of hypertrophic pachymeningitis, which potentially underlies the patients’ headache.

The authors state that they have no Conflict of Interest (COI).

References