CASE REPORT

Reversible Distension of the Subarachnoid Space around the Optic Nerves in a Case of Idiopathic Hypertrophic Pachymeningitis

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Idiopathic hypertrophic pachymeningitis (IHP) is a chronic inflammatory disease of unknown cause. We report a case of IHP with bilateral distended subarachnoid space (SAS) of the optic nerves and unilateral visual disturbance. We observed marked amelioration of magnetic resonance (MR) imaging findings after initiation of treatment with prednisolone. This radiological finding implicates optic nerve sheath involvement that affects cerebrospinal fluid (CSF) dynamics around the optic nerve.

Keywords: cerebrospinal fluid dynamics, idiopathic hypertrophic pachymeningitis, optic nerve, subarachnoid space

Introduction

Hypertrophic pachymeningitis (HP) is a rare chronic inflammatory disorder that causes localized or diffuse thickening of the dura mater.1,2 Various conditions associated with HP include infections, systemic autoimmune/vasculitic disorders, and neoplasms. Some cases in which the contributing pathogenesis of HP is unknown are currently diagnosed as “idiopathic” HP (IHP).2 IHP is clinically characterized by headache and multiple cranial polyneuropathies. Neuro-ophthalmic complication is the second most common symptom after headache in IHP,2 but the mechanism of visual impairment remains to be clarified. Pathological features of IHP include fibrotic change and inflammatory cell infiltrates of lymphocytes, plasma cells, and epithelioid histiocytes.2 Abnormal magnetic resonance (MR) imaging findings of dural enhancement after administration of gadolinium are a neuroradiological hallmark of HP.1,2 We report a first case of IHP with the notable finding of bilateral enlarged subarachnoid space (SAS) of the optic nerves.

Case Report

A 57-year-old man visited an ophthalmologist because of left visual disturbance with no other symptoms. Goldmann visual field testing indicated left central scotoma, and fundoscopic examination showed no abnormality. Ischemic optic neuropathy was diagnosed, but his visual impairment gradually worsened. Brain MR imaging after administration of gadolinium performed one month after symptom onset demonstrated thickened and enhancing dura mater at the falx cerebri and cranial base, leading to a diagnosis of HP.

The patient was admitted to our hospital for further examination. Physical examination showed no abnormality. Neurological examination revealed no sign of cranial nerve dysfunction except for left optic nerve involvement. No finding of cerebellar, pyramidal, or extrapyramidal signs was observed. Laboratory findings showed no abnormality in peripheral blood cell counts, serum chemistry, or urinalysis, other than mild elevation of alkaline phosphatase and γ-glutamyltransferase. The following laboratory tests were all normal: serologic test for syphilis, anti-human T-cell leukemia virus type 1 (HTLV–I) antibody, antinuclear antibody (ANA), anti-Ro (SS-A) and anti-La (SS-B) antibodies, myeloperoxidase-specific anti-neutrophil cytoplasmic antibody (MPO-ANCA), proteinase 3-
specific ANCA (PR3-ANCA), anti-cyclic citrullinated peptide antibody (anti-CCP antibody), serum levels of IgG4, angiotensin converting enzyme (ACE), and soluble interleukin–2 receptor. Lumbar puncture showed an opening pressure of 125 mm H2O. Cerebrospinal fluid (CSF) examination revealed neither elevated protein nor pleocytosis. CSF cultures for bacteria, fungi, and acid-fast bacillus were all negative. Polymerase chain reaction (PCR) to detect *Mycobacterium tuberculosis* and antigen of *Cryptococcus neoformans* were also negative in the CSF.

The patient underwent brain MR imaging examination using a 1.5-tesla unit (Signa HDxt, General Electric Medical Systems, USA) 6 weeks after onset. T1-weighted images with fat suppression after gadolinium administration (T1WI-Gd) showed uniformly enhanced thickening of the falx cerebri and the dura mater at the cranial base extending into the thickened optic nerve sheath (Fig. 1). Coronal sections on short inversion time inversion recovery (STIR) images revealed bilateral expansion of the SAS surrounding the optic nerve at the intraorbital ("donut configuration") (Fig. 2) but not canaliculc region. We found neither localized hypertrophic dura compressing the optic nerve directly nor dural venous sinus occlusion causing central retinal vein occlusion.

The patient was diagnosed with IHP and prescribed one mg/kg daily prednisolone. Two weeks after initiation of treatment with prednisolone, brain MR imaging on T1WI-Gd showed marked improvement of the thickened dura (Fig. 1) and bilateral expansion of the intraorbital SAS (Fig. 2), but visual field and acuity tests showed only partial amelioration.

**Discussion**

This is the first report to show reversible bilateral distended SAS of the optic nerves in a case of IHP. Enlarged SAS has been reported in intracranial hypertension and in local pathological conditions that affect CSF dynamics around the optic nerve, such as optic nerve sheath meningioma, Tolosa-Hunt syndrome, and optic neuritis. In our case,
local factors seemed to cause the dilated SAS. No findings suggested intracranial hypertension; papilloedema was not detected, and lumbar puncture revealed normal opening pressure.

The most plausible explanation of the distended perioptic SAS is obstruction between the SAS of the optic nerve and the chiasmal cistern. Hickman and associates observed enlarged SAS in 45.5% of patients with optic neuritis6 and attributed dilatation of the optic nerve sheath mainly to mechanical factors rather than local inflammation based on its occurrence even without enhancement of the sheath. Histological examination of the meninges of the human optic nerve showed the perioptic SAS most narrow in the intracanalicular region. In view of the anatomical features and our observation of notable SAS dilation at the intraorbital region, a check valve phenomenon at the intracanalicular region may have caused local CSF entrapment.

Another possible explanation is impaired CSF absorption. The optic nerve is covered by the meninges and surrounded by CSF throughout its entire length. Owing to the cul-de-sac structure, CSF dynamics between the intracranial SAS and the SAS of the optic nerve remain to be determined in detail. Killer and colleagues reported that lymphatics present in the optic dura offers an outflow pathway of CSF at the optic nerve sheath. In our case, MR imaging demonstrated enhancing dura mater along with the optic nerve sheath. Therefore, inflammation and/or thickening of the dura mater may have impaired CSF resorption, resulting in dilatation of the SAS. The above 2 hypotheses may suggest that the reversibility of the enlarged perioptic SAS after initiation of treatment with prednisolone reflects relief of mechanical obstruction or amelioration of an inflammatory process.

At present, the mechanism underlying visual dysfunction in HP is unknown. Direct compression, spread of inflammation from adjacent thickened meninges, and secondary ischemic damage of the optic nerve are the candidate mechanisms.6 Our patient experienced unilateral visual dysfunction despite bilateral dilated SAS of the optic nerves. Interestingly, ophthalmic examination revealed central scotoma, a characteristic visual disturbance that suggested ischemic damage as the likely pathogenesis of optic nerve involvement. Though the exact reason for our patient’s unilateral visual disturbance is uncertain, the severity of impairment of the vascular system penetrating the dura mater and/or the difference in vascular anastomosis may contribute to the pathogenesis of optic nerve impairment.

In conclusion, the noteworthy MR imaging finding of bilateral distended SAS of the optic nerves was demonstrated in a case of IHP, a rare clinical entity. However, we should consider this disease as a differential diagnosis, even in patients with isolated neuro-ophthalmic complications.

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