Interval progression of idiopathic hypertrophic pachymeningitis

Nitin K. Sethi, MD

A 38-year-old right-handed woman first presented at age 16 with compressive myelopathy. A C7-T2 decompressive laminectomy was performed after imaging revealed diffuse thickening of spinal and intracranial dura. Biopsy revealed lymphocytic infiltration of the dura (infiltration of the dura by a population of monomorphic small cells with scant cytoplasm and dense nuclei) with no features to suggest neoplasia. No giant cell, granuloma, or microorganisms were seen. Mitoses were not seen and negligible nuclear atypia was reported. Immunohistochemical staining for lymphocyte common antigen was positive in most of the inflammatory cells. Glial fibrillary acidic protein and neuron-specific enolase staining was not seen. Taking her history of rheumatoid arthritis, glomerulonephritis, and Hashimoto thyroiditis into consideration, after an exhaustive evaluation, a diagnosis of idiopathic hypertrophic pachymeningitis was made. She was initially maintained on prednisone and later methotrexate for about 10 years but self-discontinued it in 2005 as she had no symptoms. In February 2009, brain MRI was carried out to evaluate for occipital headaches (figure 1). Amitriptyline resulted in headache relief and she was lost to follow-up but presented again

Figure 1 Brain MRI with contrast (2009)

Sagittal T1-weighted imaging sequence shows diffuse thickening of cranial dura mater.

Department of Neurology, New York–Presbyterian Hospital, Weill Cornell Medical Center, New York.

Funding information and disclosures are provided at the end of the article. Full disclosure form information provided by the author is available with the **full text of this article** at Neurology.org/cp.

Correspondence to: sethinitinmd@hotmail.com

Figure 2 Brain MRI with contrast (2013)

Sagittal T1-weighted imaging sequence shows interval progression of pachymeningeal thickening with mass effect on the inferior aspect of the fourth ventricle with slight protrusion of the cerebellar tonsils through the foramen magnum, ventriculomegaly, and hydrocephalus.

in March 2013 with headaches. Neurologic examination was nonfocal, funduscopy was normal, and there were no myelopathic signs. Interval increase and worsening of the diffuse pachymeningeal thickening with enhancement involving the posterior fossa and extending into the cervical canal along with mass effect on the inferior aspect of the fourth ventricle with slight protrusion of the cerebellar tonsils through the foramen magnum was documented on MRI. The progressive ventriculomegaly was consistent with hydrocephalus (figures 2 and 3). Cervical spine MRI revealed exaggerated lordosis with diffuse thickening and enhancement of the dura causing mass effect and compression of the cervical cord up to T2-T3 level (figure 4). Her headaches were believed to be congruous with either chronic tonsillar herniation or

Figure 3 Brain MRI with contrast (2013)

Coronal T1 with contrast imaging sequence shows interval progression of pachymeningeal thickening with mass effect on the inferior aspect of the fourth ventricle with slight protrusion of the cerebellar tonsils through the foramen magnum, ventriculomegaly, and hydrocephalus.

Figure 4 Cervical spine MRI (2013)

Sagittal proton density imaging sequence of cervical spine shows lordosis with diffuse thickening and enhancement of the dura causing mass effect and compression of the cervical cord up to T2-T3 level.

progressive noncommunicating hydrocephalus. While endoscopic third ventriculostomy was entertained as a therapeutic option, since on examination there was no objective evidence of raised intracranial pressure (i.e., papilledema, altered cognition, long tract signs), it was decided to follow her with surveillance imaging and serial ophthalmologic examinations. She is currently asymptomatic on prednisone. Idiopathic hypertrophic pachymeningitis is an uncommon disorder that causes localized or diffuse thickening of the cranial and spinal dura mater. Untreated, the clinical course is marked by severe headaches, progressive neurologic deterioration, and vision loss. Though steroid-responsive, clinical manifestations frequently recur when steroid and immunosuppressive agents are tapered off.¹

REFERENCE

1. Kupersmith MJ, Martin V, Heller G, Shah A, Mitnick HJ. Idiopathic hypertrophic pachymeningitis. *Neurology* 2004;62:686–694.

STUDY FUNDING

No targeted funding reported.

DISCLOSURES

N. Sethi serves as an Associate Editor of the *Eastern Journal of Medicine*. Full disclosure form information provided by the author is available with the **full text of this article at Neurology.org/cp**.