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Idiopathic Pachymeningitis Presenting with Progressive Sensorineural Hearing Loss, Tinnitus and Confusion

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The patient was a 74-year-old female who presented with progressive left hearing loss, nonpulsatile tinnitus, and confusion. She denied headache, visual symptoms, and vertigo. She had a normal neurotologic examination. Pure tone audiometry showed left low frequency sensorineural hearing loss (SNHL) and a bilateral symmetric high frequency SNHL, with word recognition of 100% right, 46% left (Fig. 1). Serologic studies revealed a nonspecific monoclonal gammopathy. Other serologic tests were negative, including CBC, ANA, Lyme titer, syphilis, angiotensin converting enzyme, anti-double stranded DNA and ANCA antibodies. Cerebrospinal fluid testing revealed normal cytology with a mild increase in protein. Computed tomography of the abdomen and chest was normal, as was bone marrow biopsy. Magnetic resonance imaging (MRI) demonstrated diffuse smooth dural thickening and enhancement consistent with idiopathic hypertrophic pachymeningitis (IHP) (Fig. 2).

The patient was initially treated with oral prednisone with minimal improvement. She then received IV methylprednisolone infusions every two weeks for two months with improvement in left hearing, tinnitus, mental status, and dural enhancement on MRI. Six months following initial presentation, she showed no further progression of her symptoms while off steroid therapy.

DISCUSSION

IHP is a rare inflammatory disease that causes thickening of the dura mater (1, 2) with sparing of the leptomeninges and brain.(3) The peak age of presentation is in the sixth decade; and the male-to-female ratio is 1.2:1(1). Symptoms include headache often accompanied by multiple cranial neuropathies, ataxia, diplopia, and loss of visual acuity (1, 2). Less commonly, memory loss and confusion can occur (1, 2). Cranial nerve palsies result from compression of nerve roots by hypertrophic dura (2). The vestibulocochlear nerve is

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the most commonly affected cranial nerve. SNHL is thought to occur due to pressure from affected dura within the internal auditory canal (4). Other etiologies for SNHL from IHP have been hypothesized including cochlear damage due to compression of internal auditory artery from thickened dura and autoimmune-induced damage (4). Previously reported cases found progressive SNHL with disproportionately poor word recognition scores and tinnitus accompanied by imbalance and vertigo (3–5).

IHP is a diagnosis of exclusion (1). Due to the broad differential of hypertrophic pachymeningitis, full workup includes extensive serologic studies, CSF studies, and contrast-enhanced MRI of the brain. Findings should not be explained by trauma, neoplasms, systemic vasculitides, fibrosclerotic/autoimmune disease, or infection (Lyme, syphilis, tuberculosis, and fungi). In a minority of patients, ANA, ANCA, RF, ESR and CRP levels may be mildly elevated. CSF protein is elevated in 50–60% of cases (2). Directed dural biopsy may be indicated to exclude other causes of hypertrophic meningitis, (1) especially in cases of rapid disease progression or those that are refractory to treatment (3).

Gadolinium-enhanced MRI is used to evaluate localization, extent, and pattern of abnormal dura and to monitor progression (2). The dura is iso- or hypointense and thickened with avid dural contrast enhancement on T1-weighted images. On T2-weighted images the dura is hypointense (1, 4). In over 80% of cases symptoms are directly associated with the location of the abnormal dura (1, 2). Most cases display diffuse linear falcine, tentorial, and clival enhancement extending to the posterior fossa. 25% have parasellar and cavernous sinus wall enhancement (3). Dural involvement may be linear or nodular, and both patterns may be seen in the same patient (2). Well-defined hyperintense T2-weighted borders and homogenous enhancement on T1-weighted images frequently correlate with short clinical course and best prognosis. Despite improvement in symptoms, abnormal dural enhancement on MRI can persist, possibly due to progression of disease to fibrotic stages (2). The imaging differential includes sarcoidosis, intracranial hypotension, infectious pachymeningitis and dural metastases. Dural metastases demonstrate nodular enhancement.

High dose corticosteroid therapy is the initial treatment of choice, followed by immunomodulators such as cyclophosphamide, which may allow steroid taper (2). To avoid long-term side effects, pulse steroid therapy has been proposed (1). Steroids typically improve hearing, with word recognition improving more markedly than pure tone thresholds (4, 5). Surgery has been advocated for localized nodular IHP. Procedures such as optic nerve decompression and CSF shunting are strongly recommended to avoid catastrophic complications such as blindness (3, 4). Favorable prognostic features include diffuse dural enhancement and improvement in symptoms following treatment with corticosteroids (1). Despite response to treatment, IHP remains a chronic disease hallmarked by a progressive, relapsing and remitting clinical course.

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Figure 1.

Pure tone audiograms from 2009 through 2012, demonstrating progressive hearing loss in the left (A) and right (B) ears. At baseline (solid black line), the patient had a left 30 dB SRT and 100% WR, and a right SRT of 20 dB and WR 100%. After onset of symptoms (dotted black line), the patient had a left 65 dB SRT and 46% WR and a right 35dB SRT and 100% WR. Following treatment (grey line), the left SRT was 50 dB and WR 96%; however, right hearing had worsened (60 dB SRT, 80% WR).

Figure 2. Axial T1-weighted contrast-enhanced MRI of brain from prior to treatment (A) demonstrating initial enhancement of dura of the internal auditory canals left > right (arrowheads) prior to treatment. Following pulse steroid treatment (B), a significant decrease in dural enhancement is noted, particularly in the internal auditory canals (arrows).