Hypertrophic pachymeningitis in the context of the treatment of polymyalgia rheumatica

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DESCRIPTION
A woman in her mid-70s with a history of polymyalgia rheumatica (PMR) and on oral prednisolone of 2.5 mg per day presented with a 4-week history of progressive weakness with diffuse myalgia. On admission, neck stiffness and tenderness on the posterior neck, shoulders and thighs were appreciated. Notably, pupils were dilated and non-reactive to light bilaterally. Bilateral visual acuities and auditory functions were impaired. Horizontal ocular movements were full and smooth, but the vertical gaze was impaired. Bilateral papilledema was noted. Motor and sensory functions were intact in all modalities. Deep tendon reflexes were diffusely absent. Her gait was short-stepped and ataxia was noted. Her higher cerebral function was almost intact, except for working memory.

Laboratory findings included elevated erythrocyte sedimentation rate of 3–15 mm/1 h (reference range 3–15) and C reactive protein of 146.6 mg/L (0.0–1.4). Cerebrospinal fluid (CSF) analysis revealed opening pressure of 32 cmH2O (5–20), protein level of 156 mg/dL (15–45) and white blood cell counts of 21 (mono 19, poly 2) ×10^6/L cells/mm^3 (0–5). Gram stain and acid-fast stain of CSF detected no organism, and cultures were negative for bacteria, fungus or mycobacterium. No malignant cells were detected on the cytopathology.

Gadolinium-enhanced brain MRI showed diffuse thickening of the entire meninges with late gadolinium enhancement without evidence of tumour or abscess (figure 1A,B: horizontal; C: sagittal; D: coronal). Although there was a finding of enlarged supratentorial ventricles without cerebral cortical atrophy, the aqueduct was not obviously occluded. Serum ACE, immunoglobulin G4 (IgG4), antinuclear antibody and antineutrophil cytoplasmic antibody were negative. She was diagnosed as idiopathic hypertrophic pachymeningitis (HP) and treated with three courses of intravenous pulse steroid therapy, followed by maintenance therapy with oral prednisolone and cyclophosphamide. Although a follow-up imaging study would have been ideal, we did not conduct that. At 3 months of follow-up, her myalgia, weakness, short-stepped gait and hearing loss improved, while her sight remained unchanged.

HP is a disease associated with chronic meningeval inflammation and dural thickening, typically resulting in progressive cranial nerve palsy, headache and cerebellar dysfunction. The diagnosis of HP is often difficult, because of its rarity and its non-specific neurological symptoms and signs, as seen in this case.

HP can be classified as either idiopathic or secondary. The causes of secondary HP encompass tumours, infections, IgG4-related diseases and inflammatory conditions including rheumatoid arthritis, sarcoidosis and vasculitis. The causal relationship between HP and PMR is not clear, but the reported association between rheumatic disease and HP may at least suggest the association between HP and PMR. The diagnosis was made with gadolinium-enhanced contrast MRI in this case, as described in the past reports as useful. In retrospect, the learning point from this case is that in patients with rheumatic disease and multiple cranial nerve abnormalities of unknown causes, HP should be listed as a differential diagnosis.
Images in... a differential diagnosis. Besides, while obvious obstruction was not observed in this case, communicating hydrocephalus secondary to HP might have existed based on the image findings, which could explain the patient’s symptoms, such as gait disturbance.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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