Hypertrophic Pachymeningitis Accompanying Scleritis

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An elderly woman undergoing treatment for organizing pneumonia and Graves’s disease complained of severe headache after a reduction of her oral prednisolone dose (5 mg/day to 2.5 mg/day). Although her headache resolved spontaneously, it recurred with severe ophthalmalgia and she showed bloodshot eyes. Enhanced MRI revealed a thickened dura mater with a contrast effect (Picture A and C: coronal, Picture B: sagittal), leading to a diagnosis of hypertrophic pachymeningitis. The patient was positive for myeloperoxidase anti-neutrophil cytoplasmic antibodies and her immunoglobulin G4 level was elevated. Moderate scleritis was ophthalmologically evident (Picture D). Intravenous methylprednisolone (500 mg for 3 days) followed by oral prednisolone (30 mg/day) improved her headache, eye pain, and MRI findings.

The typical symptoms of hypertrophic pachymeningitis include headache with or without intracranial neurologic manifestations such as cranial neuropathy and cerebellar dysfunction (1). Despite the rarity, this disease combination should be kept in mind when we encounter patients with headache and eye pain (2).

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References


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