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CASE REPORT

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A case of suspected MPO-ANCA-related hypertrophic pachymeningitis with atypical presentation

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Abstract

A 66-year-old man presented to our emergency department with acute consciousness disorder, headache, and fever. Initial laboratory data indicated a high level of inflammatory reaction. Enhanced magnetic resonance imaging revealed a thickening of the right cranial dura mater on T1-weighted images. We presumed the patient was diagnosed as having myeloperoxidase antineutrophil cytoplasmic antibody (MPO-ANCA)-related hypertrophic pachymeningitis (HP). Acute impaired consciousness without focal signs is an unusual presentation of HP. This treatable disease should be considered as a differential diagnosis when a patient presents with drowsiness and headache.

KEYWORDS

consciousness disorder, headache, hypertrophic pachymeningitis, MPO-ANCA

1 | INTRODUCTION

Hypertrophic pachymeningitis (HP) is a rare inflammatory disorder demonstrating local or diffuse thickening of the intracranial or spinal dura mater. This report describes an atypical case of myeloperoxidase antineutrophil cytoplasmic antibody (MPO-ANCA)-related HP along with a literature review.

2 | CASE PRESENTATION

A 66-year-old man presented to our emergency department with impaired consciousness and headache, which had worsened over the previous 2 days. He suffered from chronic headaches, and he had been treated for polymyalgia rheumatica. A neurologist prescribed 10 mg of prednisolone, which he had been taking for 1 year. He had no history of chronic sinusitis or otitis media.

He was restless and unable to follow our instructions. On physical examination, his Glasgow Coma Scale was 14 (E4V4M6), blood pressure was 105/80 mm Hg, heart rate was 113 beats per minute, body temperature was 38.8°C, and a respiratory rate was 20 breaths per minute. His cardiopulmonary and gastrointestinal findings were normal. On neurological examination, his patellar and Achilles tendon reflexes were bilaterally hyperactive. His biceps, triceps, and radial tendon reflexes were normal. His Chaddock reflexes were bilaterally positive, cranial nerve examinations were normal, and he did not have a stiff neck. Initial laboratory data indicated leukocytosis (11,200/μL) and increased C-reactive protein (1.65 mg/dL). Other data, including glucose, hepatobiliary enzyme, creatinine, and electrolyte levels (sodium, potassium, and calcium) were within normal limits. His blood gas analysis showed slight respiratory alkalosis due to mild hyperventilation. His cerebrospinal fluid was clear, and the opening cerebrospinal fluid pressure was 250 mH_2O. His cell count was 2/μL, total protein was 74 mg/dL, cerebrospinal fluid-to-serum glucose ratio was 0.6, and gram-staining results were negative. Noncontrasted computed tomography of the head revealed normal findings. Further evaluation concerning his impaired consciousness and headache was required. Therefore, we consulted a neurologist and ordered enhanced brain magnetic resonance imaging (MRI). A thickness of the right cranial dura mater was revealed on T1-weighted MRI (Figure 1). No other venous or parenchyma abnormalities were shown on MR venography and diffusion-weighted images and fluid attenuation inversion

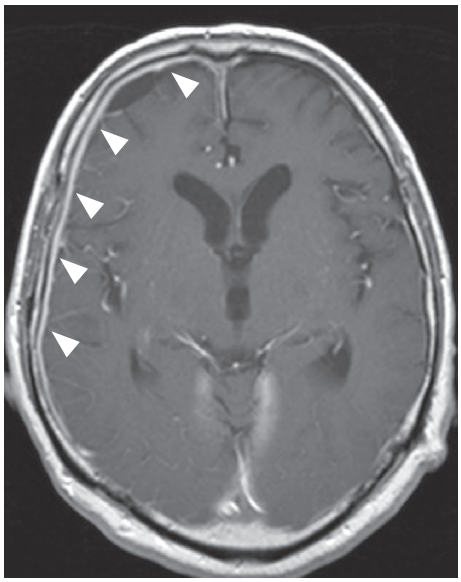

FIGURE 1 Gadolinium (Gd)-enhanced T1-weighted magnetic resonance imaging (MRI) of the brain showing a thickening of the right cranial dura mater

recovery images, respectively. HP was suspected, and he was hospitalized in the neurology ward for a thorough examination and treatment. He was treated with 1000 mg of intravenous methylprednisolone daily for 5 days, and his impaired consciousness and headache improved from the day following treatment. After that 50 mg of oral prednisolone per day was administered followed by gradual tapering to 10 mg per day without relapse of symptoms. Later, additional laboratory data revealed a high MPO-ANCA level (16 U/mL). Tests for PR3-ANCA, IgG4, and other autoantibodies, including antinuclear antibody, were all negative. We finally presumed the patient was diagnosed as having MPO-ANCA-related HP.

3 | DISCUSSION

Hypertrophic pachymeningitis is a rare inflammatory disorder, characterized by intracranial or spinal dura mater thickening. Most cases are idiopathic, and secondary HP can be associated with miscellaneous diseases such as ANCA-related disease, rheumatoid arthritis, IgG4-related disease, syphilis, tuberculosis, and cancer.

Headache is the most common symptom (71.1%).[1] Neurological findings, including cranial nerve deficits, are also common (62.3%).[1] Two or more cranial nerves are usually affected, and ophthalmological and otological symptoms are frequently presented. Enhanced brain MRI is the most useful imaging technique for the diagnosis of HP, and it can exclude other differential diseases as required.

We excluded common causes of acute consciousness disorder including hypo/hyperglycemia, abnormal electrolytes, stroke, and infectious meningitis. Major causes of secondary HP including PR3-ANCA and IgG4 related HP were also excluded. We consider autoimmune encephalopathy and relative adrenal failure are the most difficult differential diagnoses. Chan[2] reported a case of carcinomatous HP associated with metastatic breast carcinoma. Our case did not have such a progressive carcinoma, but we measured no antineuronal antibodies. Our case showed no abnormalities of glucose, electrolytes, and eosinophil level which suggest relative adrenal failure. But we could not completely rule out because we did not measure cortisol level. These are our limitation of diagnosis.

The MRI findings of our case showed no signs of parenchymal abnormality. As far as we could search, previous studies on HP with impaired consciousness have reported some kind of parenchymal abnormality on MRI findings. For example, Yokoseki et al.[3] demonstrated that all patients with PR3-ANCA-related HP and a consciousness disorder had brain parenchymal involvement on MRI. Saito et al.[4] also reported a case of MPO-ANCA-related HP and cerebral venous thrombosis with a consciousness disorder.

Our case had a vague clinical presentation, indicating that there may be instances where HP is overlooked as a diagnosis.

4 | CONCLUSION

We reported an atypical case of MPO-ANCA-related HP that presented with an acute consciousness disorder without cranial nerve deficits. We should consider this treatable disease as a differential diagnosis when a patient presents with drowsiness and headache.

CONFLICT OF INTEREST

The authors have stated explicitly that there are no conflicts of interest in connection with this article.

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