Sir,

Hypertrophic pachymeningitis (HP) is a comparatively rare disease characterized by marked inflammatory hypertrophy of the dura mater, which can be classified into hypertrophic cranial pachymeningitis (HCP) and hypertrophic spinal pachymeningitis (HSP) depending on the location of the lesion. Most of the cases involved the intracranial dura mater, whereas cases of HSP were rarely reported, especially when the spinal cord was involved. To our knowledge, this is the first case of antineutrophil cytoplasmic antibody (ANCA) - associated HSP presenting with longitudinally extensive transverse myelitis.

A 65-year-old man was admitted to our department in October 2018 complaining of a 6-month history of low back pain, numbness, and weakness of both lower limbs over 4 months. The man first visited other hospital; there were no characteristic findings about magnetic resonance image (MRI) of his head, but MRI of spine showed thickening of spinal cord at T8-L2 level. Thus, the patient was diagnosed as myelitis, and his symptoms were improved significantly by treating with prednisolone. However, the patient suffered a relapse when he reduced the oral prednisone to 10 mg/day. This time, he was admitted to our hospital. The patient had hypertension and diabetes, with no history of trauma, tuberculosis, or any other diseases.

On admission to our department, he was found to have Grade 4/5 power in bilateral lower limbs and all modalities of sensation below xiphoid plane decreased apparently. Laboratory data were as follows: The erythrocyte sedimentation rate (ESR) was 90 mm/h; hypersensitive C-reactive protein level (hsCRP) was 71.1 mg/L; and c-ANCA was positive. Complete blood count, urinalysis, fecal routine, liver function tests, renal function tests, electrolyte, coagulation profile, myocardial enzyme, tumor markers, rheumatic profile, and thyroid tests were all within the normal ranges. Proteinase-3(PR3)-ANCA, myeloperoxidase (MPO)-ANCA, and p-ANCA were all negative.

He also underwent lumbar puncture. Within 24 h, his weakness markedly worsened with complete loss of power (Grade 0/5) in both lower limbs. Furthermore, he developed urinary retention and had to be catheterized. The examination of cerebrospinal fluid (CSF) revealed slight turbidity, yellow cerebrospinal fluid, and the CSF pressure was 80 mmHg. The total protein level of CSF was markedly elevated to 37 g/L (0.15–0.45) with the cell count of 2 × 10^6 per L (Leukocyte 2). The cytological examination of CSF showed no malignant cells, and examinations of bacterial, fungal, and mycobacteria were all negative in CSF. The highly specific and sensitive serological biomarkers of neuromyelitis optica (NMO) spectrum disorders such as oligoclonal banding (IgG-OB), NMO antibody, MAP antibody, and MOG antibody investigation were all negative both in the sera and CSF. A gadolinium (Gd)-enhanced MRI scan of spinal cord demonstrated thickening and enhancement of the soft and dura mater, especially at the level of T4 and below [Figure 1a and b]. The T2-MRI of the spine showed swelling of the spinal cord at the level of T4-T12 and thickening of soft tissue around the spinal canal at the level of T4 and below [Figure 1c]. The electrocardiography and CT scan detected no remarkable findings.

Then, a diagnosis of ANCA-associated HSP was made treated with pulse intravenous methylprednisolone (500 mg daily) for 3 days and followed by oral prednisone (50 mg/day) with gradual tapering of the dose. His symptoms improved with muscle strength in the lower limbs recovering to grade 4/5 and the level of sensation disturbances lowering to umbilicus plane. His urination also recovered. ESR and hsCRP decreased to 70 mm/h and 34.2 mg/L, respectively. Then, the patient was discharged from our hospital; lumbar puncture and spinal MRI were recommended for follow-up after 3 months.

HSP is a rare inflammatory disorder characterized by local or diffuse thickening of the spinal dura mater. A variety of conditions can cause HSP, including syphilis, tuberculosis, fungal infection, neoplastic diseases, trauma, and autoimmune diseases such as ANCA-associated systemic vasculitis (AASV), IgG4-related disease, sarcoidosis, or being labeled idiopathic in the absence of an identifiable cause. A nationwide survey of HP from Japan showed that idiopathic
HP was most frequent (accounts for 44%), followed by ANCA-associated HP (accounts for 34%). From clinical symptoms, laboratory examinations and imaging findings, the patient, in this case, can be excluded from infection, neoplastic diseases, NMO spectrum disorders, and systemic lupus erythematosus et al., then ANCA-associated with HSP was first considered.

The main symptoms of HSP were back pain, sensory/motor disturbances, and even paraplegia resulting from the compression of adjacent spinal cord.[2] The contrast-enhanced MRI of spinal dura mater was reported to be the most valuable diagnostic imaging technique, and the typical imaging findings were iso- or low-intense signal on T1-weighted images and lower-intense signal on T2-weighted images which mostly affect thoracic vertebral levels in a consecutive or discrete manner and can be markedly enhanced after injection of the contrast media.[4] The MRI findings in our case were consistent with the imaging features of HSP and previous reports.[2,5] There was also some difference between our case and previous reports. The T2-MRI of the spine in our case showed the spinal cord was swelling. For all we know, this was the first case report of a patient with ANCA-associated HSP involving longitudinally extensive transverse myelitis (LETM). LETM is generally taken to mean myelitis that extends over a continuous lesion at least 3 vertebral segments in length;[6] the most common cause of LETM was NMO. Tobin et al.[7] discussed the differential diagnosis in patients presenting with LETM in detail. However, ANCA-associated HSP was not mentioned in that article. Our case suggested that ANCA-associated HSP may be a rare cause of LETM. The exact mechanism of transverse myelitis in ANCA-associated HSP remains elusive. Yokoseki et al.[8] proposed that ANCA-associated HP might be a limited neurologic form of ANCA-associated systemic vasculitis.

In conclusion, HSP is a rare disease, especially when presented with transverse myelitis, making it difficult for clinicians to consider this disease in the first place. We strongly suggest that patients diagnosed with transverse myelitis also should be screened with ANCA test or other autoimmune assays and contrast-enhanced MRI, which is of great importance for clinical diagnosis.

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Conflicts of interest
There are no conflicts of interest.

Jun Shu, Chunhui Gu, Yiqing Ren, Wenshi Wei
Department of Neurology, Huadong Hospital Affiliated to Fudan University, No. 221, West Yan An Road, Shanghai, China

Address for correspondence: Dr. Wenshi Wei, Department of Neurology, Huadong Hospital Affiliated to Fudan University, No. 221, West Yan An Road, Shanghai - 200040, China.
E-mail: wenshiwei1999@163.com

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