

# Myeloperoxidase-Antineutrophil Cytoplasmic Antibody Positive Hypertrophic Spinal Pachymeningitis at the Cervicothoracic Junction: A Case Report

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A 49-year-old female was referred to our department with a history of neck pain and right upper extremity numbness lasting for three months. Gadolinium-enhanced magnetic resonance imaging (MRI) showed a diffusely enhanced epidural mass from C6 to T2 (Fig. 1, 2a). Surgical treatment was recommended, but she rejected it because her clinical symptoms were mild. Four months after the initial visit, she developed interstitial pneumonia and was admitted to the pulmonary medicine department of the regional medical center. Her myeloperoxidase-antineutrophil cytoplasmic antibody (MPO-ANCA) level was elevated to 14.7 IU/mL; she was treated with methylprednisolone (mPSL) pulse therapy and maintenance steroid administration. No immunosuppressive agents were used. After the treatment, her numbness and pain completely disappeared. MRI taken two months after the treatment showed a significant reduction of the mass (Fig. 2b).

However, three months after the treatment, she visited our emergency room complaining of acute back pain, numbness/pain on the ulnar side of the left arm, and clumsiness. She had difficulty standing or walking due to severe back pain. She had muscle weakness of MMT grade 4 in the biceps, triceps, iliopsoas, and quadriceps on both sides. She had no bladder or rectal disturbance. MRI showed regrowth of the mass from C5 to T3 (Fig. 2c). A blood test revealed a white blood cell count of  $15.5 \times 10^9/L$  and C-reactive protein at 16.4 mg/L. No definitive diagnosis could be made, but because of the rapid progression of myelopathy, we performed an emergent expansive laminoplasty from C7 to T1 with a

concurrent biopsy. Intraoperatively, we found diffusely thickened dura mater compressing the underlying spinal cord (Fig. 3). According to the intraoperative findings, clinical symptoms, and MRI findings, we made a diagnosis of hypertrophic spinal pachymeningitis (HSP). The pathological examination showed fibrosis with infiltration of inflammatory cells surrounding relatively large vessels, findings seen in epithelioid granuloma and vasculitis (Fig. 4). Electromyography showed peripheral neuropathy of both legs and the left arm, which are seen in polyneuritis resulting from vasculitis. She was diagnosed with granulomatosis with polyangiitis, one of the ANCA-associated systemic vasculitis, based on her history of interstitial pneumonia, peripheral neuropathy, and the pathological findings, as the cause of HSP.

With support from rheumatologists, she was treated with mPSL pulse therapy, intravenous cyclophosphamide (IVCY), and maintenance steroids. As IVCY-induced liver injury occurred, IVCY was then changed to rituximab 71 days after surgery. Her neurological symptoms improved, and she could walk using a cane at discharge. A follow-up MRI taken seven months after the surgery showed a decrease in the degree of dural thickening (Fig. 2d). Two years after the surgery, she could walk with no pain, and no findings were indicating a recurrence.

HSP is a rare disorder caused by chronic inflammatory hypertrophic fibrosis of the spinal dura mater. It is a spinal form of hypertrophic pachymeningitis first reported by Charcot and Joffroy in 1869<sup>1)</sup>. The present case was an



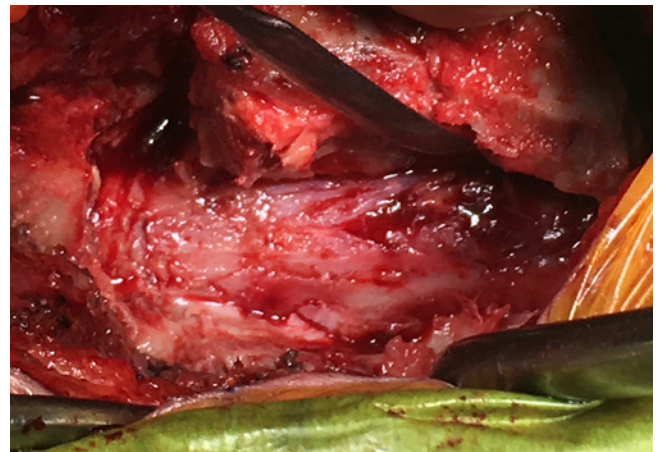
**Figure 1.** The spinal cord was compressed by a diffusely enhanced epidural mass (white arrow) from C6 to T2, with isointensity on a T1-weighted magnetic resonance imaging (MRI) and low intensity on a T2-weighted MRI.

- a: T1-weighted sagittal image.
- b: T2-weighted sagittal image.
- c: Gadolinium-enhanced sagittal image.

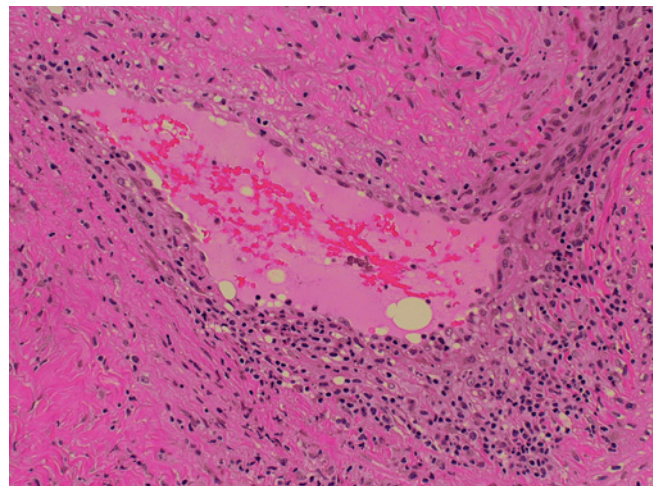


**Figure 2.** T2-weighted sagittal MRI of the cervical spine.

- a: The epidural mass (white arrow) compressed the spinal cord from C6 to T2 at the initial visit.
- b: Significant reduction of the epidural mass was observed after the methylprednisolone pulse therapy for interstitial pneumonia.
- c: Regrowth of the mass was observed from C5 to T3 at admission.
- d: The mass had reduced, and there was no recurrence seven months after surgery.



**Figure 3.** Intraoperative findings of the dura mater. The dura mater was found to be diffusely thickened and was compressing the underlying spinal cord.



**Figure 4.** Pathological findings of the surgically removed specimen. Hematoxylin and Eosin staining sections revealed fibrosis with infiltration of inflammatory cells, such as lymphocytes and plasma cells, surrounding relatively large vessels (original magnification  $\times 200$ ).

MPO-ANCA-related HSP. To the best of our knowledge, only six cases of MPO-ANCA positive HSP have previously been reported in the English literature<sup>2-6</sup>. Among seven cases, including this case, only two cases could diagnose HSP from MRI findings as it is difficult to differentiate HSP from other diseases, such as an epidural abscess or atypical spinal tumor, by preoperative MRI<sup>7</sup>. Biopsy of the dura mater is reported to be the standard diagnostic test<sup>2</sup>. Out of the seven cases, our case was the only case experiencing a short-term recurrence. To date, there is no established treatment for MPO-ANCA-positive HSP. All the past six cases were treated with mPSL pulse and immunosuppressants, but the present case was treated with only mPSL at first. This suggests that the administration of only steroids for MPO-ANCA-positive HSP may lead to a higher rate of recurrence.

The present case suggests the importance of suspecting

HSP in patients with an epidural mass. It is also important to consider ANCA-associated vasculitis as a cause of HSP and to examine the systemic symptoms, take blood tests, including ANCA and immunoglobulins, and take whole body contrast computed tomography.

**Conflicts of Interest:** The authors declare that there are no relevant conflicts of interest.

**Ethical Approval:** None. 20110141

**Author Contributions:** Ryo Ogaki wrote and prepared the manuscript. Eijiro Okada performed the surgery and supervised the manuscript. All of the authors had read, reviewed, and approved the manuscript.

**Informed Consent:** Before submission, informed consent was obtained by the patient.

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