A rare IgG4-related acute sclerosing pachymeningitis leading to spinal cord compression: diagnosis and treatment

Sirs,

Immunoglobulin G4-related disease (IgG4-RD) is an autoimmune inflammatory disorder characterised by the infiltration of IgG4-positive plasma cells, and it is recognised as a distinct clinical entity that can involve various organs, including the pancreas, bile ducts, kidneys, salivary glands, lymph nodes, aorta, nervous system, and retroperitoneum (1, 2). IgG4-RD typically presents as tumour-like lesions at the affected sites usually accompanied by elevated serum IgG4 concentrations, resulting in local obstruction and compression symptoms (1).

IgG4-RD is characterised by elevated serum levels of IgG4 and infiltration of IgG4-positive plasma cells in affected organs, and its pathogenesis is thought to be influenced by genetic factors, environmental triggers, and infectious stimuli (2).The histopathological features of this disease typically include non-specific lymphoplasmacytic infiltration and fibrosis, which can also be observed in various chronic inflammatory conditions such as systemic vasculitis and multicentric Castleman disease (2). As a result, it can be confused with malignancies, infections, or other immune-mediated diseases. Currently, the diagnostic and therapeutic criteria for IgG4-RD are largely based on consensus statements from domestic and international experts (1-4). The diagnosis of this condition primarily relies on the comprehensive assessment of histopathology, clinical manifestations, serology, and imaging findings.

Based on the international consensus guidance statement on IgG4-RD, the mainstream treatment approach remains to be steroids and immunosuppressants (1, 3, 5, 6). Glucocorticoids are considered the first-line therapy for treating IgG4-RD patients, and they not only alleviate symptoms but also provide effective prophylaxis against relapse through maintenance therapy with low-dose glucocorticoids (1, 5, 6). Immunosuppressants can serve as a substitute option for suboptimal steroid efficacy in medical practice (1, 5).

A 59-year-old previously healthy man presented to us with a 10-day history of abdominal pain. Two days after admission, he was unable to walk and both lower limbs were hypalgesic. Thoracic MRI showed a strip lump with a well-defined outline in the spinal canal at the T3-T6 level, compressing the spinal cord. A spinal haematoma evacuation procedure was performed, and postoperative pathology indicated a significant elevation of IgG4+ plasma cells (Fig. 1). The patient was diagnosed with IgG4-related pachymeningitis based on histopathological features.

The patient initially received methylprednisolone pulse therapy at a dose of 500 mg for 3 days. Subsequently, the methylprednisolone dosage was adjusted to 160 mg and then 80 mg, with treatments lasting for one week each, in combination with cyclophosphamide. After treatment, the most frequently used inflammatory markers including ESR and CRP decreased to the normal range. When the patient was discharged, the muscle strength in the both lower limbs had returned to grade 1/5. Follow-up at 3 months after discharge, the patient's condition was stable, and the muscle strength in the both lower limbs had returned to grade 2/5. Follow-up at 8 months after discharge, the muscle strength in the both lower extremities had returned to grade 3/5.

In conclusion, a multi-disciplinary team is highly advantageous in the diagnostic and therapeutic management of IgG4-RD, with the active involvement of rheumatology and pathology specialists being particularly pivotal. In cases where patients exhibit characteristic tissue morphological features or organ involvement suggestive of IgG4-RD, a comprehensive preoperative case discussion and differential diagnosis are imperative. Furthermore, performing an early biopsy of the involved lesion region is of paramount importance in confirming the diagnosis of IgG4-RD, excluding concomitant systemic neoplasms, and avoiding inappropriate surgical interventions that may lead to unnecessary iatrogenic damage. Regular follow-up visits are also crucial for early detection of disease progression, thereby mitigating the risk of organ damage and optimising patient outcomes.

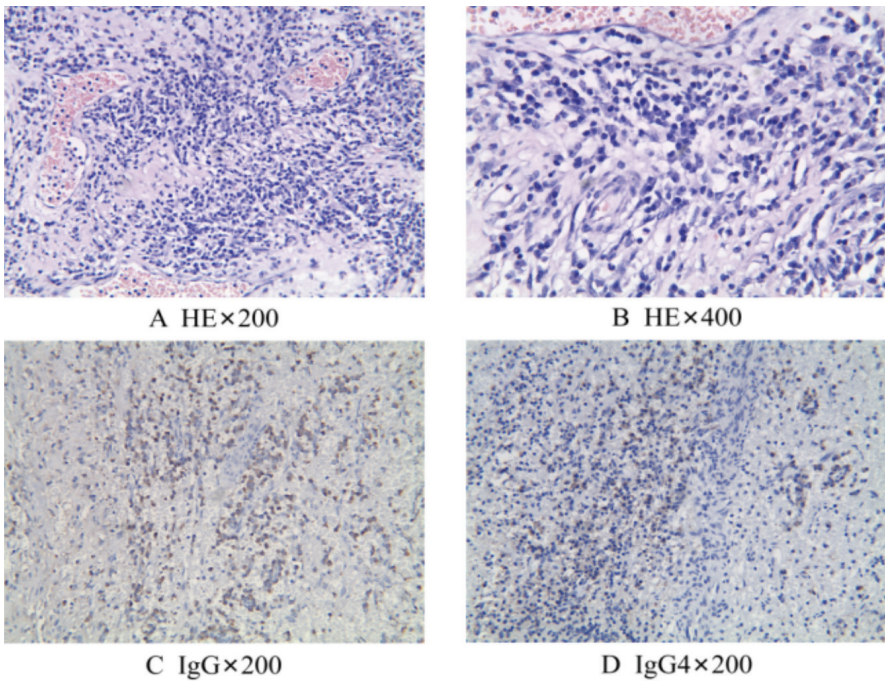

Fig. 1. Histopathological and immunostaining examination of the thoracic subdural lesion which was resected. Haematoxylin-eosin staining showed numerous plasma cell, lymphocytes, and neutrophils infiltrates. Plasma cells were positive for IgG (C) and IgG4 (D), and the IgG 4 / IgG ratio exceeded 40% with an average of more than 50 IgG 4 + plasma cells per high magnification.

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Competing interests: none declared.

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