eP-24

IgG4-related hypertrophic spinal pachymeningitis as a rare cause of cervical myelopathy: case report and an updated review of the literature.

Ahmet Parlak[1], Christian-Andreas Müller[1], Hans Clusmann[1], Tobias Schmidt[1], Ulf Bertram[1], Christian Blume[1]

[1]University Hospital RWTH Aachen, Neurosurgery and Spine Surgery, Aachen, Germany

Background: Hypertrophic spinal pachymeningitis (HSP) is a rare cause of cervical myelopathy. We present a case of IgG4-related hypertrophic spinal pachymeningitis treated by surgical decompression and immunosuppressive treatment. Furthermore, we provide a literature review on IgG4-related HSP.

Methods: We describe the case of a 45-year-old male presenting with cervical myelopathy. MR-imaging showed a circumferential thickening of the meninges (fig 1). The patient underwent surgical decompression through laminectomy and removal of the dural thickening. The pathological findings showed HSP with large-scale dural infiltration of IgG4-positive lymphocytes. No signs of systemic IgG4-related disease were present. Adjuvant therapy with methylprednisolone and rituximab resulted in full recovery with no clinical or radiological signs of recurrence 9 months postoperatively (fig 2).

An updated review of the literature regarding IgG4-related HSP was performed according to PRISMA-guidelines. Patient characteristics, treatment modality and outcome were reviewed.

Results: The literature review provided a summary of 53 available cases, which included the one cases from our centre. The lesions involved the craniocervical junction in 5 (9%), cervical spine in 28 (53%), thoracic spine in 32 (60%), lumbar spine in 11 cases (21%) and sacral spine in 1 case (2%). Surgical decompression was performed in 43 (81%) cases. Eight cases (15%) were not treated with steroids. Recurrence has been described after surgery or corticosteroid therapy in 7 (13%) cases.

Conclusions: IgG4-related HSP can occur without the presence of systemic IgG4-disease. Prompt diagnosis with surgical decompression and thereby dural biopsy followed by immunosuppressive treatment is essential to prevent permanent spinal cord injury.

Fig. 1

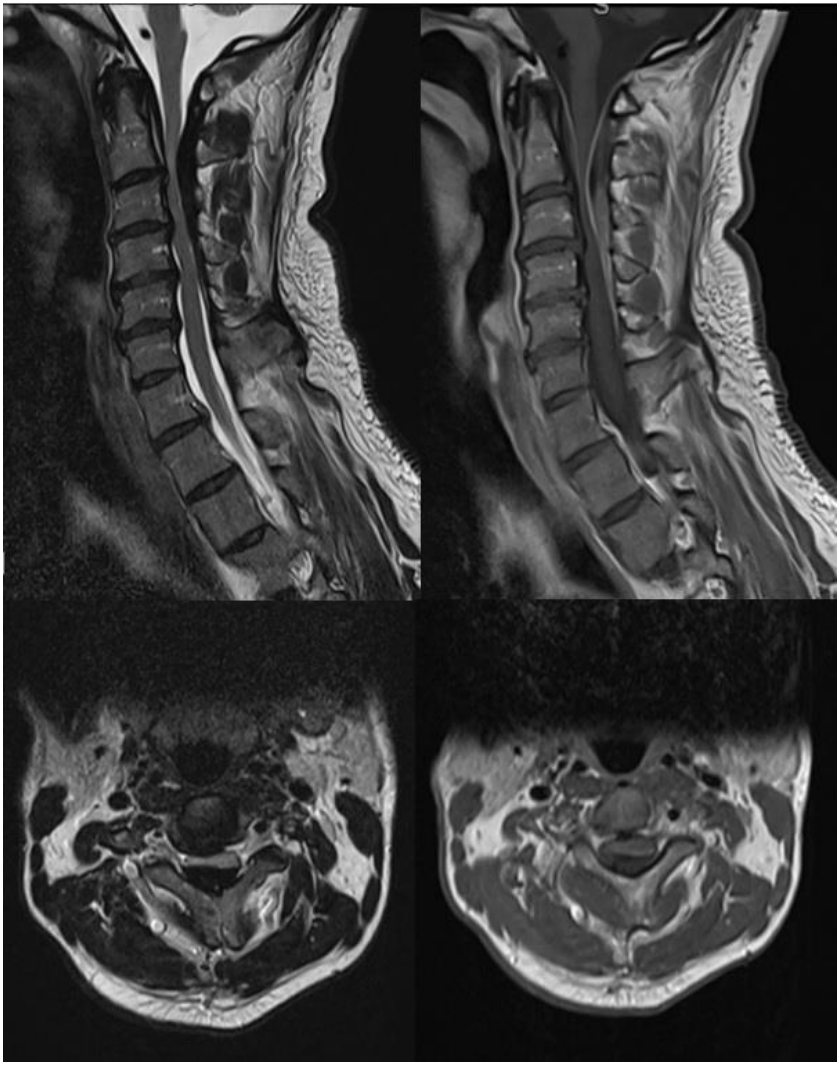

Fig. 2