ANCA-associated systemic vasculitis presenting with hypertrophic pachymeningitis: a case report

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INTRODUCTION

Hypertrophic pachymeningitis (HP) is a condition characterized by a chronic fibrosing inflammatory process usually involving the intracranial or spinal dura mater, but rarely both. Headache, cranial neuropathies, radiculopathies and seizures are the most common clinical manifestations. It was first described in 1869 as a cryptogenic thickening of the dura mater. Since then, several etiologies, including syphilis, tuberculosis and fungi, connective tissue disorders such as rheumatoid arthritis or sarcoidosis and various malignant diseases, have been identified. Furthermore, HP has been considered a rare complication of **granulomatosis with polyangitis** (GPA), occurring later in the disease course and during clinical relapses.

CASE REPORT

Sixty-one year old female admitted to our department with an 8 months history of persistent **nuchal headache**, associated with bilateral **hypoacusia**. Neurological examination was negative. Blood chemistry analysis exhibited elevated erythrocyte sedimentation rate and positive **p-ANCA levels**[1]. T2 and T2 FLAIR weighted MRI images showed pericerebellar and cervicodorsal **dural thickening**, also positive with contrast enhancement (Fig.1, Fig.2 and Fig.3a), and transependymal cerebrospinal fluid resorption *(Fig.4)*. CT scan showed bilateral **otomastoiditis**. A lumbar puncture was performed, with findings consistent with a non-specific inflammatory process. Serologic and cerebrospinal fluid testing for infectious diseases associated with chronic meningitis were negative. A diagnosis of **granulomatosis with polyangitis** was made using the recent algorithm for the classification of systemic vasculitides proposed by *Watts and colleagues*[2]. Treatment with high doses of intravenous **methylprednisolone** was started, followed by a cycle of therapeutic **plasma exchange**, with clinical, laboratoristic and radiological improvement *(Fig.3b)*. The patient was discharged with the indication of two more methylprednisolone cycles.
Shortly after the last cycle the patient presented clinical and radiological relapse, with persistent nuchal headache and **drowsiness**. She was admitted again to our department where she underwent corticosteroids and plasma exchange treatment, with no benefit. MRI findings showed signs of **subacute hydrocephalus** and initial **tonsillar herniation**.
A neurosurgical **ventriculocisternostomy** was performed with consistent symptomatic relief. The associated meningeal biopsy showed chronic inflammatory changes, lymphoplasmacytic cell infiltration and fibrous tissue hyperplasia. When the patient's conditions ameliorated she was discharged with an oral **prednisone-based maintenance therapy** and she underwent a cycle of **physical** therapy. Two months after discharge, the patient showed a gradual improvement, her chronic headache dissipated and p-ANCA levels were negative.
Rituximab immunosuppressive treatment is now being considered as a treatment option for eventual clinical disease relapse[3].

CONCLUSION

- Our case report emphasizes that **HP**, affecting both the intracranial and spinal dura mater, **may be the first clinical sign of GPA** without the recurrence of other lesions.
- Neurological manifestation, MRI findings and laboratory evaluation are essential for making an accurate diagnosis.
- A prompt surgical approach and dura mater biopsy are often required if symptoms are progressing and malignancy is suspected.
- Steroid therapy and close observation for recurrence are necessary to ensure a good long-term outcome.

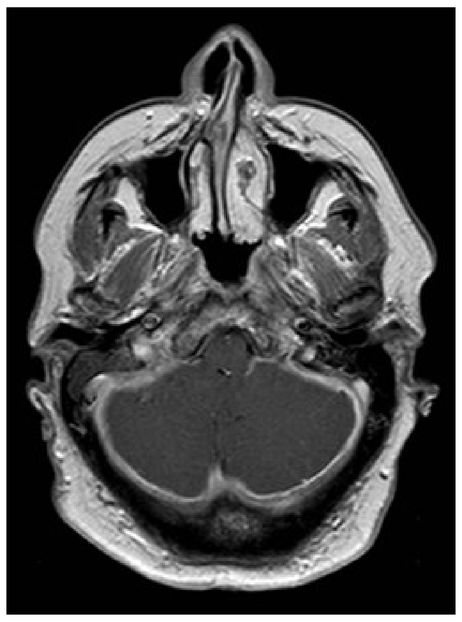

Fig.1: Axial T1 weighted MRI with gadolinium showing pericerebellar dural enhancement and thickening.

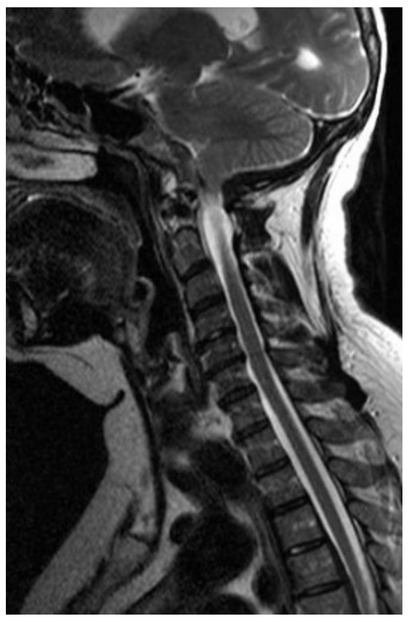

Fig.2: Sagittal T2 TSE weighted MRI showing cervical dural thickening.

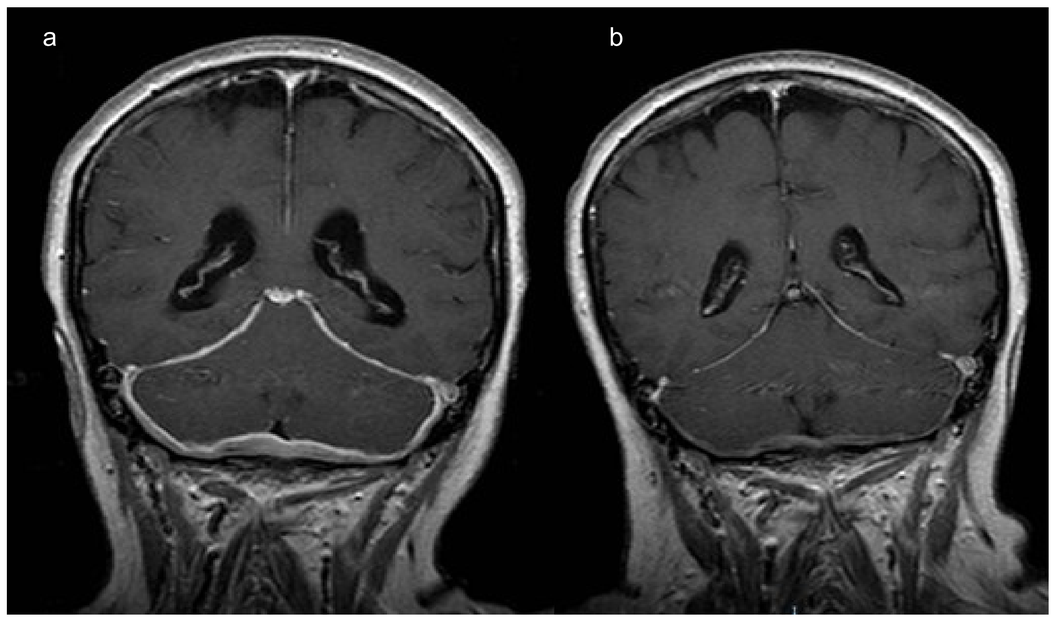

Fig. 3: Coronal T1 weighted MRI with gadolinium showing pericerebellar dural enhancement and thickening (**a**). After methylprednisolone treatment and plasma exchange therapy the same sequence showed marked reduction of meningeal post-contrast enhancement (**b**).

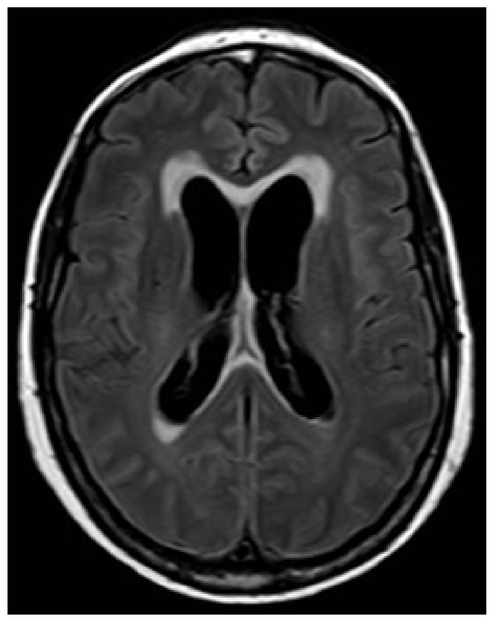

Fig.4: Axial T2 FLAIR weighted MRI showing transependimal cerebrospinal fluid resorption.

References:

1.Yokoseki A. et al. "Hypertrophic pachymeningitis: significance of myeloperoxidase anti-neutrophil cytoplasmic antibody", Brain; 2014 Feb; 137(Pt 2):520-36.
2.Watts R. et al., "Development and validation of a consensus methodology for the classification of the ANCA-associated vasculitides and polyarteritis nodosa for epidemiological studies", Ann Rheum Dis; 2007 Feb; 66(2): 222–227.
3.Just SA. et al. "Wegener's granulomatosis presenting with pachymeningitis: clinical and imaging remission by Rituximab", ISRN Rheumatol; 2011; 2011: 608942.