Hypertrophic pachymeningitis and cerebral venous thrombosis in myeloperoxidase-ANCA associated vasculitis

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DESCRIPTION

Hypertrophic pachymeningitis (HP) is a circumscribed inflammatory process that thickens meninges with fibrous adhesions. Among the causes of HP, vasculitis and autoimmune disease should be considered; myeloperoxidase (MPO)-antinuclear cytoplasmic antibodies (ANCA)-positivity can be the only feature of HP and ANCA-related vasculitis association.¹

A 47-year-old woman was admitted because of persistent severe frontotemporal headache.

Neurological examination was unremarkable. In the previous months, she had frequent affections of the upper airways and underwent surgery for bilateral maxillary sinusitis and otitis media. A previous brain MRI had shown mild white matter abnormalities and bilateral signs of otomastoiditis.

A new brain MRI showed linear contrast enhancement of the pachymeninx in the right middle cranial fossa involving the middle ear and the tegmen of the tympani (figure 1A,B). The ophthalmology evaluation showed bilateral papilloedema and visual evoked potentials revealed bilateral latency prolongation. A laboratory workup was made focusing on inflammation and antibody-mediated disease: raised acute phase proteins (erythrocyte sedimentation rate of 75 mm/h and C reactive protein level of 12.30 mg/dL) and ANCA were detected with a perinuclear pattern (p-ANCA). The cerebrospinal fluid examination showed mildly raised proteins (52 mg/dL) and no cells. Chest CT was normal, but abdomen CT demonstrated bilateral focal renal lesions (figure 1C,D), a biopsy of which showed inflammatory pathology.

The patient was suspected to have an MPO-associated vasculitis. Treatment with high-dose intravenous methylprednisolone (1000 mg/day) was administered for 5 days, followed by oral prednisone (1 mg/kg daily) with good symptomatic improvement. After 20 days, she was discharged asymptomatic.

A year later, the patient returned to our clinic complaining about headache and numbness of the left forearm. A brain MRI demonstrated signs of intracerebral sinus thrombosis (figure 1E,F). Oral anticoagulants were prescribed and a follow-up MRI after 2 weeks showed evidence for recanalisation. The patient was started on cyclophosphamide, followed by rituximab, with a complete clinical and radiological response.

We report a rare case of ANCA-associated vasculitis with MPO-ANCA-positive HP with ocular, meningeal and upper-airway involvement. Our patient did not satisfy diagnostic criteria for WG, but she had a 'limited form' with an oculo-nose-meningeal-restricted presentation.² ANCA are usually detected in WG: in particular, c-ANCA is the most frequent pattern, leading to a systemic disease involving lung and kidney.²

It has been reported that HP is more often an initial presentation of WG rather than a late complication of this disease. In our patient, HP was the presenting feature of ANCA-associated vasculitis, as previously reported.³

Figure 1 Neuroimaging data. (A and B) MRI of the brain performed at clinical onset: white arrows show linear contrast enhancement of the pachymeninx in the right middle cranial fossa involving the middle ear and the tegmen of the tympani. (C and D) Hypodense signal of bilateral focal renal lesions on abdomen CT (white arrows). (E and F) MRA with TOF performed during cerebral venous thrombosis. Hyperintense signal on FLAIR sequences of the postcentral gyrus and associated parietal cortex (arrow) due to venous infarction (E). In a coronal view the white arrow demonstrates thrombosis of right superior sagittal sinus, transversus and sigmoid sinus (F). FLAIR, fluid attenuated inversion recovery; MRA with TOF, magnetic resonance angiography with time-of-flight.

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Meningeal involvement in ANCA-associated vasculitis might confuse and complicate the differential diagnosis. However, as HP is a fibrosing process, early recognition and treatment are required, even in the absence of previous pulmonary or renal involvement. In these patients, neuroimaging and neurological examination have

a key role in making the correct and timely diagnosis, in order to reduce the progression of the disease and prevent life-threatening complications, such as cerebral venous thrombosis.

Learning points

- ▶ Myeloperoxidase (MPO)-antinuclear cytoplasmic antibody (ANCA)-positive can present as a limited form with ocular, meningeal and upper-airway involvement.
- ▶ Hypertrophic pachymeningitis (HP) and MPO-ANCA associated vasculitis should be considered in the evaluation of young patients with headache and inflammatory indices elevation.
- ▶ As HP is a fibrosing process, early diagnosis of HP in the acute setting and prompt treatment can prevent or reduce a more severe neurological damage in ANCA-associated vasculitis, considering its good clinical response to immunosuppressive therapy.

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