ANCA-ASSOCIATED HYPERTROPHIC PACHYMENINGITIS, A CENTRAL NERVOUS SYSTEM LIMITED TYPE OF SYSTEMIC VASCULITIS

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LEARNING POINT FOR CLINICIANS

Hypertrophic pachymeningitis (HP) is characterized by a thickening of the dura mater that in some cases has been related to systemic diseases. Some hearing loss may be the first clinical manifestation of PH as a form of ANCA-positive vasculitis limited to the brain. Treatment with rituximab appears to be a good therapeutic option for these cases.

CASE PRESENTATION

Hypertrophic pachymeningitis (HP) is a dura mater thickening that has been linked to some systemic diseases. We present the case of a 64-year-old woman with hearing loss who was diagnosed with HP associated with antineutrophil cytoplasmic antibodies (ANCA).

Our patient initially consulted in 2014 for acufen and hearing impairment, along with xerophthalmia. Her main comorbidities were hypercholesteremia, subclinical hypothyroidism and areata alopecia. Examination was normal except for bilateral, right-dominant, mixed hearing loss. An ophthalmological evaluation found bilateral keratitis and dry eye (Schirmer test 0 mm). Laboratory tests showed an Hb of 11.3 g/dL, ESR of 70 mm/h and CRP of 0.9 mg/dL (N<0.5). QuantiFERON-TB, HIV and hepatotropic virus serologies were negative. Autoimmunity studies found ANA + 1/160, negative anti-DNA antibodies and ENAS, ANCA + 1/80 with perinuclear pattern and anti-myeloperoxidase antibodies (MPO) 53.9 AU/mL (N< 20). IgG4 was normal. A cranial CT showed occupation of both tympanic boxes.

She was initially treated with 3 pulses of 250 mg 6-methylprednisolone (6MP), 0.5 mg/kg/d prednisone and 15 mg/week methotrexate, with progressive hearing improvement. However, six months later she developed left trigeminal neuralgia. A cranial MRI revealed HP and vasogenic temporal edema (Image 1). Spinal MRI and CSF were normal. ANCA-associated HP was diagnosed and she was therefore treated with 3 pulses of 1 g 6MP and 2 g Rituximab (RTX) with clinical improvement. An MRI 12 months later showed HP resolution (Image 2).

After 2 years she consulted for fever, cough, malaise, headache and asthenia. She did not refer polymyalgia, jaw claudication or visual symptoms. Physical examination was normal. Analysis showed Hb 9.9 mg/dL, ESR 119 mm/h, CRP 11.5 mg/dL and ANCA-MPO 218.4 U/mL (N <1.0). Blood cultures and thoracic-abdominal CT were normal. A new cranial MRI found no meningeal involvement but ultrasound of temporal arteries showed the halo sign. Right temporal artery biopsy and FDG-F18 PET/CT were both normal. Final diagnosis was CNS limited MPO-ANCA vasculitis with HP. This time she was given 3 pulses of 125mg 6MP, 30 mg/d prednisone and 2 gr RTX with clinical remission and normalization of inflammatory parameters.

Discussion

HP is an inflammatory disease diagnosed by meningeal thickening and dural contrast uptake in T1 sequences of cranial MRI. Although most are labeled as idiopathic, some cases have been
associated with infections, neoplasia, sarcoidosis, IgG4 disease, rheumatoid arthritis, and vasculitis. Yokoseki et al reviewed 17 cases of ANCA-MPO HP diagnosed between 1996 and 2012 at a Japanese hospital. 29% presented chronic sinusitis, 65% otitis media and 47% mastoiditis, typical clinical features of granulomatosis with ANCA-antiproteinase3 polyangiitis. In 76% of those patients, hearing loss preceded HP, and was due to either otitis media or damage to the VIII pair. Recently Kobayashi et al. described 7 other patients with ANCA-MPO HP treated with RTX between 2013 and 2018. RTX treatment was effective in all patients, even those who were refractory to cyclophosphamide.

Our patient initially responded to RTX and was in remission for 2 years before developing systemic inflammation. The halo sign, although characteristic of vasculitis, has also been described in amyloidosis, neoplasia and can be a false positive. We believe that in some mixed hypoacusia HP should be considered, which can be a limited form of vasculitis and where RTX can be an effective treatment.

Conflict of interest. None declared.
Imag 1: A) Coronal T1 after IV contrast: homogeneous dural enhancement in the floor of the right middle cranial fossa (red arrow). B) Coronal T2: Right dural thickening with low signal (red arrow), associated severe temporal vasogenic edema (green arrow) and occupation of mastoid cells by high density inflammatory material (blue arrow).

Image 2: A) Coronal T1 after IV contrast: significant decrease of homogeneous dural enhancement in the right temporal fossa (red arrow) B) Coronal T2: resolution of right temporal dural thickening and associated vasogenic edema (green arrow)

Reference


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IMAGE 2: A) Coronal T1 after IV contrast: significant decrease of homogeneous dural enhancement in the right temporal fossa (red arrow) B) Coronal T2: resolution of right temporal dural thickening and associated vasogenic edema (green arrow)

975x1087mm (57 x 57 DPI)