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Idiopathic cranial pachymeningitis – diabetes was not the brains

Emmanuel Sagui, Arnaud Jouvion, Mathieu Planchard, Michel Bregigeon, Christian Brosset,
Service de Neurologie, Hôpital d’Instruction des Armées Laveran, Marseille Cedex 13, France
Correspondence to Emmanuel Sagui, emlsmg@gmail.com

DESCRIPTION
A 58-year-old diabetic woman presented with a 5-week history of sudden-onset diplopia and progressive headache. One week before admission, she also complained of nausea. She reported no history of allergic rhinitis or arthritis. Physical examination revealed isolated left-cranial-nerve VI palsy. Routine biological tests were unremarkable, apart from hyperglycaemia 300 mg/dl and glycosylated haemoglobin 12.4%. Eosinophilia was normal. Lumbar puncture revealed pleocytosis with 8/mm³ cells and protein at 170 mg/dl.

MRI of the brain disclosed abnormal linear dural enhancement and T2 hyperintensity of dura mater, including falx and tentorium (figure 1).

On clinical and MRI grounds, hypertrophic cranial pachymeningitis (HCP) was diagnosed, and idiopathic HCP was ascertained after second-line investigations failed to ascertain a known aetiology: serological tests to syphilis and Lyme disease were negative; all cerebrospinal fluid stains and cultures to fungi and Mycobacterium tuberculosis were negative; chest CT was normal; c-ANCA were negative; minor salivary glands biopsy was normal; biopsy of the right superficial temporal artery revealed no arteritis. Lep-tomeningeal biopsy showed infiltrates of mature lymphocytes, without evidence of granuloma, vasculitis or neoplastic cells.

HCP is a rare disorder due to local or diffuse inflammation of the dura mater with abnormal dural enhancement revealed on MRI. Aetiological diagnosis of HCP is still a challenge. Once intracranial hypotension is ruled out on the basis of clinical features – orthostatic headache – four conditions should be evoked: (1) infectious diseases, mainly tuberculosis, syphilis, HTLV1, Lyme disease and fungal infections; (2) neoplasia, mainly lymphoma and dural carcinomatosis; (3) granulomatous diseases such as neurosarcoïdosis or Wegener's disease; (4) vasculitis, either primary or secondary to rheumatoid arthritis or temporal arteritis.

Idiopathic HCP is ascertained after these diagnoses are ruled out.
COMPETING INTERESTS
None.

PATIENT CONSENT
Obtained.

REFERENCES