Idiopathic hypertrophic pachymeningitis presenting with occipital neuralgia

Laurent Auboire1, Jonathan Boutemy2, Jean Marc Constans3,4, Thomas Le Gallou2, Philippe Busson5, Boris Bienvenu2,3

1. Université François-Rabelais de Tours, Inserm, Imagerie et Cerveau UMR U930, Tours, France
2. CHU de Caen, Department of internal medicine, 14000-Caen, France
3. Université de Caen Basse-Normandie, Medical School, 14000-Caen, France
4. CHU de Caen, Department of Radiology, 14000-Caen, France
5. CH d’Avranches, Department of Medicine 2, 50300-Avranches, France

Abstract:
Background: Although occipital neuralgia is usually caused by degenerative arthropathy, nearly 20 other aetiologies may lead to this condition.
Methods: We present the first case report of hypertrophic pachymeningitis revealed by isolated occipital neuralgia.
Results and conclusions: Idiopathic hypertrophic pachymeningitis is a plausible cause of occipital neuralgia and may present without cranial-nerve palsy. There is no consensus on the treatment for idiopathic hypertrophic pachymeningitis, but the usual approach is to start corticotherapy and then to add immunosuppressants. When occipital neuralgia is not clinically isolated or when a first-line treatment fails, another disease diagnosis should be considered. However, the cost effectiveness of extended investigations needs to be considered.
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Case report
A 56-year-old man was admitted with moderate-intensity pain that originated at the base of the skull and radiated to the left side of the occipital scalp. Left occipital neuralgia was diagnosed. A few weeks later, the patient was admitted again, this time for an intense headache that required opioid painkillers. Magnetic-resonance imaging (MRI) and computed tomography (CT) scans showed thickening of the dura mater, ranging from the cerebellar tentorium (see arrow, Figure 1) to the meninges of the second cervical vertebra (see arrow, Figure 2).

Corresponding author:
Boris Bienvenu,
Service de Médecine Interne,
CHU Côte de Nacre, CS 30001,
14033 Caen cedex 9, France
Tel : +33(0)231064584;
e-mail: bienvenu-b@chu-caen.fr

Figure 1: MRI T1 axial image showing thickening of the left cerebellar tentorium.

Figure 2: CT-scan axial image showing thickening of the meninges between the C1–C2 vertebrae.
MRI excluded cerebral venous sinus thrombosis. Cerebrospinal-fluid analysis (CSF) showed hyperproteinorrachia (0.86 g/L) with normal glycorrhachia, 64 leukocytes/mm³ with 93% activated non-clonal lymphocytes, mostly CD4+. A PCR assay to detect Mycobacterium tuberculosis DNA was negative. An examination and culture of the CSF was negative which allowed eliminating bacterial causes, especially tubercular. There were no abnormal cells in the CSF. Serological testing for HIV 1 and 2 was negative. A body CT scan revealed no carci-

One case report of idiopathic hypertrophic cranial pachymeningitis (IHP) has been described, for the first time to our knowledge, a case of idiopathic pachymeningitis revealed by isolated occipital neuralgia. We found one similar case reported in the literature, but occipital neu-

Although occipital neuralgia is relatively common and is predominantly caused by osteoarthritis, the literature provides no formal evidence on its prevalence and incidence, probably because of its frequent ambulatory management. Its diagnosis is based on cri-

A clinical examination, a complete blood count, sedi-

In a nationwide survey in Japan between 2005 and 2009, which included 70 cases of idiopathic hyper-

Clinical examination, a complete blood count, sedi-

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| Table 1: The main etiologies of occipital neuralgia |
| --- | --- | --- | --- |
| Cause | Personal history | Clinical symptoms | Imaging |
| Cervical spondylitis [6] | Occipital artery, 
vertebral artery, or posterior inferior cerebellar artery. | None | Neurological abnormality | None |
| Cervicobrachialgia [7] | Angina pectoris, 
Arteriovenous malformation [8] | None | None |
| Neurosyphilis [10] | History of syphilis, 
gummatous or cardiovascular syphilis, HIV infection or compromised immune status | Dark-field microscopy of an active chancre, VDRL and RPR test, enzyme immunoassay not for nonvenereal or other causes, fluorescent treponemal antibody-absorption test, lumbar puncture (white blood cell count, VDRL test, TPHA test). |
| Herpes zoster infection [11] | History of recent facial herpes lesion | Herpes viral culture of a skin lesion and blood serologies (Herpes simplex virus IgG and IgM). |
| Joint and bone diseases [12]: hypermobility posterior arch of atlas, osteolytic lesion of unknown cause, or exuberant calcification formation | Arthritis | Protein electrophoresis (for myeloma). |
| Myelitis [13] | None | Neurological abnormality | None |
| Rheumatoid arthritis [14] | Polyarthritis | CRP, Rheumatoid factor, anti-citrullinated protein antibodies | X-ray of the hands, MRI |
| Tumor [15] | Personal history of cancer, other localization known | Neurological abnormality | None |
| Trauma [16] | Clinical history of trauma | Neurological abnormality | None |

Abbreviations: CRP: C-reactive protein; PMR: polymyalgia rheumatica; MRI: magnetic resonance imaging; VDRL: Venereal Disease Research Laboratory; RPR: rapid plasma reagent; TPHA: Treponema pallidum hemagglutination assay.
then to add immunosuppressants. We suggest that additional exploration of occipital neuralgia is warranted when conventional treatments fail. However, the cost-effectiveness of extended investigations needs to be considered.

References