Idiopathic hypertrophic pachymeningitis presenting with occipital neuralgia

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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).

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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 hyperproteineorachia (0.86 g/L) with normal glycorraclia, 64 leucocytes/mm² 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 tuberculosis. There were no abnormal cells in the CSF. Serological testing for HIV 1 and 2 was negative. A body CT scan revealed no carcinoma or other disease except for the cerebral anomaly (described above). We did not find any evidence of lymphomatous or systemic disorders (granulomatosis with polyangiitis, Sjögren's syndrome, rheumatoid arthritis, mixed connective-tissue disease). Therefore, a meningeal biopsy was performed and revealed lymphoplasmacytic infiltration with no abnormal cells. Accordingly, we diagnosed idiopathic hypertrophic cranial pachymeningitis (IHP).

During our investigations, a nerve block was performed to reduce the pain, and was repeated, but the results were insufficient. Slight asymmetry in the muscles of the neck suggested damage to the 12th left cranial nerve. After this diagnosis, we initiated corticosteroid therapy. However, the patient was still cortico-dependent (at 15 mg/day) at 2 years after starting therapy. Consequently, we decided to introduce cyclophosphamide therapy. The patient received six pulses of cyclophosphamide at 0.7 g/m² per month. A MRI scan then showed significant improvement, in particular next to the median parietal cortex.

Cyclophosphamide therapy was followed by a maintenance treatment. Our patient remains clinically stable under methotrexate therapy (17.5 mg/week and prednisone 5mg/day). The patient no longer needs painkillers.

**Discussion**

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 criteria from the current International Classification of Headache Disorder-2 (ICHD-2). The principal causes of occipital neuralgia are summarized in Table 1.

A clinical examination, a complete blood count, sedimentation rate, C-reactive protein, viral and bacteriological tests and/or serologies, in addition to imagery from a CT-scan or MRI, are required to exclude most secondary causes.

<table>
<thead>
<tr>
<th>Causes</th>
<th>Personal history</th>
<th>Clinical symptoms</th>
<th>Histological tests</th>
<th>Imaging</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypertrophic pachymeningitis (HP)</td>
<td>None</td>
<td>Neurological abnormality</td>
<td>None</td>
<td>MRI</td>
</tr>
<tr>
<td>Multiple sclerosis (MS)</td>
<td>Age &gt;50 years. Personal history of PMR</td>
<td>Weight loss, long-term low-grade fever, abolition or reduction of the temporal pulse, jaw claudication, occipital hyperesthesia</td>
<td>Sedimentation rate, CRP, temporal-artery biopsy</td>
<td>Digital ultrasound, high-resolution MRI</td>
</tr>
<tr>
<td>Pott's disease [9]</td>
<td>Personal history of tuberculosis or contact with tuberculosis</td>
<td>Back pain, fever, night sweats, anorexia, weight loss, spinal mass, tuberculin test positive, muscle weakness of the legs</td>
<td>Erythrocyte sedimentation rate, tuberculin skin test, bone biopsy</td>
<td>MRI, X-ray of the spine, CT scanner</td>
</tr>
<tr>
<td>Neurosyphilis [10]</td>
<td>Personal history of syphilis, granulomatous or cardiovascular syphilis. HIV infection or compromised immune status.</td>
<td>Sarcoidosis, ataxia, apathy, paragangliomatosis, personality changes, cognitive disturbance, visual changes, hearing loss, neuropathy, loss of beard or bladder function or sign of gummatous vessels or posterior inferior cerebellar artery blood supply.</td>
<td>Dark-field microscopy of an active chancre, VDRL and RPR test, enzyme immunoassay for antiproliferal IgG, fluorescent antibody test, antinuclear antibodies, antinuclear antibody test, IgG and IgM</td>
<td>None</td>
</tr>
<tr>
<td>Herpes zoster infection [11]</td>
<td>History of a recent facial herpes lesion</td>
<td>Reddening of the occipital skin with vesicles</td>
<td>Herpes viral culture of a skin lesion and blood serology (Herpes simplex virus IgG and IgM)</td>
<td>None</td>
</tr>
<tr>
<td>Joint and bone diseases [12]: hypermobile posterior arch of atlas, osteolytic lesion of unknown cause, or exuberant callus formation</td>
<td>Arthritis</td>
<td>Pain exacerbated by motion</td>
<td>Protein electrophoresis (for myeloma)</td>
<td>MRI</td>
</tr>
<tr>
<td>Myelitis [13]</td>
<td>None</td>
<td>Neurological abnormality</td>
<td>None</td>
<td>MRI</td>
</tr>
<tr>
<td>Rheumatoid arthritis [14]</td>
<td>Polyarthritis</td>
<td>Rheumatoid nodules, ulnar deviation, boutonniere deformity, swan-neck deformity</td>
<td>CRP, Rheumatoid factor, anti-citrullinated protein antibodies</td>
<td>X-ray of the hands, MRI</td>
</tr>
<tr>
<td>Tumor [15]</td>
<td>Personal history of cancer, other localization known</td>
<td>Neurological abnormality</td>
<td>None</td>
<td>MRI</td>
</tr>
<tr>
<td>Trauma [16]</td>
<td>Clinical history of trauma</td>
<td>Neurological abnormality</td>
<td>None</td>
<td>MRI</td>
</tr>
</tbody>
</table>

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