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.

Keywords: neuralgia/pathology, meningitis, neck pain/aetiology, review

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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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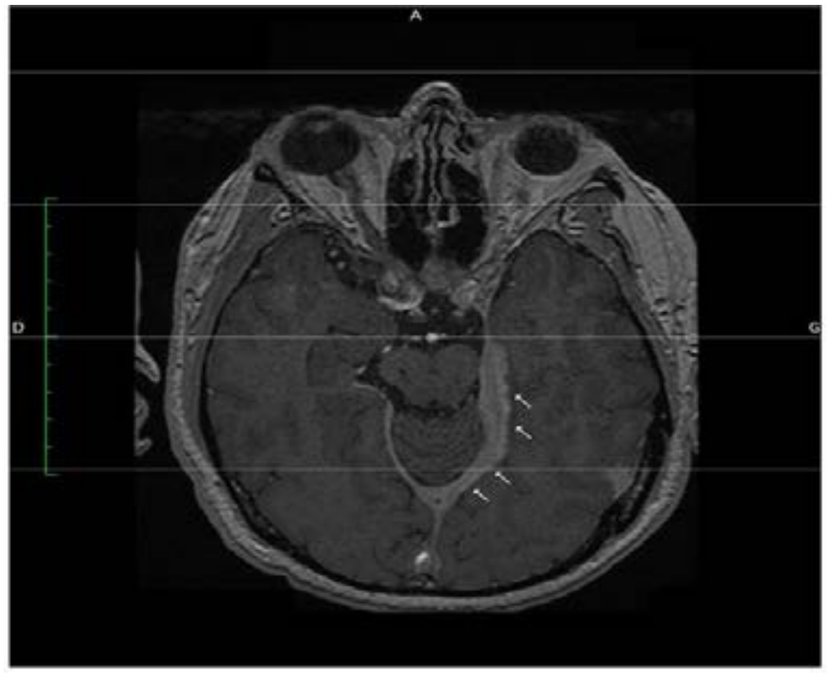

Figure 1: MRI T1 axial image showing thickening of the left cerebellar tentorium.

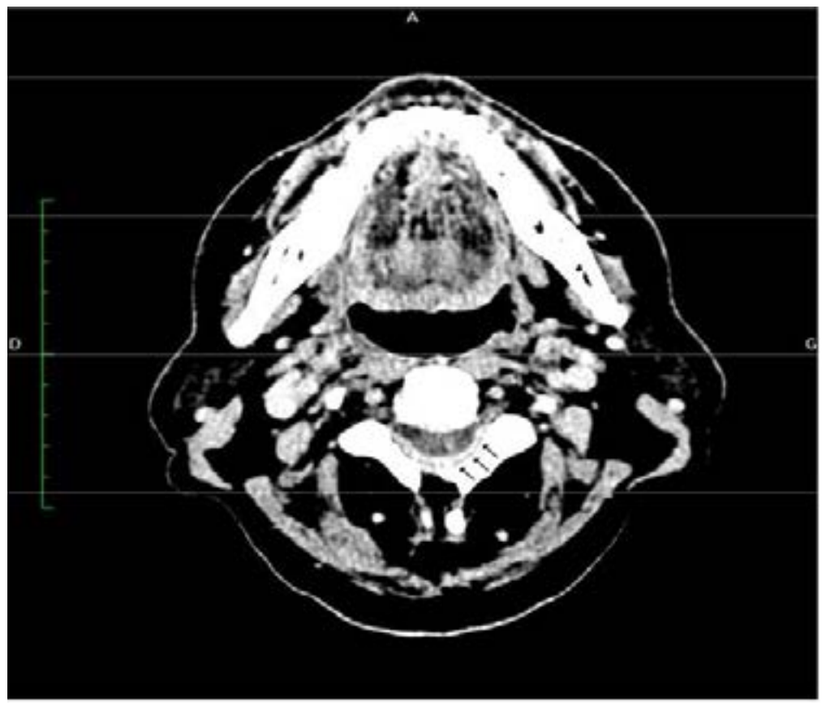

Figure 2: CT-scan axial image showing thickening of the meninges between the C1–C2 vertebrae.

MRI excluded cerebral venous sinus thrombosis.
Cerebrospinal-fluid analysis (CSF) showed hyperproteinorachia (0.86 g/L) with normal glycorrhachia, 64 leukocytes/mm^3 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 lymphoplasmocytic 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 pain killers.

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[1]. Its diagnosis is based on criteria from the current International Classification of Headache Disorder-2 (ICHD-2)[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.

Hypertrophic pachymeningitis (HP) is a rare entity. Yonekawa et al.[3] reported that 159 cases were found during a nationwide survey in Japan between 2005 and 2009, which included 70 cases of idiopathic hypertrophic pachymeningitis. The causes of HP are various: infections, autoimmune disease, tumours, trauma, or idiopathic. Considering only IHP, in Yonekawa et al.'s study, females were predominant (gender ratio of 1/0.75). The overall mean age was 54.8 +/- 16.5 years. Headache (62.9%) was the most common symptom and was often accompanied by cranial-nerve palsy (55.7%). Corticotherapy alone was effective in 58.7% of cases. Immunosuppressants were added when there was a suboptimal response to corticosteroids: this led to remission in 21.4% of these cases. Bosman et al.[4] reported similar proportions when they reviewed treatment of IHP cases published between 1990 and 2008. They identified 60 patients: 93% were given a corticotherapy, of which 65% were monotherapies: this led to 46% relapse rate. Some patients required radiotherapy or surgery when their condition became too serious (e.g., hydrocephalus).

Herein, we have 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 neuralgia was not isolated as the patient also presented with tinnitus and deafness at admission. Corticosteroids did not improve the clinical situation and, similar to our patient, a sparing treatment had to be introduced[5].

Conclusion

IHP is a possible cause of occipital neuralgia and may present without cranial-nerve palsy at the beginning. There is no consensus on IHP treatment, but the most common approach is to initiate corticotherapy and

Table 1: The main etiologies of occipital neuralgia

Causes	Personal history	Clinical symptoms	Biological tests	Imaging
Vascular compression [6]: occipital artery, ectatic vertebral artery, or posterior inferior cerebellar artery	None	Neurological abnormality	None	MRI
Vasculitis [7]: giant-cell arteritis	Age >50 years. Personal history of PMR	Weight loss, asthenia, long-term low-grade fever, abolition or reduction of the temporal pulse, jaw claudication, occipital hyperesthesia	Sedimentation rate, CRP, temporal-artery biopsy	Doppler ultrasound, high-resolution MRI
Arterio-venous malformation [8]	Other localization of arterio-venous malformation	Vascular bruit	None	MRI
Pott's disease [9]	Personal history of tuberculosis or contact with tuberculosis	Back pain, fever, night sweats, anorexia, weight loss, spinal mass, paresthesia, or muscle weakness of the legs	Erythrocyte sedimentation rate, tuberculin skin test, bone biopsy	MRI, X-ray of the spine, CT-scanner
Neurosyphilis [10]	Personal history of syphilis, gummatous or cardiovascular syphilis. HIV infection or compromised immune status.	Seizures, ataxia, aphasia, paresis, hyper-reflexia, personality changes, cognitive disturbance, visual changes, hearing loss, neuropathy, loss of bowel or bladder function or signs of gummatous syphilis or cardiovascular syphilis.	Dark-field microscopy of an active chancre, VDRL and RPR test, enzyme immunoassay test for antitreponemal IgG, fluorescent treponemal antibody-absorption test, lumbar punction (white blood cell count, VDRL test, TPHA test)	None
Herpes zoster infection [11]	History of a recent facial herpes lesion	Reddening of the occipital skin with vesicles	Herpes viral culture of a skin lesion and blood serologies (Herpes simplex virus IgG and IgM)	None
Joint and bone diseases [12]: hypermobile posterior arch of atlas, osteolytic lesion of unknown cause, or exuberant callus formation	Arthritis	Pain exacerbated by motion	Protein electrophoresis (for myeloma)	MRI
Myelitis [13]	None	Neurological abnormality	None	MRI
Rheumatoid arthritis [14]	Polyarthritis	Rheumatoid nodule, ulnar deviation, boutonniere deformity, swan-neck deformity	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	MRI
Trauma [16]	Clinical history of trauma	Neurological abnormality	None	MRI

Abbreviations: CRP: C-reactive protein; PMR: polymyalgia rheumatica; MRI: magnetic resonance imaging; VDRL: Venereal Disease Research Laboratory; RPR: rapid plasma reagin; 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.

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