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Occipital neuralgia as a presentation of multiple sclerosis

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Abstract

Occipital neuralgia (ON) is a sharp, jabbing, electric shock like pain in the distribution of the greater, lesser and/or third occipital nerves and is often associated with decreased sensation or dysesthesia in the area. The etiology may be idiopathic, traumatic or from compressive lesions of the area. Headaches have been reported to be initial symptoms of multiple sclerosis (MS) recently. Neuralgia form headache is common in MS. However, the prevalence of ON in MS is still unknown and is under-recognized as a symptom of MS. We report two patients who had ON without any other neurological deficit at the time of presentation who were found to have demyelinating cervical lesions on magnetic resonance imaging. Moreover, in one of the patients, ON itself was the initial symptom of MS. ON of both patients improved with intravenous 1000 mg/day methyl prednisolone (IVMP) treatment. In conclusion, neuralgia form headache is increased in patients with MS. However, headache may be overlooked by both patients and clinicians, and may not be diagnosed as a symptom of MS in clinical practice. ON without any other neurological finding is rare as a symptom in MS. It should be kept in mind that ON by itself may be the initial symptom of MS and will respond to IVMP. The consideration of MS as a cause of ON may enable more rapid diagnosis and treatment of MS.

Keywords: Headache, multiple sclerosis, occipital neuralgia, spinal cord

INTRODUCTION

Occipital neuralgia (ON) is characterized by paroxysmal intense pain that feels like a sharp, jabbing, electric shock in the distribution of the greater, lesser and/or third occipital nerves. It is often associated with decreased sensation or dysesthesia. ON is usually idiopathic, but may be associated with trauma, compression or inflammation of the occipital nerves and/or higher cervical segments of the spinal cord (C2-C3).[1] Although the specific incidence and prevalence rates for ON remain unknown, it is not very common.[2]

Multiple sclerosis (MS) is a chronic, autoimmune, neuroinflammatory disease with a worldwide prevalence of 2.3 million. Although the incidence of headache in MS varies considerably, headache has been increasingly recognized as a symptom of MS.[3,4] Researchers reported that 50% of MS patients experienced headache, of which 27.6% were migraines and 37.2% were tension-type headaches. Neuralgiform headaches are increased in MS due to demyelination of the central myelinated part of the nerves.[5] The relationship between trigeminal neuralgia and MS is well-documented, with a prevalence ranging from 1.9% to 4.9%, however, there is no definite information about the incidence or prevalence of ON in MS.[2] There is limited study about ON in MS. We report-2 patients where ON was the sole symptom of underlying MS.

CASE REPORTS

Patient 1

A 28 year-old woman with MS who was using glatiramer acetate (20mg/day) for maintenance treatment was admitted to hospital with severe, acute-onset, left sided occipital headache that gradually increased and had persisted for 3 days. The headache was defined as sharp and jabbing like an electric shock and repetitive in short intervals, lasting for a maximum of 15 minutes. It was unresponsive to non-steroidal anti-inflammatory drugs. She had no previous history of headache. The only pathological finding in

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neurological examination was bilateral increased deep tendon patellar reflexes.

T2 weighted magnetic resonance imaging (MRI) revealed multiple ovoid plaques in periventricular, subcortical, infratentorial and juxtacortical areas in the brain. There was a gadolinium enhancing lesion at the C2 level (Figure 1). She was diagnosed with s a relapse of current MS presenting as ON and was treated with 1000mg/day intravenous methylprednisolone (IVMP) for 7 days. The headache completely improved after treatment and the high intensity signal in C2 disappeared on follow-up MRI scanning. Six months after the treatment, she was still pain-free without any medication.

Patient 2

A 40-year-old woman with no known disease was admitted to hospital with a thunderclap pain and dysesthesia localized in the left occipital region. Her headache was in the form of recurrent episodes of a few seconds repeated about 15 times during the day, and it could be triggered with massage. Neurological examination was normal.

In T2 weighted MRI, there were multiple, hyperintense, ovoid lesions in periventricular and juxtacortical areas and a single, 1 cm length, hyper intense lesion in C2 (Figure 2). Oligoclonal band was detected in lumbar puncture. Vasculitic and infectious markers were negative. She had a cousin with MS. She was diagnosed with MS and her complaints improved completely after 1000 mg IVMP treatment for 5 days.

DISCUSSION

Stabbing pain in the distribution of the greater, lesser and/or third occipital nerves, sensitization of the affected nerve and temporary relief of headache with local blockade of the relevant nerve are defined as the diagnostic criteria for ON by International Headache Society (IHS). ON is paroxysmal, sharp, electric shock-like pain which originates in 90% of cases from the greater occipital nerves, 10% from the lesser occipital nerves and very rarely, from the third occipital nerve. The incidence of ON has been reported as 3.2 per 100,000 people and the mean age of diagnosis was 54.1 years.[6]

The etiology is usually idiopathic, however trauma, compression or demyelination in the root of occipital nerves or in higher cervical segments of spinal cord can also be causes. ON has been reported secondary to neurosyphilis, C1-C2 arthrosis, cervicomedullary dural arteriovenous fistula, upper cervical cavernous angioma, vascular compressions and MS.[5]

Neuralgiform headache, particularly trigeminal neuralgia is well-defined in MS. However, the association of ON with MS has been reported only in case series, and it is quite rare that an ON by itself is found to be a relapse or initial finding of MS. Although neuroimaging findings were not performed to prove the relationship, Erikson *et al*. reported ON as a MS relapse in 5 of 255 MS patients during a 25-year follow-up.[7] De Santi *et al*. reported 3 MS patients with ON where C2 lesions improved after IVMP.[8] Kissoon *et al*. reported ON in 8 patients with primary progressive MS and in 12 patients with relapsing remitting MS in 2019.[5]

MS patients have an increased incidence of headache. Recent studies reported that headache should be evaluated as an early symptom in MS.[4] New onset headache might be related to a new active lesion in MS, however headache is still overlooked by both patients and clinicians in practice. Although various kinds of headaches can be detected in MS, neuralgiform headache has a particular relationship with MS. Trigeminal neuralgia is the neuralgiform headache whose relationship with MS is mostly understood, however ON is also another neuralgiform headache which is also associated with MS.

Treatment of ON varies according to the etiology; and includes oral analgesics, anticonvulsants, cervical traction, electrical nerve scattering, local anesthetic injections ganglion blockade. However, ON caused by MS lesions are unresponsive to pain killers and it can only be treated by IVMP.

In the current report, ON was diagnosed as a relapse of MS in the first patient, and as an initial symptom of MS in the second. They both had new lesions in C2 and pain was relieved after IVMP.

In conclusion ON may be a symptom of MS relapse or the initial symptom of MS. IVMP is the most appropriate treatment. Suspicion of MS in cases with ON could reduce delay in diagnosis of MS and can decrease disability by enabling early treatment.

DISCLOSURE

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