Occipital neuralgia as a manifestation of neuro-Behçet’s relapse: a case report

Sir,

According to the International Headache Association, occipital or C2 neuralgia is a paroxysmal, painful sensation in the major or minor occipital nerve dermatome. Although its etiology remains unclear in most cases, reported secondary causes include cervical compression, musculoskeletal system pathologies or myelitis (1).

Behçet’s syndrome (BS) is a chronic inflammatory disease of unknown origin characterised by orogenital ulcerations and uveitis. Neuro-Behçet’s syndrome (NBS) can occur in up to 10% of the patients and include mainly two types: parenchymal NBS and cerebral venous sinus thrombosis (2). Parenchymal NBS is a serious morbidity of BS leading often to disability or mortality; the anatomical regions most commonly affected in NBS are the brain stem (3). Spinal cord involvement can be rarely observed (4, 5).

Headache is the most common neurological symptom seen in BS and can be of several types. Our group had reported a prevalence of 66.2% in a cohort of randomly selected BS patients (6). We now describe a different type of headache which is called occipital neuralgia probably due to spinal cord involvement in a patient with NBS. The patient was a 28-year-old male with a remote history of BS diagnosed 13 years ago with recurrent oral ulcers, uveitis and positive Pathergy test. He was off treatment for about 6 years due to being clinically silent. The patient was admitted to our emergency department with a headache, dysarthria, and right-side weakness. Cranial magnetic resonance imaging (MRI) revealed a pontomesencephalic lesion consistent with the neurological involvement of BS. He was treated first with intravenous methylprednisolone (IVMP) 1 g daily × 10 days. Afterwards, oral prednisolone was also given (40 mg/day) for 1 month than tapered to 5 mg/day. Infliximab was added as a preventive treatment. After 8 months while he was receiving infliximab at 6 weeks intervals, he presented with dysarthria, right-sided weakness, and a new headache that had developed over 1 month. He described his headache as stabbing, located in the right occipital region, lasting seconds, and occurring more than 10 times a day, which was compatible with occipital neuralgia. He did not have active ocular inflammation and his ophthalmological examination revealed chronic signs of uveitis. His rheumatological examination was found to be normal. MRI of the cervical spine showed a lesion extending from the medulla oblongata to the C1-C2 level with gadolinium enhancement at the medulla oblongata (Fig. 1a, b). He was treated with IVMP 1 g daily × 10 days. While the right-sided weakness and dysarthria recovered completely following the IVMP treatment, the neuralgiform pain remained in the right occipital region. Additionally, he did not benefit from the optimum doses of gabapentin or pregabalinal. Cervical MRI was repeated to exclude a new relapse and revealed a sequel lesion on the right lateral side of C2, which explained the ongoing neuralgiform pain (Fig. 1c). As the right greater occipital nerve (GON) was tender on palpation and he did not respond to medical treatment, a GON block was performed with 2 mL of lidocaine and 80 mg of methylprednisolone. The neuralgiform pain resolved completely after the GON block and did not recur during 1-year follow-up. Neurological involvement, referred to as NBS, occurs in 5–10% of the patients with BS (5). Headache is the most common neurological symptom of NBS and can be due to various causes. Given that NBS may present with upper spinal cord involvement, occipital neuralgia may occur during the spinal cord relapse of NBS. The GON arises from the medial branch of the C2 dorsal ramus, receiving a bifurcation from the medial branch of the dorsal ramus of C3, the third occipital nerve, which innervates the skin of the back of the scalp up to the vertex. Therefore, high cervical inflammatory lesions can cause occipital neuralgia, and clinicians should suspect spinal cord relapse of NBS in patients with severe occipital neuralgia and perform cervical MRI.

Idiopathic occipital neuralgia is usually refractory to medication; in neuroinflammatory diseases, however, an almost complete response is obtained with high-dose IVMP (7). In patients unresponsive to IVMP, as in our case, a GON block should be considered as a treatment option. Although occipital neuralgia due to high cervical lesions occurs in demyelinating syndromes (8, 9), our case is, to our knowledge, the first case of occipital neuralgia secondary to spinal cord involvement of NBS.

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Fig. 1. (a) T2-FLAIR sagittal image demonstrates a lesion extending from the medulla oblongata to the C1-C2 level of the spinal cord. (b) T1 sagittal post contrast image shows gadolinium enhancement at medulla oblongata. (c) T2 axial image reveals a lesion at the right side of C2 on follow up MRI.

Letters to the Editors

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1
References