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Teaching Case: Occipital neuralgia in a young patient

**Expert commentary** 

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This 27-year-old man developed a daily headache in the occipito-cervical area bilaterally following a motor vehicle accident. His daily headache frequently exacerbated to an intense hemicranial pain with few associated symptoms. The symptoms of neck strain and residual neck pain suggest that he had sustained neck trauma during the accident. It was unclear whether or not he had also sustained head trauma.

The patient's headache characteristics, the finding of occipital tenderness on examination, and the temporary headache relief after an occipital nerve block all support the diagnosis of occipital neuralgia (ON). In fact, the patient's headache meets the International Headache Society (IHS) criteria for ON (Table 1). It is noteworthy that the IHS does not define specific limits for the duration or frequency of the acute headaches of ON.

Although typically unilateral, ON can be bilateral.<sup>2</sup> Posterior head or neck trauma is a preceding event in some patients, but in other cases, no specific cause is found. Hypoesthesia in the distribution of the greater or lesser occipital nerves may be present. The IHS criteria state that ON should be distinguished from pain that originates in upper cervical spine structures (e.g. facet joints, ligaments). The general term that has been given to these conditions is cervicogenic headache, and it is included in the 2nd edition of

the IHS headache classification (ICHD-II). Distinguishing between cervicogenic headache and ON can be difficult, since both may be associated with neck trauma and both typically respond to greater occipital nerve (GON) block. A cervical spine MRI may help in differentiating between the two entities.

Cervical arterial dissection (of the vertebral or internal carotid artery) may result from neck trauma and cause unilateral headache. Typically, patients with internal carotid artery dissection have a Horner syndrome ipsilateral to the pain. This may or may not be accompanied by other focal neurological signs. A dissection of the vertebral artery may present with unilateral headache and cranial nerve palsies (e.g. diplopia due to ophthalmoparesis). An MRA of the neck will detect the dissection in the majority of cases. The long duration of headache in this case makes an arterial dissection unlikely.

The patient was given a diagnosis of bilateral hemicrania continua (HC). I question this diagnosis for a number of reasons: 1. HC, as the name implies, is typically a strictly unilateral headache without side shifts. Bilateral cases have rarely been reported. 2. The diagnosis of HC requires the presence of at least one symptom of cranial autonomic dysfunction (e.g. conjunctival injection, tearing, nasal congestion) ipsilateral to the pain, and this patient was not reported to have any of these symptoms. 3. The patient's headache did not resolve completely after treatment with indomethacin.

Occipital neuralgia is, therefore, the likely diagnosis in this case. GON block is the first-line treatment for ON.<sup>3</sup> The response to GON block is usually prompt, and headache relief may last days and even weeks after the anesthetic effect of the procedure subsided.

The mechanism of GON block's prolonged therapeutic effect on head pain is unclear, but it may be related to a modulating effect of the nerve block on afferent input that travels through the trigeminal system.<sup>4</sup> Tenderness at the area of the GON was found be a predictor of a good response to GON block, but anesthesia in the territory of the GON was not.<sup>5</sup>

Headache relief after GON block is an important feature of ON, but it is not specific, as GON block may effectively alleviate head pain of other headache disorders, such as migraine and cluster headache.<sup>6</sup> Therefore, the diagnosis of ON should not rely solely on a good response to GON block. Other headache features and findings on physical examination should be considered before making the diagnosis.

This patient had both lidocaine and a corticosteroid to block the GON. The use of corticosteroids in addition to local anesthetics when performing GON block for headaches is controversial. In a recent randomized study, adding triamcinolone to local anesthetics when performing GON block in patients with transformed migraine was not associated with improved outcome. In a controlled study of patients with cluster headache, however, GON block using a mixture of lidocaine and betamethasone was effective in preventing CH attacks, whereas lidocaine alone was not. Since injecting corticosteroids has been reported to cause local and even systemic adverse effects, I do not use them routinely when performing GON block for headaches.

Neurostimulation of the GON has recently gained attention as a potential treatment for patients with refractory headaches.<sup>3</sup> Several studies on its efficacy in treating various types of headache have reported positive results. However, these studies were small and

non-controlled. Data from on-going controlled studies on the effect of GON stimulation for this indication will hopefully clarify the role of this treatment for headaches.

This patient responded well to GON stimulation. Placement of a GON stimulator is a reasonable treatment choice, considering his favorable initial response to this treatment and his relatively short-lasting response to GON block.

### Table 1: ICHD-II criteria for occipital neuralgia

- A. Paroxysmal stabbing pain, with or without persistent aching between paroxysms, in the distribution(s) of the greater, lesser and/or third occipital nerves.
- B. Tenderness over the affected nerve.
- C. Pain is eased temporarily by local anaesthetic block of the nerve.

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