

## Resident and Fellow Section

*Morris Levin, MD, Editor*

### TEACHING CASE: OCCIPITAL NEURALGIA IN A YOUNG PATIENT

**Alexios G. Carayannopoulos, DO, MPH**  
Fellow in Pain Medicine, Department of Anesthesiology,  
Dartmouth Hitchcock Pain Fellowship Program

#### Case Presentation

A 27-year-old left-handed male, with medical history significant only for exploratory laparotomy for intussusceptions in childhood, was initially seen by the neurology service at our institution for a 10-year history of headaches. He was seen in consultation specifically for consideration of occipital nerve stimulator. He did have a history of 2 MVAs proximate to the time of headache onset. In the first, he was a restrained driver of a vehicle that rolled over, causing loss of consciousness for a few seconds without posttraumatic amnesia or other injuries. In the second, which was about a year after the first, there was no loss of consciousness, but he did suffer a neck strain with residual neck pain, which went away after several weeks. It was at this time he started to experience new daily persistent headache.

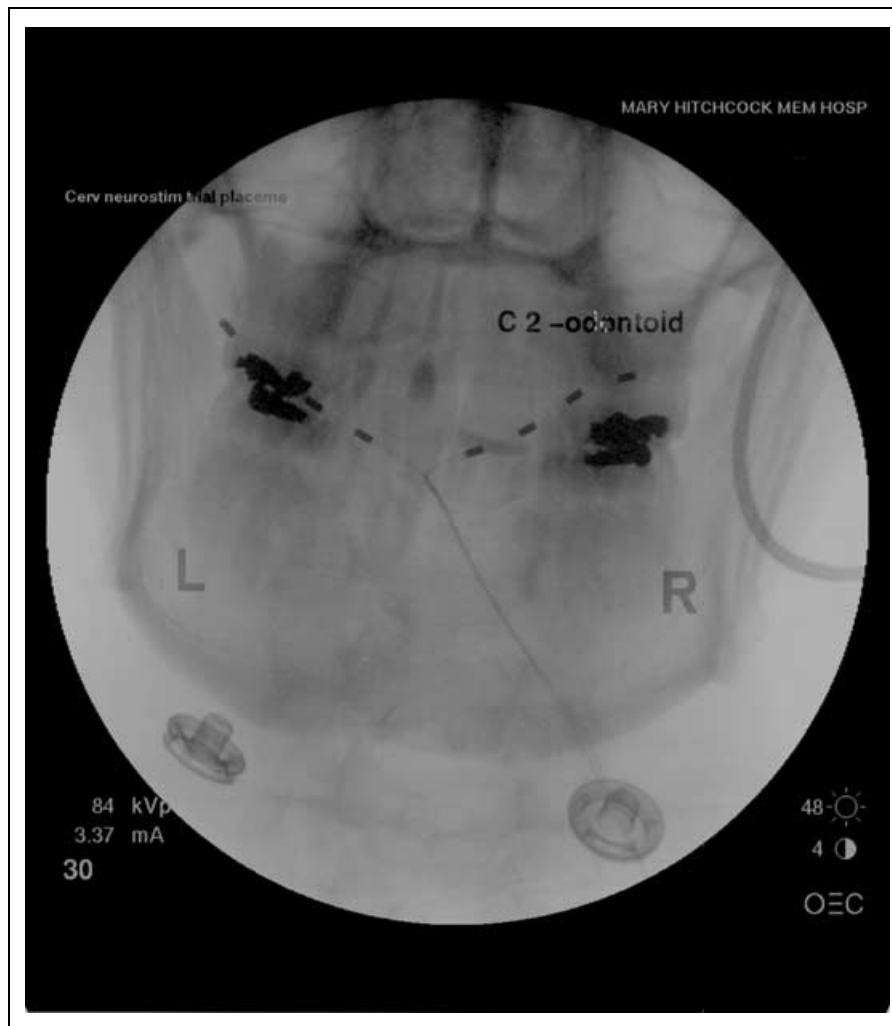
The headache quality was described as variable. The less intense headache, which was more common, was described as mild posterior pressure sensation located over the bioccipital-nuchal region. It had an average intensity of 1 to 2/10 on a verbal numeric score (VNS). The headache would exacerbate frequently to a more intense headache, which was almost always left hemicranial, rarely right, which was severe and sharp, without nausea, vomiting, aura, or prodrome, but with mild phono but no photophobia. This was rated as a 10/10 on VNS. Triggers included caffeine, sweets, onions, hot dogs, and laughing, which would trigger a headache within a few seconds, but not a true Valsalva. His symptoms were worse with heat or warm temperatures and watching television. He had a vision check and was prescribed new glasses for myopia, which helped his vision but had no effect on his headache. He had a cousin with similar symptoms, but no first degree relative. He did have a history of motion sickness, fainting, but did not have cold hands or feet. His sleep was good, mood was "okay," and energy level was appropriate. He worked as an installer of industrial air ducts and was a volunteer fireman.

Prior to being seen by Neurology at our institution, the patient was being managed by a neurologist in the community. His medications were eletriptan, indomethacin, nadolol, and topiramate. He was trialed on divalproex sodium, which exacerbated his headache, and was discontinued. On initial examination, his vitals revealed blood pressure on the low

range of normal, completely normal neurological, vascular, and musculoskeletal exams with the exception of tenderness bilaterally over the occipital nerves. Initial management included referral for an MRI examination to rule out Chiari malformation, and gradual reduction of nadolol. On follow-up examination, his BP was 86/51 with a pulse of 40, and his nadolol was tapered off. He continued to have tenderness bilaterally over the occipital nerves and underwent occipital nerve block using 1% lidocaine and 20 mg Depo-Medrol<sup>®</sup>. He was started on Indocin SR<sup>®</sup> for exacerbations and given famotidine for prophylaxis. He was tapered off topiramate. Patient was called the next day after block to report that he had no headache, but returned 6 days later with a lesser intensity. He then continued to have a low background daily headache with more intense exacerbations several times per week. He was then diagnosed with bilateral hemicrania continua and started on zonisamide. His baseline CBC and liver enzymes were normal. He continued to respond to indomethacin at 50 BID and his zonisamide was raised to 200 mg at bedtime. Based on the persistence of his symptoms, he was then referred to the Pain Clinic for evaluation and consideration for peripheral nerve stimulation.

Evaluation in the pain clinic revealed persistent tenderness in the occipital region bilaterally, left greater than right as well as tenderness over the spinous processes from C2 to C6-7 with a normal neurological and vascular examination. The patient and his wife were then presented with information regarding occipital nerve stimulation, including the risks and benefits of the procedure. They were explained that trial would be necessary to assess effectiveness followed by permanent placement. They elected to proceed with trial, feeling that something different needed to be attempted given persistence of symptoms and its effect on his function. Psychological clearance, as required by most insurance companies, was then obtained as was prior authorization from his insurance company.

The patient then underwent surgical placement of a percutaneously placed Medtronic "Pisces Quad" lead over the greater occipital nerve, bilaterally (see the Figure) without complication. The stimulation paresthesias were tolerable and provided excellent coverage of his headache bilaterally intra-operatively. The trial leads were then secured to the skin and connected to an external generator placed in a belt pack. The patient was admitted overnight for pain control, monitoring, and for education regarding the use of the system. Prior to discharge the next morning, his stimulator system was reprogrammed to optimize stimulator coverage of his occipital nerves bilaterally. He was instructed how to use the Medtronic "MyStim" remote control system and given limited control of certain parameters to customize stimulation during his trial period outside the hospital. He was



**Figure.—Bilateral occipital nerve leads.**

discharged the next day to home and was followed up in 1 week.

At follow-up, he reported no adverse side effect from the procedure. He had been given 2 days of oxycodone for post-operative pain, of which he only had to take 3 pills total. He noted that his continuous baseline headache over the bioccipital-nuchal region had broken. He was able to discontinue his daily indomethacin. He suffered only a few exacerbations of the more intense headache but stated was much less intense and only took a few doses of PRN indomethacin. Examination of his neck revealed no sign of infection or dislodgement of the stimulator leads. The leads were then removed and the site was cleaned and dressed. Based on the patient's successful trial, he elected to undergo permanent placement of the stimulator leads and implantation of the generator, which is pending at the time of this write-up. He was advised that he could restart his daily indomethacin as needed and remain on the topiramate and zonisamide.

Pending scheduling of the permanent implantation, the patient was asked to return 1 more time to address certain occupational issues. Because the patient had changed his

principal job and began working for a home heating company delivering oil to clients, which involved carrying a large hose over his shoulder, and because he continued to work as a volunteer fireman, he was asked to come back with all of his equipment and to don his gear in an effort to avoid any pressure points. A custom-sized catheter was then ordered based on these specifications. At the time of this write-up, permanent placement is still pending.

#### EXPERT COMMENTARY

**Avi Ashkenazi, MD**

**Assistant Professor of Neurology, Jefferson Headache Center, Thomas Jefferson University, Philadelphia, PA, USA**

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

**Table—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 anesthetic block of the nerve.
- 

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).<sup>1</sup> 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 (eg, 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).<sup>1</sup> 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 2 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 (eg, diplopia due to ophthalmoparesis). A magnetic resonance angiogram 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.<sup>1</sup> Bilateral cases have rarely been reported. (2) The diagnosis of HC requires the presence of at least 1 symptom of cranial autonomic dysfunction (eg, 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 to 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.<sup>7</sup> 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.<sup>8</sup> 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 noncontrolled. Data from ongoing 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.

## REFERENCES

1. Headache Classification Committee. The International Classification of Headache Disorders, 2nd edition. *Cephalalgia*. 2004;24:1-160.
2. Ashkenazi A, Levin M. Three common neuralgias. How to manage trigeminal, occipital, and postherpetic pain. *Postgrad Med*. 2004;116:16-4, 31.
3. Ashkenazi A, Levin M, Dodick DW. Peripheral procedures: Nerve blocks, peripheral neurostimulation and botulinum toxin injections. In: Silberstein SD, Lipton RB, Dodick DW, eds. *Wolff's Headache and Other*

- Head Pain*. New York: Oxford University Press (in press).
4. Bartsch T, Goadsby PJ. Stimulation of the greater occipital nerve induces increased central excitability of dural afferent input. *Brain*. 2002;125:1496-1509.
  5. Afridi SK, Shields KG, Bhola R, Goadsby PJ. Greater occipital nerve injection in primary headache syndromes—prolonged effects from a single injection. *Pain*. 2006;122:126-129.
  6. Ashkenazi A, Levin M. Greater occipital nerve block for migraine and other headaches: Is it useful? *Curr Pain Headache Rep*. 2007;11:231-235.
  7. Ashkenazi A, Matro R, Shaw JW, Abbas MA, Silberstein SD. Greater occipital nerve block using local anesthetics alone or with trimacinalone for transformed migraine: A randomized comparative study. *J Neurol Neurosurg Psychiatry* published online 6 August 2007 doi:10.1136/jnnp.2007.124420.
  8. Ambrosini A, Vandenheede M, Rossi P, Aloj F, Sauli E, Pierelli F, et al. Suboccipital injection with a mixture of rapid- and long-acting steroids in cluster headache: A double-blind placebo-controlled study. *Pain*. 2005;118:92-96.

#### QUESTIONS FOR DISCUSSION

1. What are the ICHD-II criteria for occipital neuritis? For cervicogenic headache?
2. What is the proposed pathophysiology of occipital neuritis?
3. What risks are there to GON block? To upper cervical root blocks? To occipital nerve stimulation?

This case presentation and discussion meets the ACGME requirements for residency training in the following core competency areas: Patient Care, Medical Knowledge, Practice Based Learning and Improvement, and Systems Based Practice.