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Case Information

Presenting Symptom: Left Occipital pain, headache

Case Specific Diagnosis: Left Occipital Neuralgia

Learning Objectives:

1. To develop an algorithmic approach to the patient with occipital head pain and develop a differential diagnosis.
2. To understand the diagnosis and workup of Occipital Neuralgia.
3. To understand the evidence for Occipital Nerve Stimulation for treatment of Occipital Neuralgia in refractory cases.

History:

59-year-old man with a history of CAD, adrenal insufficiency, depression, and pituitary adenoma that was resected in 2007 followed by cranial radiation with a total dose of 65 Gy, presents with left sided occipital pain. Over the subsequent 6 months, he developed left occipital pain which radiated over the left temporal and frontal regions to his eyes. He described his headaches as dull and aching, rating 7/10 average on the visual analog scale. Intermittently he felt an incapacitating, sharp and stabbing sensation over the left occiput. These headaches occurred daily, with a constant dull pain component that lasted 2-4 hours. His pain was worse at night, with aching and muscular tightness in the upper neck which interfered with his sleep. He denied any associated aura, but did have nausea and occasional photophobia. Pain was exacerbated by activity.

The patient denied any recent weight loss, fever/chills, night sweats, visual or hearing changes.

Pertinent Physical Exam Findings

He appeared in discomfort, but cranial nerves were all intact. He had full range of motion in the neck with normal rotation, extension and flexion.

Tinel's sign over the left occiput was positive, with tapping just lateral to the occipital protuberance evoking pain that radiated anteriorly over the scalp. There was mild tenderness to palpation over the occiput on the left side and Spurling's test was negative. There was no associated weakness. He had mildly diminished sensation to light touch over the left occipital scalp region, with cold allodynia on that side. DTR were all 2+ and motor function was normal.

Diagnostic Imaging and Results

MRI BRAIN without contrast

Impression: Normal MRI of the brain.

MR Neurogram of the bilateral Greater Occipital Nerves (GON)

Impression: 3T MRN demonstrating left GON neuropathy associated with with left occipital neuralgia, Coronal 3D PSIF and 8-mm-thick MIP reconstruction show an asymmetrically thickened and hyperintense left GON.

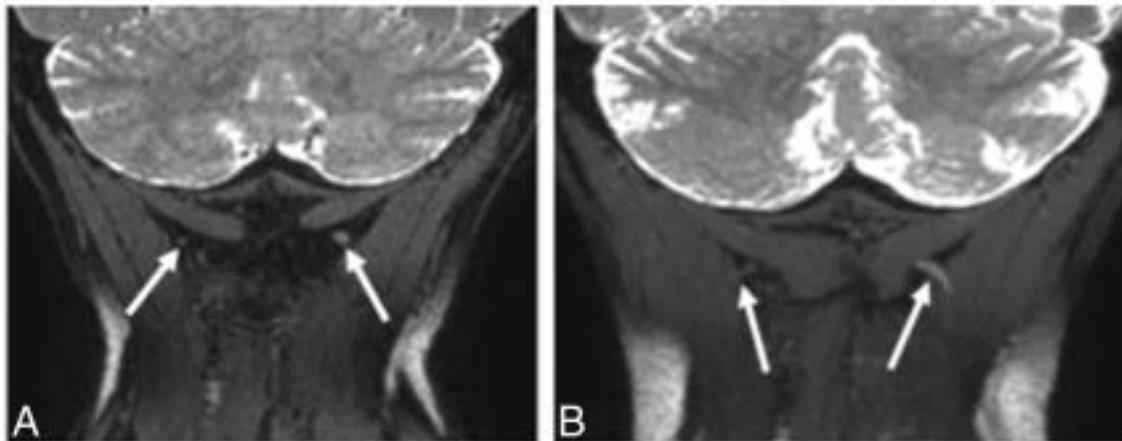
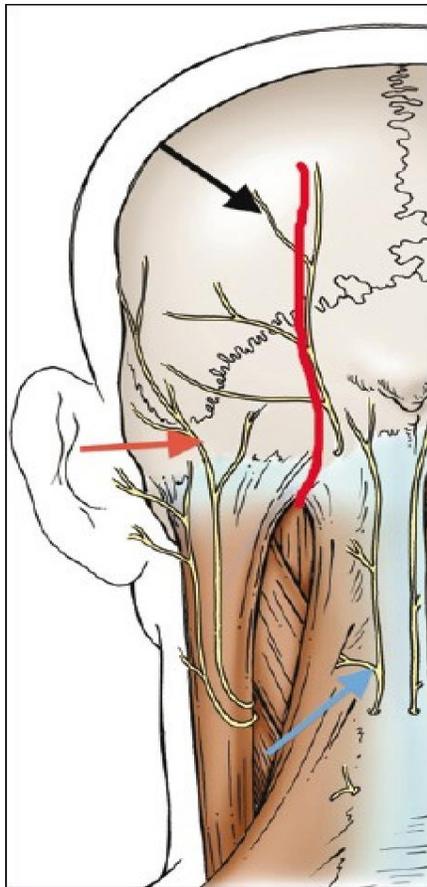


Figure 1 MR Neurogram of the bilateral Greater Occipital Nerves. The left greater occipital nerve demonstrates larger diameter and increased signal compared to the right, suggesting left greater occipital neuropathy (Hwang et al., 2017)



- Greater Occipital Nerve
- Lesser Occipital Nerve
- Third Occipital Nerve

Figure 2: Anatomy of the Greater, Lesser and Third Occipital Nerves (from Kemp et al, [Surg Neurol Int.](#) 2011)

Differential Diagnosis

1. Left Occipital Neuralgia, radiation induced.
2. Chronic migraine without aura
3. Cervical facet arthropathy at C2-3 level (Cervicogenic headache)
4. Paroxysmal Hemicrania (subtype of Trigeminal Autonomic Cephalgias)
5. Cervical radiculopathy at C2

Medications and Interventions:

These headaches were refractory to multimodal medical therapy including NSAIDs, TCA, SNRI, sodium channel blockers, triptans, beta blockers, acupuncture and biofeedback.

After presenting to the pain clinic, he underwent successful Left sided Greater Occipital Nerve block with 2.5cc of 1% lidocaine and 2.5cc of 0.25% bupivacaine distributed in a fan like fashion, just medial to the occipital artery. His pain improved from a 7/10 to 2/10 within 15 minutes, with analgesic relief lasting 2 hours. This confirmed left greater occipital neuralgia as a cause of his symptoms.

After discussion, he opted not to have Greater Occipital nerve radiofrequency ablation trial but rather decided to trial an occipital nerve stimulator given a desire for longer lasting relief.

Weeks later, the patient underwent percutaneous spinal neurostimulator electrode implantation under fluoroscopic guidance in the operating room for a 6 day stimulator trial. An 8 contact percutaneous SCS lead was advanced and placed at the left mastoid in the region of the greater, lesser, and least occipital nerves. Ultrasound was used to identify the depth of the lead needle and assess skull location. During the 6 day trial he reported pain relief ranging from 80-100%. He was able to sleep and “do light garden work”. He noted that his stimulator was working “relatively well” in preventing his headaches. VAS ranges were 1-3/10.

Evidence Based Indications for Occipital Nerve Stimulation in refractory Occipital Neuralgia

Diagnosis of Occipital Neuralgia.

Occipital Neuralgia (ON) is a common cause of headaches in the occipital region and is the third most common behind migraine and tension-type headaches (Headache Classification Committee of the International Headache Society (IHS), 2013; Liu et al., 2017). The greater occipital nerve arises from the dorsal ramus of the C2 nerve root, but the lesser or third occipital nerve may be involved as well. While the pathophysiology of most occipital neuralgia is uncertain, most hypotheses include damage to the C2-C3 nerve roots through various mechanisms (Elias and Burchiel, 2002). The most common causes include whiplash injury and C2 nerve compression from arthritic changes (e.g. facet arthropathy) (Liu et al., 2017). While 90% of cases of occipital neuralgia present unilaterally, bilateral ON has been reported. According to the International Classification of Headache Disorders 3rd Edition there are a set of signs and symptoms that must be met in order to diagnose this disease as follows:

The International Headache Society diagnostic criteria for occipital neuralgia (2013)

A. Unilateral or bilateral pain fulfilling criteria B–E
B. Pain is located in the distribution of the greater, lesser and/or third occipital nerves
C. Pain has two of the following three characteristics:
1. recurring in paroxysmal attacks lasting from a few seconds to minutes
2. severe intensity
3. shooting, stabbing or sharp in quality
D. Pain is associated with both of the following:
1. dysesthesia and/or allodynia apparent during innocuous stimulation of the scalp and/or hair

2. either or both of the following:
a. tenderness over the affected nerve branches
b. trigger points at the emergence of the greater occipital nerve or in the area of distribution of C2
E. Pain is eased temporarily by local anesthetic block of the affected nerve
F. Not better accounted for by another ICHD-3 diagnosis.

Other headache syndromes such as migraine or tension headache may produce similar pain as ON by acting through the trigeminal nucleus caudalis. A diagnosis of occipital neuralgia may be confirmed by pain relief from a greater occipital nerve block. Therapeutic options for this disease have been limited to conservative management with heat application and medications; persistent symptoms are often treated with repeat occipital nerve blocks (Ward, 2003). Nerve block of the GON can be performed together with the lesser occipital nerve (LON) for therapeutic effect, however the third occipital nerve may be missed and must be targeted separately.

Efficacy of Occipital Nerve Stimulation

As of Oct 2017, there has been no randomized controlled trial evaluating the efficacy of occipital nerve stimulation against placebo for ON. However, there have been multiple randomized controlled trials of occipital nerve stimulation for intractable chronic migraine which demonstrate >50% pain reduction (VAS) in 30-40% of patients (Dodick et al., 2015; Mekhail et al., 2016).

A recent case study reported successful treatment of bilateral ON with unilateral GON stimulation (Liu et al., 2017). The same study reviewed other retrospective studies combining 78 total patients with ON, of which slightly greater than half received >50% benefit from GON stimulation. More recently, a single center retrospective case series of 29 patients with ON reported a trial to permanent implant ratio of 69% for GON stimulation (20/29, (Keifer et al., 2017)). Of the implanted patients, 85% had more than 50% pain relief after 1 year (mean 410 days), with average pre-implant VAS scores of 7.4/10 falling to an average of 2.9 after implant.

Safety of Occipital Nerve Stimulation

Occipital nerve stimulation carries significant risk, with the main complication being lead migration or fracture given the location of the lead near the occipital protuberance. Other complications include skin erosion, infection or loss of effect; incidence of adverse events has not been accurately calculated owing to small sample sizes in most studies.

Treatment recommendations for this case:

This patient proceeded to left GON stimulator implant and his average VAS score at 3 months was 2.5. Given his large benefit from the trial, lack of other risk factors or contraindications, we recommended permanent implant of a GON stimulator.

Take home points

1. Occipital Neuralgia can be confused with migraine or cervicogenic headache because of common features (nausea, photophobia, neck pain).
2. Physical exam may reveal positive Tinel's sign, allodynia or musculoskeletal tightness in the splenius/trapezius regions which is usually unilateral (90%) but can be bilateral.
3. MR Neurogram may show an enlarged and hyperintense GON on the symptomatic side.
4. GON Block with local anesthetic may be diagnostic, but consider LON or third occipital nerve block if this fails.
5. There are no randomized controlled trials to support the use of occipital nerve stimulation to treat refractory ON, though careful retrospective case series provide level 5 evidence (Sackett criteria).
6. In carefully selected patients that have a response to GON, LON or third occipital nerve block, consider RFA or peripheral nerve stimulation trial for occipital neuralgia.

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