Ganglionectomy of C-2 for the treatment of medically refractory occipital neuralgia

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Occipital neuralgia is a result of neuropathic pain transmission in the distribution of the greater occipital nerve. Because it is well anatomically localized, occipital neuralgia has been the focus of various surgical treatments. Ablation, decompression, and modulation of the C-2 nerve have all been described as effective treatments. The C-2 dorsal root ganglionectomy provides effective treatment for this disorder with a low incidence of unpleasant side effects. In this review the authors summarize the current treatment of occipital neuralgia.

KEY WORDS • occipital neuralgia • ganglionectomy • headache • rhizotomy

Occipital neuralgia is characterized by pain confined to the distribution of the upper cervical nerve roots. This condition most commonly affects the greater occipital nerve and may be related to antecedent trauma. Although various terms have been used to describe the subjective nature of this discomfort, a lancinating or paroxysmal quality is characteristic.

Since the initial description of traumatic injury to the occipital nerve by Hunter and Mayfield in 1949,⁵ numerous treatments have emerged to manage occipital neuralgia. For patients in whom conservative management strategies have failed to resolve symptoms, ablative, decompressive, and neuromodulatory procedures can provide relief from chronic pain.

ANATOMY OF THE C-2 NERVE

The C-2 nerve rootlets emerge from the spinal canal surrounded by a dural sleeve, which terminates before the exit of the C-2 nerve root from the atlantoaxial interlaminar space. The extradural root then passes posterolaterally to lie beneath or just inferior to the C-1 arch. Here it becomes heavily invested within the vertebral venous plexus as it passes between the posterior atlantoaxial ligament and facet joint. Two to four millimeters after its dural exit,

the nerve root gives rise to the dorsal root ganglion, which averages 4.5 mm in width. The obliquus inferior muscle overlies the ganglion.

The ganglion gives rise to dorsal and ventral rami. The dorsal ramus contributes several branches, the largest of which is the greater occipital nerve. This nerve passes through the semispinalis muscle and then through the attachment of the trapezius muscle to the occipital bone. It then branches to innervate the scalp.

The greater occipital nerve provides cutaneous sensation to the medial posterior neck and scalp up to the coronal suture. Its dermatome extends laterally to the mastoid, where sensation is provided by the lesser occipital nerve. Although occipital neuralgia classically produces pain in this dermatomal distribution, nociceptive inputs appear to congregate with the spinal trigeminal nucleus. This may explain why some patients also experience retroorbital pain. Occipital neuralgia may also occur in the distribution of the lesser occipital nerve.

Occipital neuralgia has characteristic features that allow the clinician to distinguish it from other causes of neck and head pain. Pain is characteristically in the dermatomal distribution of either the greater or lesser occipital nerve. In two thirds of the cases the pain is unilateral. Descriptors of the pain that are predictive of a surgery-related good

result include those typically associated with neuropathic pain transmission: lancinating, electric, and shocklike pain is most characteristic.⁹ In cases being considered for surgical treatment, percutaneous nerve blocks with administration of local anesthetic agents can be useful for confirming the origin of pain.

ORIGIN OF PAIN IN OCCIPITAL NEURALGIA

Although the underlying cause of occipital neuralgia remains unclear, it is likely that there are various causes for abnormal neuronal activity. Whereas mechanical irritation of the nerve is a commonly proposed explanation, this entity has been associated with temporal arteritis,⁶ neurosyphilis,¹³ vascular compression,⁴ and herpetic neuralgia. Occipital neuralgia has also been reported to be associated with arthrosis of the C1–2 facet joint² and scarring from previous surgeries in the area.

The dorsal ramus of the C-2 nerve is unique in its anatomical relationship to neighboring osseous and soft-tissue structures, and it has been postulated that the C-2 nerve is susceptible to mechanical compression at three sites: 1) the exit zone between the cervical laminae, 2) the perforation of the atlantoaxial membrane, and 3) the tendinous portions of the trapezius muscle. Hunter and Mayfield⁵ proposed that because the C-2 nerve root and ganglion are unique in not being protected by surrounding bone, they are susceptible to mechanical trauma. Rotation and extension of the atlantoaxial joint was thought to irritate these nervous structures. This original explanation was concluded to be unlikely in normal patients by the authors of cadaveric studies who found that movement in this region did not cause nerve compression.¹ Degeneration of the spine producing abnormal articulations between C-1 and C-2, however, has been shown to produce mechanical nerve root compression. Atlantoaxial subluxation, as seen in cases of rheumatoid arthritis, can compress nervous structures between the laminae,³ and hypertrophy of the atlantoepistrophic ligament can entrap the exiting C-2 root.¹¹

Vascular engorgement of the vertebral venous plexus has also been postulated to cause transient occipital pain. In patients with this entity, painful exacerbations are associated with the Valsalva maneuver.⁴ Arterial compression of the C-2 root by an ectatic vertebral artery has also been described.¹²

TREATMENT STRATEGIES

The approach to patients with occipital neuralgia must initially be conservative. Frequently the symptoms will improve or resolve with therapy involving heat, rest, anti-inflammatory medications, and muscle relaxants. Oral anticonvulsant medications such as carbamazepine and gabapentin may also alleviate the pain. Patients with persistent symptoms may be treated with percutaneous injections of anesthetic and steroid medications. Nerve blockade is not only diagnostic but can also be therapeutic, often providing permanent pain relief. The opportunity to assess the patient's tolerance of an anesthetic scalp will also aid in determining if he or she will tolerate an ablative procedure.

Percutaneous neurolysis of the C-2 nerve root can be accomplished using ethyl alcohol.⁷ This technique has the advantage of avoiding the surgery- and anesthetic-related morbidities and can be an attractive option in patients who are poor surgical candidates. Pain recurrence can be problematic, however, and is seen in a large percentage of patients.

Peripheral neurectomy was the first treatment for occipital neuralgia.⁵ This procedure is simple and, because the nerve is isolated superficially, can be performed after injection of a local anesthetic. Initial pain relief, however, is durable in only approximately 50% of patients,¹⁰ and the recurrent pain is frequently more disabling than the initial symptoms.

The procedure involving removal of the C-2 ganglion was developed in response to the high failure rate of peripheral neurectomy. Postneurectomy recurrent pain and dysesthesias were presumably caused by axonal regeneration or neuroma formation. These problems are obviated by removal of the cell bodies that reside in the dorsal root ganglion. In a series of 39 patients treated at the University of Toronto, only one patient developed deafferentation pain after ganglionectomy, and this procedure was successful in treating the majority of patients in whom other ablative surgeries had failed.^{9,14}

In patients with subluxation of the atlantoaxial joint causing compression of the C-2 nerve reliable benefit can be achieved by performing nerve decompression and fixation of the joint.² Rheumatoid degeneration frequently leads to gross translation of the axis, which wedges the C-2 root between the osseous laminae. Posterior approaches such as C1–2 wiring- or transarticular screw-assisted fusion can relieve pain by correction of deformity followed by fixation. In cases in which the deformity cannot be adequately corrected, laminectomy is warranted to relieve pressure on the C-2 nerve.

Electrical stimulation of the greater occipital nerve has recently been performed to treat occipital neuralgia.^{8,15} Because destructive procedures for pain treatment are necessarily permanent, neural modulation is a theoretically attractive alternative. Because there are few case series demonstrating its efficacy, however, the results of this procedure remain unproven.

Fig. 1. Intraoperative photograph of the surgical exposure for C-2 ganglionectomy. 1 = C-1 lamina; 2 = C-2 spinous process; 3 = posterior atlantoaxial ligament; 4 = C-2 dorsal root ganglion; 5 = vertebral venous plexus.

Fig. 2. Photomicrograph of an excised dorsal root ganglion, showing afferent cell bodies and satellite nuclei. (H & E, original magnification $\times 40$).

CASE HISTORY

This 87-year-old woman complained of lancinating left-sided neck pain radiating to her forehead and orbit. This pain increased with neck flexion, extension, and rotation. The pain was rated as 10 on a 10-point scale. She did not complain of numbness or pain on the right side. After the failure of conservative treatments, she was referred for surgical treatment.

During surgery the patient was intubated and placed in the prone position with her head held in a Mayfield skull clamp. The neck was carefully flexed to expand the interlaminar space between C-1 and C-2. A 3-cm midline incision was made over the C-2 spinous protuberance. Dissection was taken through the nuchal fascia and down the C-2 spinous process and the C-1 and C-2 laminae unilaterally. A self-retaining retractor was placed to elevate the muscles off of the laminae (Fig. 1).

The C-2 nerve root was then seen laterally in the interlaminar space, exiting the vertebral venous plexus, which was cauterized with bipolar current. Careful separation of the nerve root from the venous plexus helped to minimize bleeding. There was no evidence of compression by the atlantoepistropheic ligament. Following the nerve laterally revealed the nerve root ganglion, which was seen as a dilation in the root. Distal to the ganglion the nerve was seen to divide into dorsal and ventral rami. Division of the root proximal and distal to the ganglion allowed complete excision of the afferent cell bodies (Fig. 2). There was no evidence of cerebrospinal fluid leakage (Video clip).

Click here to view video clip. Microsurgical procedure of C-2 ganglionectomy.

Postoperatively, the patient experienced numbness in the back of the left occipital region but no other neurological deficits. She was discharged to home on the 2nd postoperative day. She experienced good relief of her pain.

References

1. Bogduk N: The anatomy of occipital neuralgia. *Clin Exp Neurol* 17:167–184, 1981
2. Ehni G, Benner B: Occipital neuralgia and C1–C2 arthrosis. *N Engl J Med* 310:127, 1984
3. Hammond SR, Danta G: Occipital neuralgia. *Clin Exp Neurol* 15:258–270, 1978
4. Hildebrandt J, Jansen J: Vascular compression of the C2 and C3 roots—yet another cause of chronic intermittent hemicrania? *Cephalalgia* 4:167–170, 1984
5. Hunter CR, Mayfield FH: Role of the upper cervical roots in the production of pain in the head. *Am J Surg* 78:743–751, 1949
6. Jundt JW, Mock D: Temporal arteritis with normal erythrocyte sedimentation rates presenting as occipital neuralgia. *Arthritis Rheum* 34:217–219, 1991
7. Koch D, Wakhloo AK: CT-guided chemical rhizotomy of the C1 root for occipital neuralgia. *Neuroradiology* 34:451–452, 1992
8. Lou L: Uncommon areas of electrical stimulation for pain relief. *Curr Rev Pain* 4:407–412, 2000
9. Lozano AM, Vanderlinden G, Bachoo R, et al: Microsurgical C-2 ganglionectomy for chronic intractable occipital pain. *J Neurosurg* 89:359–365, 1998
10. Oh S, Tok S, Allemann J, et al: [Exeresis in occipital neuralgia.] *Neurochirurgia* 26:47–50, 1983 (Ger)
11. Poletti CE, Sweet WH: Entrapment of the C2 root and ganglion by the atlanto-epistropheic ligament: clinical syndrome and surgical anatomy. *Neurosurgery* 27:288–291, 1990
12. Sharma RR, Parekh HC, Prabhu S, et al: Compression of the C-2 root by a rare anomalous ectatic vertebral artery. Case report. *J Neurosurg* 78:669–672, 1993
13. Smith DL, Lucas LM, Kumar KL: Greater occipital neuralgia: an unusual presenting feature of neurosyphilis. *Headache* 27:552–554, 1987
14. Stechison MT, Mullin BB: Surgical treatment of greater occipital neuralgia: an appraisal of strategies. *Acta Neurochir* 131:236–240, 1994
15. Stojanovic MP: Stimulation methods for neuropathic pain control. *Curr Pain Headache Rep* 5:130–137, 2001

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