Microsurgical C-2 ganglionectomy for chronic intractable occipital pain

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Object. The authors evaluated the effectiveness of microsurgical C-2 ganglionectomy in 39 patients with medically refractory chronic occipital pain. In this procedure the neurons transmitting sensory inputs from the occiput are removed and, unlike peripheral nerve ablation, axonal regeneration is not possible.

Methods. The patients in this series had symptoms for 1 to 43 years. In 22 patients the occipital pain was caused by trauma; in 17 patients the pain was spontaneous. Pain relief failed in 17 patients who had undergone a previous occipital neurectomy or C-2 rhizotomy. Twenty-three patients experienced pain that was described as shocklike, electric, shooting, jabbing, stabbing, sharp, or exploding (Group I). Eight patients described their pain as dull, pounding, aching, throbbing, or pressurelike (Group II). The patients underwent unilateral or bilateral C-2 open microsurgical ganglionectomies.

The postoperative follow-up period ranged from 19 to 48 months. Nineteen patients experienced an excellent result (> 90% reduction in pain). Pain caused by trauma or that described using Group I terms responded best to ganglionectomy (80% good or excellent response). In contrast, the majority of the patients with nontraumatic pain or those described using Group II descriptors did not achieve favorable results.

Conclusions. The authors conclude that: 1) patients who suffer from chronic occipital pain after having sustained injury obtain worthwhile benefit from microsurgical C-2 ganglionectomy; 2) patients suffering from migraine, tension, and vascular headaches involving the occipital area are most often not helped by this operation; and 3) terms such as "shock," "electric," "shooting," "jabbing," and "sharp" used to describe occipital pain predict a favorable pain outcome following a C-2 ganglionectomy.

KEY WORDS • occipital neuralgia • spinal nerve root • ganglionectomy • headache • anesthetic block • rhizotomy

Chronic pain syndromes in the region of the occiput have multiple origins and a long and complicated history of surgical intervention. One disorder, occipital neuralgia, consists of a paroxysmal occipital pain extending from the suboccipital region to the vertex of the head.22,23 The quality of pain is frequently described as "sharp," "shooting," "stabbing," and "electric," terms that are usually associated with a neuropathic cause.6,27 It can be hypothesized that the pain is the consequence of abnormal somatosensory processing caused by injury or dysfunction of C-2 sensory neurons at the level of the perikarya or axons. In contrast, in occipital pain associated with migraine or tension headache, or in pain with a possible cervicogenic cause, pain is frequently described as "diffuse," "aching," "throbbing," or "pressurelike." These descriptors are consistent with a nociceptive origin1 and suggest that the pain may arise from a primary pathological disorder in soft tissues or vascular and bony structures that lie within the dermatome.

Numerous causes of occipital pain have been proposed including congenital malformations,28 neoplasms,29 degenerative disease of the spine,3,30,33 and disorders of the peripheral nervous system caused by metabolic,9,10 inflammatory,14,15 and infectious26 conditions. Of particular interest, as a consequence of its anatomical peculiarities, the C-2 sensory neurons and axons may be unusually vulnerable to mechanical injury and/or entrapment.1,19,23 The C-2 ganglion and nerve root, which lie outside the spinal canal between the arch of C-1 and the lamina of C-2, may be vulnerable to crush injury during traumatic hyperextension. The most common example of this type of injury is "whiplash," which is caused by a rear-end motor vehicle accident.13 Furthermore, the C-2 ganglia and posterior root are in direct contact with the lateral edge of the posterior atlantoaxial ligament (Fig. 1), which, when hypertrophied, is thought to cause C-2 ganglion entrainment.17,19,25. Enlargement of the extensive venous plexus that envelops the C-2 ganglia has also been implicated as a potential source of compression injury16 as have ectatic arteries.28 Peripherally, potential sites of entrapment are the points at which the greater occipital nerve passes through the semispinalis capitis or the aponeurotic attachment of the sternocleidomastoid muscle at the nuchal line.1

Surgical intervention for chronic occipital pain has been
undertaken when a headache is resistant to medical therapy and is no longer alleviated by nerve block(s) created by local anesthetic and/or steroid medications. Surgical approaches include greater occipital nerve neurectomy,\textsuperscript{15} percutaneous C-2 rhizolysis,\textsuperscript{21} C2-3 facet rhizolysis,\textsuperscript{33} dorsal root rhizotomy,\textsuperscript{8} C1-2 decompression,\textsuperscript{29} and C-2 ganglionectomy.\textsuperscript{33,32} The success of these procedures has been variable. Occipital neurectomy is a simple procedure and has the advantage of being easily performed while the patient receives local anesthesia. Neurectomy, however, can be followed by axonal regeneration with the potential for pain recurrence and, thus, the results of occipital neurectomy can be disappointing. Early postoperative recurrence of pain is frequent\textsuperscript{7} and the development of dysesthesia, hyperpathia, and allodynia, presumably due to sensory terminal regeneration and/or neuroma formation, has been reported.\textsuperscript{53} In addition, occipital neurectomy leaves the proximal C-2 elements, which can be the site of the pathological disorder, intact and free to generate ongoing dysfunction.

The C-2 ganglionectomy may offer certain advantages over competing neurosurgical pain procedures. Dorsal root ganglionectomy removes the primary sensory neuronal cell bodies. There is no possibility of axonal regeneration and no sparing of aberrant central axonal processes such as those entering the spinal cord through the posterior root,\textsuperscript{53} which could occur with posterior rhizotomy. The C-2 ganglion can be easily exposed microsurgically involving minimal bone resection. Indications for cervical ganglionectomy, however, are not well established and previous anecdotal reports involving few patients show variable results.\textsuperscript{97} To evaluate the safety and efficacy of C-2 ganglionectomy for the treatment of medically refractory chronic occipital pain, we have retrospectively analyzed outcome in 39 patients with neuralgic or nonneuralgic occipital pain who underwent this procedure. A preliminary report of this study was presented in abstract form.\textsuperscript{23}

**Clinical Material and Methods**

**Patient Population**

Patients were referred for the treatment of their medically refractory occipital pain. Medical history was gathered and physical examination was conducted. Patients were examined by means of cervical spine radiographs and magnetic resonance imaging of the craniovertebral area at the discretion of the neurosurgeon.

Pain was evaluated preoperatively by using a visual analog pain scale that rated each patient’s pain from 0 to 10. Patients were asked to describe their pain. These descriptors were assigned to one of two groups on the premise that the terms “shocklike,” “electric,” “shooting,” “jabbing,” “stabbing,” “sharp,” or “exploding” indicated a neuropathic origin (Group I) and that the terms “dull,” “pounding,” “aching,” “throbbing,” or “pressurelike” (Group II) indicated a nonneuropathic cause. Most patients had undergone a diagnostic local anesthetic block prior to their referral to neurosurgery. This block was performed under fluoroscopic control by a radiologist who used 1 ml of 0.25% marcaine deposited at the C-2 ganglion. The results of the block were not used in preoperative decision making.

Unilateral or bilateral C-2 ganglionectomy was performed in 39 patients with intractable occipital pain between 1990 and 1992. In all patients medical therapy had failed and in more than one-half of the patients a previous surgical intervention to alleviate the pain had failed. The exclusion criteria for surgery were: 1) inadequate medica-
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Evaluation of Surgical Outcome

The relevant surgical anatomy is depicted in the photograph shown in Fig. 1. Anesthesia was induced in the patients and they were orotracheally intubated and positioned prone with the head flexed in a Mayfield headrest. A midline incision extending from the base of the occiput to C-3 was made through the skin and subcutaneous tissues. The muscle attachments to the spinous processes and laminae of C-1 and C-2 were incised and retracted laterally. The vertebral artery was carefully avoided throughout the dissection. With the aid of the operative microscope, the robust epidural venous plexus enveloping the C-2 ganglion was incised and coagulated. It was necessary to remove the inferior border of the C-1 lamina to expose the C-2 ganglion in approximately one-half of the cases. The C-2 ganglion was identified as an area of focal increase in girth proximal to the C-2 primary rami. The variable fused dural/epineural sleeve containing the dorsal rootlets of the C-2 ganglion and the anterior C-2 rootlets was dissected proximally. Distally, the dorsal and ventral primary rami of the C-2 root were followed laterally by dissecting the venous plexus by using a right-angled hook, bipolar cautery, and sharp dissection. The neural elements immediately proximal and distal to the C-2 ganglion were cauterized and cut with a No. 15 blade and the ganglia were excised. In no case was there an apparent opening into the subarachnoid space or a release of cerebrospinal fluid. Hemostasis was maintained using cautery and, in some cases, gelfoam. The fascia and skin were closed in a standard fashion. Bilateral ganglionectomies were performed in a similar manner. Patients were extubated in the operating room and taken to the recovery room.

Following surgery, patients remained in the hospital for approximately 2 days (range 1–10 days). They were seen at a follow-up visit 4 weeks after surgery and their cases were followed for a median of 28 months (range 19–72 months).

Surgical Procedure

The origin of the occipital pain was traumatic in 22 patients (67%) with a median age of 43 years (range 35–60 years). The duration of symptoms prior to ganglionectomy in this group was 15 years. The remaining eight patients (24%) with nontraumatic occipital pain reported a relationship between the onset of pain and the following precipitating factors: allergic reaction, Ménière’s disease, diagnostic lumbar spinal myelogram, vaginal delivery of a full-term infant, cervical anterior discectomy and fusion, and ankylosing spondylitis, each in one patient; and no identifiable precipitating event in two patients. These patients had symptoms ranging in duration from 13 to 43 years with a median time to ganglionectomy of 18 years.

In the 39 patients, the medical history was significant for hypertension controlled by medication in two patients, peptic ulcer disease in four patients, and breast cancer treated by mastectomy in one patient. Three patients had undergone spinal surgery before the onset of their pain syndrome: two had lumbar spinal fusion after trauma and one had anterior cervical surgery for a C-6 radiculopathy. Of the 39 patients in this series, three patients had undergone previous cranial procedures. In one of these cases, which involved ventriculoperitoneal shunt placement, the occipital pain was likely related to the surgery and in the other two it was considered unrelated and classified as spontaneous.

The occipital pain was located at the level of the upper cervical spine and radiated over the occiput to the vertex in all patients. In 27 patients (69%) pain also radiated to the supraorbital, retroorbital, or frontotemporal region. Of these, 16 patients (53%) had a traumatic cause of their pain syndrome. The pain was unilateral in 24 (62%) of the 39 patients. Patients suffering from trauma were more likely to have unilateral pain (16 [73%] of 22). In contrast to patients with pain arising from trauma, patients with...
pain due to spontaneous causes were more likely to have bilateral pain (nine [53%] of 17).

Descriptors of Pain Syndromes
In 23 patients (59%), the occipital pain was described as sharp, shooting, stabbing, jabbing, electric, or exploding (Group I descriptors). The causes of injury in this group were traumatic in 17 patients (74%) and spontaneous in six (26%). In eight patients (21%) the occipital pain was described as dull, aching, throbbing, or pressurelike (Group II descriptors). In these patients, the cause was spontaneous in seven patients (88%) (six of seven patients in the migraine subgroup) and traumatic in one (nonmotor vehicle accident [13%]). Of note, no patient whose occipital pain was precipitated by a motor vehicle accident was categorized by Group II descriptors exclusively. A mixture of the descriptors was reported for eight patients (four [50%] who suffered from traumatic injury and four [50%] whose pain was spontaneous).

Diagnostic Studies and Previous Treatment
Radiological studies were available for 25 patients, 13 of whom exhibited abnormalities on cervical spine x-ray films. The most common findings were mild-to-moderate osteoarthritic changes in the facet joints and degenerative disc disease. One patient had undergone a cervical fusion prior to the injury that precipitated the occipital pain.

Almost all patients responded to local anesthetic therapy prior to C-2 ganglionectomy, although the duration of effect was highly variable. A diagnostic C-2 ganglionic block consisting of 1 ml of 0.25% marcaine injected under fluoroscopic guidance had a positive effect (> 50% pain relief) in 38 patients (97%).

Sixty-two percent of the patients (24 of 39) had undergone at least one surgical procedure as treatment for occipital pain before they underwent C-2 ganglionectomy (Table 1). Percutaneous C-2 rhizolysis had been performed in 13 patients (33%), in equal frequency for the traumatic and spontaneous pain groups. The outcome of C-2 rhizolysis was variable. Seven patients (54%) reported transient reduction or relief of pain lasting between 3 weeks and 6 months. Four patients (31%) had no relief of their pain and two patients (15%) reported an increase in pain after C-2 rhizolysis. Percutaneous facet joint rhizolysis was achieved in only 11% of patients who suffered from trauma (nine [53%] of 17), whereas 62% of the patients whose pain was caused by trauma had unilateral symptoms and unilateral surgery. There were no instances of intraoperative or postoperative cerebrospinal fluid leaks and the brisk bleeding that was often encountered from the epidural venous plexus enfolding the C-2 ganglion was readily controlled by a combination of bipolar coagulation and application of gelatin sponges and gentle pressure. There were no operative complications in any patient. Histological analysis of the surgical specimens revealed normal ganglia and nerves in 36 patients. In two patients, mild neuronal loss and mild fibrosis were noted.

The results of C-2 ganglionectomy are shown in Fig. 2. Overall 19 patients experienced an excellent result (> 90% reduction in pain), seven patients had a good result (50–90% reduction in pain), and 13 patients experienced treatment failure (< 50% reduction). Greater than 90% reduction in occipital pain was achieved in 64% of trauma patients compared with 29% of patients in whom the cause was nontraumatic or spontaneous (p < 0.05). The procedure was clinically successful (> 50% reduction in pain) in 82% of patients with occipital pain resulting from trauma, which was significantly higher (p < 0.05) than the percentage of patients with spontaneous origin (47%). Seventy-three percent of patients involved in a motor vehicle accident reported greater than 90% pain relief compared with 43% of patients who suffered trauma from another cause (p < 0.05; Fig. 3 upper). An excellent result was achieved in only 11% of patients who suffered from trauma.

Table 1: Prior surgical management of occipital pain in 24 patients who underwent microsurgical C-2 ganglionectomy

<table>
<thead>
<tr>
<th>Procedure &amp; Outcome</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>C-2 rhizolysis</td>
<td>13 (33)</td>
</tr>
<tr>
<td>transient relief</td>
<td>7 (54)</td>
</tr>
<tr>
<td>no relief</td>
<td>4 (31)</td>
</tr>
<tr>
<td>increased pain</td>
<td>2 (15)</td>
</tr>
<tr>
<td>facet rhizolysis</td>
<td>16 (41)</td>
</tr>
<tr>
<td>transient relief</td>
<td>8 (50)</td>
</tr>
<tr>
<td>no relief</td>
<td>8 (50)</td>
</tr>
<tr>
<td>occipital neurectomy</td>
<td>4 (10)</td>
</tr>
<tr>
<td>transient relief</td>
<td>3 (75)</td>
</tr>
<tr>
<td>no relief</td>
<td>1 (25)</td>
</tr>
<tr>
<td>transcutaneous stimulation</td>
<td>2 (5)</td>
</tr>
</tbody>
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Fig. 2. Bar graph depicting responses to C-2 ganglionectomy. The trauma pain group included 22 patients and the spontaneous pain group 17 patients. Results are expressed as percentages of total patients in each group who achieved the following responses: excellent (> 90% pain relief); good (50–90% pain relief); or fail(ed) treatment (< 50% pain relief).
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migraines compared with 50% of patients with pain due to a spontaneous cause other than migraine (p < 0.05; Fig. 3 lower). The highest failure rate was found in patients with migraines (67%), double the rate in patients with nonmigraine spontaneous syndromes and more than triple that in patients whose pain was due to trauma.

Response to C-2 ganglionectomy analyzed by patient grouping according to the use of pain descriptors is presented in Fig. 4. Patients who described their pain as shooting, stabbing, burning, or exploding (Group I descriptors) had a 78% success rate, with 70% reporting an excellent result. In contrast, patients whose pain was characterized as dull, aching, or throbbing (Group II descriptors) had a 63% failure rate and none of these patients achieved an excellent result with ganglionectomy. Use of both descriptor groupings predicted a 63% success rate (≥ 50% pain relief).

Although the C-2 anesthetic block had a positive effect in 97% of the patients in this series, ganglionectomy was also undertaken in the one patient who did not respond to the block because the occipital pain was temporally related to a rear-end motor vehicle accident. Interestingly, ganglionectomy had no effect on her pain syndrome.

A potential complication of the surgical management of occipital pain is the production of a deafferentation syndrome, characterized by hyperpathia and allodynia in the C-2 or C-3 dermatome. In the current series, seven patients presented with sensory loss and a component of burning pain suggestive of a deafferentation syndrome prior to C-2 ganglionectomy. Four of 13 patients developed deafferentation pain after percutaneous C-2 rhizolysis (incidence of 31%), one after occipital neurectomy, and one after alcohol injection into the occipital nerve; in one patient deafferentation pain was an initial feature of the pain syndrome. Among these cases, six were relieved by ganglionectomy. At last follow-up evaluation, only one case of deafferentation occurred as a result of ganglionectomy (2.6% incidence).

Discussion

The present study shows that C-2 ganglionectomy provides good-to-excellent reduction in chronic occipital pain in selected patients. The character of the pain and the words used to describe the pain are important predictors of the response to C-2 ganglionectomy. Patients who described their pain by using neuropathic terms, such as shocklike, electric, shooting, jabbing, stabbing, sharp, or exploding, had better results than those who described their pain in terms that suggest nociceptive activation, such as aching, pounding, throbbing, or pressurelike.

More than 80% of patients with trauma as a precipitant achieved good or excellent results. In contrast, in patients with nontraumatic occipital pain the results were poorer. More than 50% of these patients did not experience worthwhile improvement (> 50% reduction in pain) with their surgery. This was particularly the case in patients with a history of migraine headaches in whom surgery produced a satisfactory result in only 30%.

The majority of patients in this series had undergone a previous surgical attempt at treating their pain. Previous

FIG. 3. Bar graphs showing responses to C-2 ganglionectomy with respect to the role of pain origin. Upper: Trauma pain group: 15 patients in the motor vehicle accident (MVA) subgroup and seven patients in the nonmotor vehicle accident (other) group. Lower: Spontaneous pain group: nine patients suffering from migraine and eight patients with pain from another nontraumatic cause. Results are expressed as percentages of total patients in each group who achieved the responses excellent, good, or fail(led) treatment defined in the legend to Fig. 2.

FIG. 4. Bar graph showing responses to C-2 ganglionectomy with respect to the role of descriptors. There were 23 patients in Group I (pain described as sharp, stabbing, jabbing, electric, and so forth) and eight patients in Group II (pain described as dull, aching, throbbing, pressurelike). Results are expressed as percentages of total patients in each group who achieved the responses excellent, good, or fail(led) treatment defined in the legend to Fig. 2.
failure to respond to surgical treatment of occipital pain (C-2 rhizolysis, occipital neurectomy, or facet denervation) was not a predictor of the response to ganglionectomy.

In those cases in which C-2 ganglionectomy produced pain relief, it was immediate and generally sustained for the follow-up period. Similarly, those patients who did not respond experienced immediate treatment failures rather than an initial response that was subsequently lost. The reasons for failures of C-2 ganglionectomy are not clear. The most likely explanation is the persistence of pain generators outside the C-2 dermatome. The immediate time course of the surgical failures argues against pain recurrence secondary to neural sprouting from adjacent nonlesioned neural elements.

It is clear that the response to a nerve root local anesthetic block cannot be used to predict the outcome of C-2 ganglionectomy and we cannot recommend its use, as practiced in this study, in surgical decision making. How can one reconcile the apparently contradictory results in the present series: 38 of 39 patients reported significant relief of occipital pain with a C-2 nerve root block; however, only 26 of the 38 patients reported satisfactory pain relief after C-2 ganglionectomy? One possibility is that the local anesthetic block may extend beyond the intended sites. Spread to adjacent nerves, particularly the first and third cervical nerves and fibers of the semicapitais muscle, is possible. Alternatively, it is possible that the effect of the local anesthetic medication is exerted in part at the level of the spinal cord and not just the peripheral nerve. The clinical effectiveness of systemic marcaine- or xylocaine-mediated analgesia at doses below those necessary to produce conduction block in A-δ or C fibers is well documented. In addition, the effects of occipital anesthetic blocks can, in some cases, far outlast the expected biologic life of the anesthetic agent. The mechanism by which these analgesic effects are produced is poorly understood. It has recently been reported that in certain patients with pain, close to 60% of the efficacy of the local anesthetic nerve block in patients may be due to a placebo effect.

Conclusions

The results of the present study demonstrate that the syndrome of chronic occipital pain must be subdivided by origin to predict more accurately the response to C-2 ganglionectomy in patients who have experienced failed medical and/or prior surgical therapy. From our data, it can be predicted that C-2 ganglionectomy will be successful (≥ 50% reduction in pain) in 80% of patients with chronic occipital pain with an antecedent history of trauma. Patients who suffer whiplash injury from a rear-end motor vehicle accident can expect a 70% chance of reducing their pain by greater than 90%. This is in contrast to an 11% chance of the same degree of pain reduction in patients with migraine headaches. This disparity in outcome may, in part, be reflected in the underlying pathophysiological mechanisms of the pain. Patients with whiplash injury use neuropathic descriptors to characterize their pain, whereas patients with mirmage use the classic nociceptive descriptors. As shown here, successful outcome after C-2 ganglionectomy approaches 80% when neuropathic descriptors are used, but is less than 40% for patients describing nociceptive pain. Patients with occipital pain of traumatic origin, in particular those with whiplash injury who describe neuropathic pain, are most likely to benefit from C-2 ganglionectomy.

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References

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