Occipital neuralgia secondary to hypermobile posterior arch of atlas

Case report

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The authors report on the management of occipital neuralgia secondary to an abnormality of the atlas in which the posterior arch was separated by a fibrous band from the lateral masses, resulting in C-2 nerve root compression. The causes and treatments of occipital neuralgia as well as the development of the atlas are reviewed.

KEY WORDS • occipital neuralgia • atlas fracture

Occipital neuralgia is becoming increasingly recognized as a cause of posterior auricular and lateral suboccipital pain. Sometimes difficult to diagnose, the main clinical issue centers on proper identification of the “pain generator.” Myriad clinical scenarios may represent occipital neuralgia. These include sclerosis of the atlantoaxial facet, direct tethering of the greater occipital nerve, and indirect irritation of the nerve secondary to muscle spasm.

Whenever occipital neuralgia is present clinically, the surgeon must look for the lesion causing the pain. We present a case of occipital neuralgia secondary to a hypermobile posterior arch of C-1, which was diagnosed preoperatively and confirmed intraoperatively. Although uncommon, this entity should be included in the differential diagnosis of posterior auricular pain.

Case Report

Presentation. This 38-year-old man, employed as an electrician, experienced the gradual onset of retroauricular and occipital pain 6 years prior to presentation. His symptoms progressively worsened until he experienced them daily. At presentation he described the symptoms as constant dull pain in the occipital and retroauricular area associated with lancinations radiating to the vertex. The pain was aggravated by movement of his neck. He denied a history of traumatic injury immediately prior to onset of symptoms, but remarked that he was involved in a motor vehicle accident without sequelae 10 years prior to presentation.

Examination. Plain radiography and computerized tomography scanning of the occipitocervical junction revealed a bilateral symmetrical, tapered disconnection of the posterior arch of the atlas in the region of the lateral masses (Fig. 1). Magnetic resonance imaging of the brain and cervical spine demonstrated normal anatomy. Flexion–extension dynamic radiography did not demonstrate abnormal motion. The patient received nonsurgical therapy consisting of analgesic agents, acupuncture, and transcutaneous electrical nerve stimulation but experienced no relief. He underwent four courses of selective C-2 nerve root block, which resulted in 80 to 100% relief of pain for up to 3 months following each block.

Neck motion was limited secondary to pain. Extension produced pain in the retroauricular region of C-2. Sensation was decreased to pinprick in the distribution of the greater occipital nerve bilaterally. Otherwise his status was normal.

Operation. The posterior elements of the atlas and axis were exposed, and the posterior arch of the atlas appeared normal until lateral exposure revealed tapering ends attached to fibrous bands passing anterolaterally. The C-2 nerve roots passed directly inferior to these fibrous attachments and were compressed by the caudal movement of a very hypermobile posterior arch. The fibrous bands were severed, and the posterior arch of the atlas was removed. Pathological examination revealed normal bone without the presence of callus.

Postoperative Course. The patient experienced immediate relief of his occipital pain and the range of motion of his neck was increased. He has remained free of symptoms during follow up of more than 1 year.
Occipital neuralgia

Discussion

Occipital neuralgia is classically characterized by intermittent episodes of lancinating pain extending from the suboccipital region up to the cranial vertex and often superimposed on a dull, constant pain. Nausea, dizziness, vertigo, stiff neck, photophobia, and blurred vision may also be experienced, as may tenderness of the greater occipital nerve as it crosses the nuchal line.

The greater occipital nerve is formed by the medial branch of the C-2 dorsal ramus, with contributions from the C-1 and C-3 nerve roots. Whereas lesions of C-3 may cause occipital pain, those affecting C-4 and below are much less likely to do so. Irritation of the upper cervical nerve roots can also lead to pain in the frontal, periorbital, and retroorbital regions. Fibers of the upper cervical dorsal root ganglia can synapse near the nucleus caudalis of the trigeminal nerve, allowing a lesion of the C-2 nerve root to be perceived as pain in the distribution of the trigeminal nerve.

Occipital neuralgia has been associated with many different processes that produce compression or irritation of the posterior primary C1–3 rami. Many authors believe that the greater occipital nerve is susceptible to compression as it emerges from the tendinous fascia of the semispinalis capitis. Other implicated causes of occipital neuralgia include trauma-induced scars, fracture pseudarthrosis, neurosyphilis, degenerative joint disease, primary and metastatic tumors, Chiari malformation, fibrositis, myositis and temporal arteritis, and vascular compression by an anomalous vertebral artery. Nonsurgical therapy includes oral analgesics, anticonvulsants, antibiotics, cervical collars, cervical traction, electrical nerve stimulation, heat, massage, and injections of local anesthetics or neurolytic agents. Injections are given at the area of greatest tenderness along the nuchal line or at the atlantoaxial joint.

Surgical treatments are directed at lesioning or decompressing the C1–3 nerve roots, dorsal rami, or the greater and lesser occipital nerves. Complete rhizotomy of the C1–3 dorsal rootlets eliminates the pain of occipital neuralgia but is associated with scalp anesthesia or dysesthesia. A selective posterior rhizotomy may avoid these complications. In decompressive surgeries the surgeon focuses on the atlantoepistrophe ligament, posterior decompression of the C1–3 roots, suboccipital decompression for tonsillar herniation, and fusion of spinal segments in cases of atlantoaxial osteoarthritis.

Ossification of the atlas occurs in a cartilage primordium. Ossification centers are found in each lateral mass, and each center extends posteriorly into the posterior atlantal arch. In our case, the compressive entity either represented a congenital nonunion of the atlas ossification centers or a pseudarthrosis that developed after an atlas fracture. Given that the atlas forms with two major lateral ossification centers with a midline posterior synchondrosis (Fig. 2), a congenital cause is unlikely, and this case probably represents nonunion of an atlantal posterior arch fracture caused by traumatic injury. A resultant nonfixed, hypermobile arch compressed the C-2 nerve roots, producing occipital neuralgia. Resection of this hypermobile segment relieved nerve compression and pain.

A spectrum of treatment options can be used for occipital neuralgia of unknown causes. However, when occipital neuralgia is demonstrated, an attempt should be made to identify an anatomical abnormality that may be amenable to surgical treatment. Decompressive surgery in such situations can yield satisfactory results, as in the case presented here.

References

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