Occipital neuralgia and the C1–2 arthrosis syndrome

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Occipital neuralgia syndromes have been ascribed to a great many pathological alterations, some demonstrable and some hypothetical. Recently, occipital neuralgia has been attributed to developmental and posttraumatic lesions in the cervicocranial junction region, with the nerve roots at C-1 and C-2 considered to be the principal pain pathways. The authors describe a series of seven patients with an upper neck and occipital pain syndrome due to unilateral degenerative disease (arthrosis) of a C1–2 lateral articulation. Two of the cases are presented in detail. This disease is demonstrable by radiography through the open mouth by isotope bone scanning, and by computerized tomography scanning. Temporary relief may be obtained by anesthetic and steroid injection, and permanent relief achieved by C-2 dorsal rhizotomy.

KEY WORDS • occipital neuralgia • C1–2 arthrosis • cervical spine • pain

The first reference to a neuralgic syndrome of the occipital region is said to have been made in 1821 by Beruto y Lentijo and Ramos. More recently, Luff in 1913 and Osler and McRae in 1925 ascribed the syndrome to fibrositis and other organic processes. Other authors identified cervical arthritis as being responsible for certain suboccipital headaches. Hadden thought occipital neuralgia a neglected subject in 1940 and proposed, as Luff had before him, that peripherally situated abnormalities could be the cause. These included the sites where the occipital nerves penetrate the points of insertion of the trapezius muscle on the skull and are therefore subject to the effects of fibrositis and the consequences of infections of the paranasal sinus and other focal infections. Postural defects, and the effect of wearing heavy hats, of hair pulled tightly around the head, of tight headaddresses such as those worn by nuns, and of blood seepage into the subgaleal space after head trauma were also proposed as sources of pain. Cervical arthritis was deemed a cause, and several clinical varieties of occipital neuralgia were described.

In recent years, deeper-lying and more threatening lesions, including the Arnold-Chiari malformation, primary and metastatic tumors, mastoid and intraspinal infections, trauma, and metabolic disorders, including diabetes and gout, have been reported to be causes of occipital pain, sometimes with frontal and orbital extension. Several writers have devised classifications incorporating scores of causes, and one mentions headache as having some 200 causes. Tension and psychogenic etiologies, which are featured in some accounts, are denied by many. This report describes the occipital neuralgia syndromes, and suggests new cause for pain in the occipital region: unilateral C1–2 arthrosis.

Occipital Neuralgia Syndromes

Etiology

The term “occipital neuralgia,” in fairly constant use since its first mention, has a double connotation. It describes on the one hand a category of patients with occipital pains of diverse origins, although with rather stereotyped clinical complaints, and on the other hand the syndrome presented by a specific patient or homogeneous group of patients who on examination fail to yield evidence of a demonstrable lesion capable of causing the complaint. As definite causes are determined for such disorders as posterior fossa neoplasm, Arnold-Chiari malformation, and posttraumatic residua, the nonspecific appellation of neuralgia can be discarded. Discovery of additional etiologies such as we describe will diminish the neuralgia category further.

All who use the term “occipital neuralgia” recognize that it is not a disease in the sense that the trigeminal and glossopharyngeal neuralgias are entities with not only stereotypical manifestations, but also probably a single (though not yet completely agreed upon) etiology. Speculation abounds concerning the cause of occipital neuralgia not attributable to organic sources. The effect of fibrositis and other hypothetical abnormalities in the...
muscles and fascia through which the nerves pass has many supporters.\textsuperscript{6,9,12} Entrapment of the nerves between the C-1 and C-2 vertebrae,\textsuperscript{9} sympathetic disorders with arteriolar dilatation and leakage of serotonin, histamine, and bradykinin into tissue,\textsuperscript{7,23} and lesions of cervical discs at lower levels in the spine\textsuperscript{20} have all been implicated. The belief that the common type of lower cervical arthrosis can cause occipital pain has become less accepted as realization has grown that patients with undoubted spondylosis and disc rupture causing radiculopathy and myelopathy at the usual sites rarely, if ever, have occipital neuralgia as well. The increasing discovery of specific abnormalities at the C1-2 level in patients with the occipital pain syndrome has opened the possibility of causes other than degenerative changes in the lower cervical spine.\textsuperscript{2,3,6,10,13,18,21,22}

**Anatomical Features**

It appears widely accepted nowadays that occipital pain usually results from an affection in or of the C-1 and C-2 nerve roots, nerves, or their tributaries. The C-2 cord segment, endowed with a number of posterior root filaments, accepts input from the C-1–2 lateral articulation, the periosteum of these vertebrae, and the ligaments, muscles, and scalp. The C-1 segment is not devoid of dorsal root fibers as was once supposed\textsuperscript{10} and, in addition, there is the possibility of pain transmission centrally via small unmyelinated fibers traveling in anterior roots. Pain input via the C-1 roots may be responsible for the frontal pain originating from posterior fossa abnormalities producing cerebellar tonsil herniation, and there is also the possibility that the vagus nerve supply to the posterior fossa dura and blood sinuses serves as a pain pathway. Lesions at the C-3 level may cause pain simulating that produced by lesions at C1–2, but it has been our observation that lesions of C-4 and the lower roots yield pains having a hemi-collar or brachial distribution.\textsuperscript{3} Lower-level lesions at C4–5, C5–6, C6–7, and below cause neck and arm syndromes, and thus the supposition that neck muscles inserting on the skull become tense as a consequence of lower cervical arthrosis, causing occipital pain, does not enjoy the acceptance it once had.\textsuperscript{20} Therefore, the commonly present lower cervical arthrosis is rarely a cause of occipital neuralgia, and one should search for higher-level pathology.

**Treatment Methods**

Treatment ranges from therapy directed at such proven causes as diabetes, gout, Arnold-Chiari malformation, and neoplasm, to a variety of temporarily denervational and destructive interventions, sometimes based more on hypothesis than on demonstrated pathology. Such treatments include rhizotomy,\textsuperscript{1} collarplacememt,\textsuperscript{2,7} massage, infrared heat, procaine and alcohol infiltrations,\textsuperscript{4,7,9,11} avulsion of greater occipital nerves,\textsuperscript{6,14} traction and contour pillows,\textsuperscript{7} steroid injections,\textsuperscript{11} excision and alcohol injection of all of the nerves in the back of the head,\textsuperscript{13} and decompression of the upper cervical roots.\textsuperscript{18} Occipital nerve avulsion seems to have been replaced recently by decompression of the nerve by stretching and dilatation of its path through the trapezius aponeurosis. Also, in recent years patients have undergone anterior interbody fusion and other types of fusion procedures at levels of arthrosis in the mid and lower cervical spine in an effort to relieve the occipital pain by elimination of motion at the pathological sites.

Recognition of little known yet valid and demonstrable causes of occipital neuralgia increases the opportunity for better focused and more effective treatment. We have discovered such a cause: the unilateral CI–2 arthrosis syndrome.

**Unilateral CI–2 Arthrosis Syndrome**

**Description of the Entity**

Unilateral arthrosis of a C1–2 lateral articulation was first seen by one of us while examining a series of roentgenograms, including several myelograms, that had been made of a 77-year-old woman who had become bedridden after several years of increasing left suboccipital pain, great retromastoid tenderness, and severe limitation of rotation of the neck. For relief of pain she had surrounded herself by an assortment of pillows, heating devices, and traction equipment. Although the open-mouth radiographic view was quite imperfect due to her inability to extend her neck enough to display the odontoid and lateral articulations well, it was apparent that the C1–2 articulation on the right was normal while that on the left was arthritic, with thinned and roughened cartilage surfaces and associated spurs. An anesthetic injection down to this joint gave immediate temporary relief which subsequently became permanent after C-2 dorsal root section on November 17, 1965. Relief endured until her death 10 years later at age 86 years.

Since then, we have seen and successfully treated other similar cases.\textsuperscript{3} The fact that these were not unimportant and isolated observations is suggested by a subsequent case described by Schneider.\textsuperscript{21} His patient was not followed after injection of a local anesthetic agent, and reliance on eventual self-limitation of the disease is unsupported by data. Worthy of attention is Schneider's admonition against occipital neurectomy, not so much because of the risk of later neuroma formation, but because cutting a peripheral nerve for a centrally located pain process seems likely to fail.

Unilateral painful arthrosis of C1–2 is not mentioned by those who have described cervical spondylosis and disc lesions as causes of occipital neuralgia, although Martin and Fagan\textsuperscript{19} advocated checking open-mouth x-ray films for the presence of spurs around the neutral foramina. Their report included a retouched x-ray film said to show a "C-2 spur." Curiously, these writers considered rhizotomy to be a formidable operation and favored radical excision of all the nerves lying in the
Occipital neuralgia due to C1–2 arthrosis

area, from one mastoid process to the other, a procedure of which we, along with Schneider,21 disapprove.

Summary of Cases

Based on our observations in seven patients, we are persuaded that the unilateral C1–2 arthrosis syndrome is a genuine clinical entity with distinctive pathophysiological features and therapeutic requirements. Four of these patients were women and three were men, ranging in age from 63 to 79 years. The pain experienced was almost identical among these patients, being localized to the retromastoid, suboccipital, and upper cervical regions of one side only and having a deep boring and aching nature. Pain was exacerbated by motion and occasionally extended toward the vertex, but not over the ear or to the frontal region or eye. Some residual aching was felt constantly between acute attacks. The duration of symptoms ranged from several months to 3 years. No precipitating neck trauma was remembered by any of these seven patients.

Examination disclosed a decreased range of neck motion in each patient, with production of pain by almost any motion of the C1–2 articulation. Direct pressure over this joint or the transverse process of C-2 increased pain, but there were no sensitivities distally along the course of the greater occipital nerve or at its fascial outlets. Testing with pinprick and light touch stimuli did not reveal any sensory deficits in the C-2 skin field. Roentgenograms of the cervical spine through the open mouth revealed unilateral arthrosis of the C1–2 joint on the affected side of each patient.

Degenerative changes in the lower cervical spine were not uncommon, but were not invariably present. Computerized tomography (CT) and radioisotope bone scanning, when performed, were also positive for unilateral disease at the C1–2 level.

Our two most recent cases are described in some detail, since they represent our current methodology. Illustrative Case Reports

Case 1. This 68-year-old woman presented with a 4-month history of intermittent pain in the right retromastoid and low occipital regions. The pain was of spontaneous and nontraumatic onset. She had a greatly limited range of neck motion with pain elicited by right rotation, lateral tilt, and foraminal compression. Digital pressure over the C-2 spinous process and the right lateral mass of C-2 caused the same pain. She exhibited no sensory deficits in the C-2 distribution, and all lower cervical roots functioned normally. Open-mouth radiographs demonstrated almost complete obliteration of the C1–2 joint space on the right side, similar to observations that had been made in previous patients (Fig. 1 left). A radionuclide bone scan demonstrated increased activity at this site only (Fig. 1 center). The CT scans revealed degenerative changes in this joint, with minimal involvement of the asymptomatic side (Fig. 1 right).

Treatment with a soft collar and anti-inflammatory drugs produced minimal relief. Injection of the posterolateral border of the C1–2 joint with 0.5% bupivacaine and a steroid under fluoroscopic control gave immediate relief, which persisted for 1 month. Following return of pain, repeat injection and other conservative mea-

Fig. 1. Case 1. Left: Open-mouth radiographic view of the upper cervical spine of a patient with the C1–2 arthrosis syndrome. This projection, of great value in the diagnosis of developmental abnormalities and fractures of the odontoid, also images the C1–2 lateral articulation, here shown painfully arthrotic on the right. Arthritic changes in these joints may go unreported or unappreciated in significance. Center: Posterior gamma camera view. Note the pathological uptake of isotope at the site of the C1–2 right lateral articulation. Right: Computerized tomography scan of the upper cervical spine showing arthritic enlargement, joint space narrowing, and cartilage destruction on the right (shown at left in this scan).
Fig. 2. Case 2. Upper Left: Lateral radiograph of the cervical spine revealing loss of cervical lordosis, arthrotic narrowing of the C5-6 and C6-7 interspaces, anterior spurs at C5-6, and posterior spurs at C5-6 and C6-7. Changes of this sort and at these levels of the spine have been credited with causing headache in the absence of any indication of cervical radiculopathy. Lower: Open-mouth radiographic view of the C1-2 lateral articulations showing arthrotic destruction of the joint space on the painful (left) side and a normal joint on the right. Upper Right: Anteroposterior myelogram demonstrating more normal-appearing root sleeves on the left than on the painless (right) side. This lends strong support to the belief that it is upper cervical arthrosis at C1-2 on the left side that produced the pain rather than degenerative changes of the ordinary sort at lower levels without clinical radiculopathy.

Surgical treatment was unsuccessful. The patient then underwent intradural rhizotomy of the C-2 root and fusion of the C1-2 motion segment. This gave immediate relief, which has continued to the present, 2 years later.

Case 2. This 67-year-old man presented with left upper neck, retromastoid, and low occipital pain aggravated by neck motion. The pain was variable in intensity, but never absent since becoming established several months before. It had not proven amenable to aspirin, ibuprofen, or heat. Neurological examination, including a search for occipital and upper cervical sensory loss, was normal. Neck motion was limited, and there was sharply localized tenderness at the C1-2 level, just behind the mastoid tip on the left. Roentgenograms disclosed multilevel arthrosis in the lower cervical spine (Fig. 2 upper left) and unilateral arthritic changes in the left C1-2 articulation (Fig. 2 lower). Due to a suggestion that the lower changes might be more significant than those at C1-2, myelography was performed showing extradural lower cervical root compression on the right, asymptomatic, side rather than on the left (Fig. 2 upper right).

The patient underwent injection of 5 ml of 0.5% lidocaine down to the posterolateral aspect of the C1-2 joint. This immediately relieved the pain but also produced some difficulty in swallowing for 1 hour. Pain relief lasted for 1 1/2 weeks, after which the dorsal root of C-2 was cut on March 23, 1982. At follow-up examination on September 24, pain was absent, the neck was more mobile than it had been for years, and only a tiny area of hypalgesia to pinprick could be found in the left occipital scalp.

Comment. Of the remaining five patients, three were seen early after we recognized this condition and have been lost to follow-up review. One patient, treated conservatively because of an associated medical condition, obtained only partial relief. The three patients who underwent surgery achieved relief lasting from 2 to 10 years.

Discussion

The C1-2 segment is unique in its great rotatory mobility, having an average range of motion to each side of 47°. In contrast with this, the occipital C-1 condyle has essentially no rotatory motion. From 10° to 13° of flexion-extension motion is present at both of these highest-motion segments. The C-2 nerve and ganglion complex lie on the posterior surface of the C1-2 lateral articulation, passing through the lateral capsule of this joint, but we believe that the pain suffered by our patients arose not from compression of the nerve, but as a primary irritative phenomenon in the joint itself, mediated and transmitted by the anterior ramus of C-2. Spontaneous fusion of such a severely arthrotic articulation may result in disappearance of pain, as should surgical fusion of the joint. We have seen patients who had arthrosis of C1-2 revealed during the course of radiography for other conditions with no symptoms attributable to it, possibly due to the fact that fusion had already occurred. Our patients who required treatment had some pain aggravated by motion in arthrotic joints that presumably still possessed mobility.

Summary

We have described a group of elderly patients suffering from an occipital neuralgic pain syndrome aggravated by neck motion due to degenerative changes in a C1-2 lateral articulation. X-rays films, CT scans, and
Occipital neuralgia due to C1–2 arthrosis

Radionuclide studies confirmed the severe arthrotic involvement. Associated cervical spondylosis at lower levels appeared asymptomatic in our patients, several of whom had been studied myelographically. No neurological deficits were encountered in any of these patients. Local anesthetic blocks produced temporary relief, and the addition of steroids at the time of injection seemed to be reinforcing. Return of intractable pain after failure of conservative treatment justified intradural C-2 rhizotomy for denervation of the painful joint or C1–2 fusion. Such treatment gave permanent relief to the three patients so treated.

References


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