Surgical Neurology International

OPEN ACCESS

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Editor: **James I. Ausman,** MD, PhD University of California, Los Angeles, CA, USA

Case Report

Occipital neuralgia secondary to unilateral atlantoaxial osteoarthritis: Case report and review of the literature

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Received: 28 June 15 Accepted: 10 October 15 Published: 23 December 15

Abstract

Background: Atlantoaxial osteoarthritis (AAOA), either in isolation or in the context of generalized peripheral or spinal arthritis, presents most commonly with neck pain and limitation of cervical rotational range of motion. Occipital neuralgia (ON) is only rarely attributed to AAOA, as fewer than 30 cases are described in the literature.

Case Description: A 64-year-old female presented with progressive incapacitating cervicalgia and occipital headaches, refractory to medications, and local anesthetic blocks. Computed tomography and magnetic resonance imaging studies documented advanced unilateral atlantoaxial arthrosis with osteophytic compression that dorsally displaced the associated C2 nerve root. Surgical decompression and atlantoaxial fusion achieved rapid and complete relief of neuralgia. Ultimately, postoperative spinal imaging revealed osseous union.

Conclusions: Atlantoaxial arthrosis must be considered in the differential diagnosis of ON. Surgical treatment is effective for managing refractory cases. Intraoperative neuronavigation is also a useful adjunct to guide instrumentation and the intraoperative extent of bony decompression.

Key Words: Atlantoaxial fusion, occipital neuralgia, osteoarthritis

Access this article online

Website: www.surgicalneurologyint.com

DOI: 10.4103/2152-7806.172531

Quick Response Code:

INTRODUCTION

Atlantoaxial osteoarthritis (AAOA) typically presents with neck pain and restricted cervical rotation. Occipital neuralgia (ON), characterized by lancinating pain and dysesthesia in the distribution of the greater and lesser occipital nerves, is most commonly idiopathic but may occur when the C2 or C3 nerve roots are statically or dynamically compressed. Potential surgical etiologies include instability following prior trauma or surgery, compression from neoplasms, gross atlantoaxial instability secondary to rheumatoid arthritis, nerve entrapment by a hypertrophic atlanto-epistrophic ligament, or mechanical compression from a severely congested C2 venous plexus, or ectatic vertebral artery.[3,15,17,19] True ON is a rare presentation of C1–C2 facet osteoarthritis with fewer than 30 cases in the literature to date.

Here, we present a patient who has ON and severe upper cervical pain was attributed to unilateral AAOA. As she was refractory to medical therapy, she underwent osteophyte decompression with posterior atlantoaxial

How to cite this article: Guha D, Mohanty C, Tator CH, Shamji MF. Occipital neuralgia secondary to unilateral atlantoaxial osteoarthritis: Case report and review of the literature. Surg Neurol Int 2015;6:186.
http://surgicalneurologyint.com/Occipital-neuralgia-secondary-to-unilateral-atlantoaxial-osteoarthritis:-Case-report-and-review-of-the-literature/

fusion utilizing a frameless stereotactic navigation system.

CASE HISTORY

A 64-year-old female presented with several years of progressive intractable cervicalgia and occipital headache; there was no trauma history. She had restricted cervical rotation/left head tilt and neck spasms, accompanied by pain in the left suboccipital region, radiating to the left occipital scalp, and left ear (paraorbital region). There was no history of peripheral or spinal osteoarthritis or any other rheumatologic conditions.

Notably, two attempts at left occipital nerve blocks were made in the year prior to surgery; both resulted in clear albeit modest immediate improvement, but without lasting benefit.

On examination, the patient was neurologically intact but demonstrated exquisite tenderness to light touch over the left occipital scalp.

The computed tomography (CT) of the cervical spine revealed advanced unilateral arthrosis on the left involving the C1–C2 facet joint, accompanied by significant posterior osteophytic protrusion into the left C1–C2 foramen [Figure 1]. The magnetic resonance imaging (MRI) of the cervical spine confirmed the displaced and compressed C2 nerve root on the left side [Figure 2a], but the right side was normal [Figure 2b].

Surgical procedure

A posterior upper cervical approach was implemented, and intraoperative CT three-dimensional stereotactic navigation was utilized. At surgery, the C2 nerve root was retracted while the osteophytic complex was removed. Autograft was placed into both C1–C2 facet joints and with navigation, bilateral C1 lateral mass, and C2 pedicle screws/rods were placed without complication.[9]

Postoperative course

Postoperatively, the patient remained neurologically intact with a complete and immediate resolution of her cervicalgia and lancinating occipital headache. She was placed in an Aspen cervical orthosis for 12 weeks after which it was weaned, and active neck physiotherapy was implemented. At 3-month postoperative follow-up, she remained free of her ON and neck pain with radiographic confirmation of osseous union across the C1–C2 facet joints and excellent foraminal decompression [Figure 3].

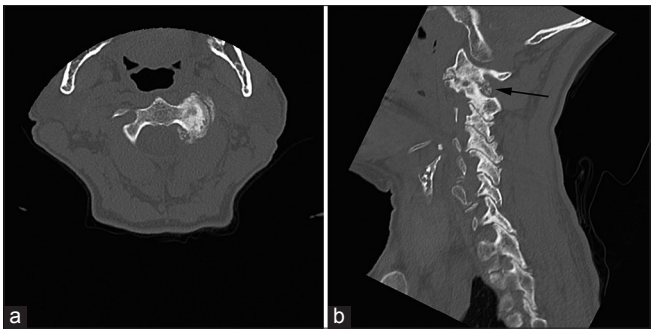

Figure 1: Axial (a) and left paramedian sagittal (b) preoperative computed tomography of the cervical spine. Unilateral arthrosis of the left C1–C2 facet joint is shown with obliteration of the joint space and extensive osteophyte formation. The posterior osteophyte encroaches into the expected site of the left C2 dorsal ramus (black arrow)

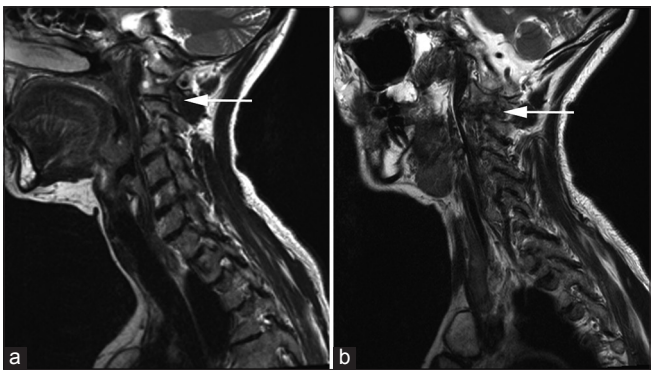

Figure 2: Right (a) and left (b) paramedian sagittal T2-weighted magnetic resonance imaging of the cervical spine. The exiting C2 nerve root is severely compressed on the left, in contrast to the right (white arrows)

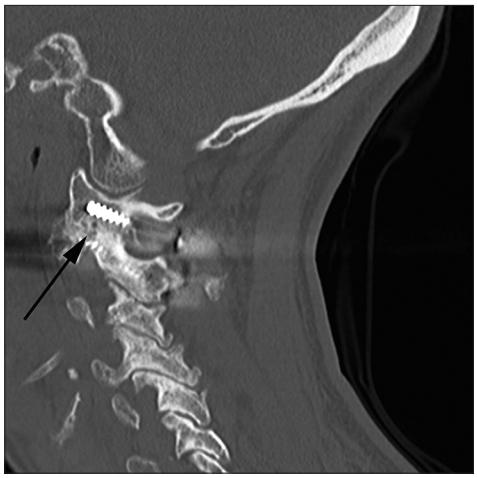

Figure 3: Left paramedian sagittal computed tomography of the cervical spine, 3-month postoperatively. The osteophyte complex at C1–C2 on the left has been eliminated with osseous union of the facet joint (black arrow)

DISCUSSION

ON was first described formally by Hunter and Mayfield in 1949 and is characterized by paroxysmal lancinating pain in the distribution of the greater or lesser occipital nerves, variably accompanied by a dull ache between episodes.[10] Diagnostic criteria include a pain history consistent with typical ON, along with dysesthesias in the affected dermatome during or following episodes,

reproducible scalp tenderness or Tinel's sign over the affected nerves, and reliable relief with local anesthetic blocks.[14]

AAOA refers to degenerative arthrosis of the lateral (facet) or central atlantoaxial joints, afflicting the facet joint in 12%, atlanto-odontoid articulation in 17%, and both in 71%.[8] Asymptomatic radiographic lateral AAOA has a reported prevalence of 5.4% in the sixth decade, increasing to 18.2% by the ninth decade.[20] Such C1–C2 facet osteoarthritis is symptomatic in 4% of patients with known peripheral or spinal osteoarthritis, occurring in females in 74%, and presenting unilaterally in 76% in one large series.[7] The symptomatic disease typically presents with restricted cervical rotation and neck and suboccipital pain that may be exacerbated by rotation but is distinct from the lancinating pain of ON.[6] ON secondary to lateral AAOA is rarely reported, limited to case reports or small series in the literature, summarized in Table 1.

The first formal description of ON secondary to lateral AAOA was by Ehni and Benner in a series of seven patients described in Table 1.[3] Several others have subsequently described small case series of ON from unilateral AAOA, with posterior C1–C2 instrumented fusion affording permanent relief in all cases, and one pseudoarthrosis as the only reported surgical complication [Table 1].[5,11,12,18] More recently, Pakzaban reported on 2 cases with clear osteophytic compression of the C2 dorsal ramus in the C1–C2 interlaminar space.[15] Both patients had immediate and complete resolution of their pain following posterior C1–C2 fusion without decompression. In a large study correlating clinical radiculopathies with MRI findings, Kucinski *et al*. reported on 1 case of ON secondary to AAOA, treated unsuccessfully with oral analgesia alone.[13]

Our review of the literature revealed 29 cases of true ON secondary to lateral AAOA, 17 treated surgically. Operative treatment included either intradural C2 dorsal rhizotomy or C1–C2 instrumented fusion, without decompression even in cases with clear osteophytic nerve root compression.[15,18] While atlantoaxial fusion is an effective option for cervicalgia secondary to AAOA, its utility for ON is less clear.[4,16] The pathogenesis of pain in ON has been postulated to include direct compression of the occipital nerves, as well as micromotion of the degenerated C1–C2 facet joint itself, with pain transmission via the anterior ramus of C2.[2,3] Elimination of facet micromotion through fusion can, therefore, be expected to remove one of the primary pain generators in this syndrome.

Our case uniquely underwent decompression of a posterior atlantoaxial osteophyte in addition to fusion. This has been replicated in other series of ON

Table 1: Summary of the existing literature

Author	Year	Number of patients	Diagnosis	Treatment	Outcome	Surgical complications
Ehni *et al.*	1984	7	Open-mouth XR ± CT ± bone scan	Facet injection ($n=4$)	Temporary relief	-
				Intradural C2 dorsal rhizotomy ($n=2$)	Permanent relief	None reported
				Intradural C2 dorsal rhizotomy + posterior C1–C2 instrumented fusion ($n=1$)	Permanent relief	None reported
Star *et al.*	1992	9	Multiplanar XR ($n=9$) Bone scan ($n=7$) MRI ($n=4$) CT ($n=3$)	NSAIDs ($n=4$)	Permanent relief (2 required ongoing daily NSAIDs)	-
				Posterior C1–C2 instrumented fusion ($n=5$) Brooks fusion ($n=2$) Gallie fusion ($n=3$)	Permanent relief	1 pseudoarthrosis requiring reoperation, 1 protracted ileus, 1 UTI
Joseph *et al.*	1994	4	Open-mouth XR	Gallie fusion ($n=4$)	Permanent relief	None reported
Fuentes *et al.*	2001	3	XR + CT + MRI	CT-guided facet injection ($n=2$)	Permanent relief	-
				Posterior C1–C2 instrumented fusion ($n=1$)	Permanent relief	None reported
Tancredi *et al.*	2004	1	XR + CT + MRI	Facet + GON injections	Permanent relief	-
Kafer *et al.*	2004	1	XR + CT + MRI + bone scan	Posterior C1–C2 instrumented fusion, with transarticular screws + atlas clamps	Permanent relief	None reported
Pakzaban	2011	2	XR + CT + MRI + bone scan	Posterior C1–C2 instrumented fusion, with transarticular screws + modified Sonntag cables	Permanent relief	None reported
Kucinski *et al.*	2015	1	MRI	Oral analgesia	No relief	-

CT: Computed tomography, GON: Greater occipital nerve, MRI: Magnetic resonance imaging, NSAIDs: Nonsteroidal anti-inflammatory drugs, UTI: Urinary tract infection, XR: X-ray

secondary to posttraumatic atlantoaxial arthrosis and hyperostosis.[1] Fusion alone, however, has also been shown effective in cases with osteophytic protrusion into the C1–C2 interlaminar space, similar to our case, where decompression was deemed unsafe intraoperatively.[15] Our case, therefore, highlights, for the 1st time in the literature, the utility of intraoperative neuronavigation as an adjunct for assessing the adequacy of osteophyte decompression, improving safety by eliminating excess drilling beyond the minimum required.

CONCLUSIONS

ON is a rare sequela of lateral AAOA. Diagnosis is suspected clinically and confirmed with radiographic evidence of facet arthrosis. Workup also includes ruling out other structural lesions with CT and MRI, and investigating instability with dynamic radiographs. Atlantoaxial fusion is an effective option for refractory cases, and intraoperative frameless stereotaxy is a useful adjunct to guide hardware placement as well as to define the extent of osteophyte decompression. Atlantoaxial focal foraminal decompression and instrumented fusion is an effective option.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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