



Case Report

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Atlantoaxial Osteoarthritis: A Well-Established Entity that Remains Frequently Overlooked

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ABSTRACT

Painful atlantoaxial (C1-2) osteoarthritis (AAOA) has been described over 40 years ago. The condition may cause severe pain symptoms and disability related to the unilateral suboccipital pain and, in some cases, occipital neuralgia. One of the greatest challenges with AAOA is making the diagnosis. Diagnosis is commonly missed or delayed when headaches are treated in isolation or when pain is attributed to subaxial spondylosis.

Here we present an illustrative case involving a 67-year-old male presenting with classic painful AAOA. After failing conservative treatments, he was evaluated with morphologic, radiological studies and a diagnostic injection. He was successfully treated with bilateral, navigation guided C1 lateral mass and C2 pedicle screw fixation and fusion.

To conclude, when there is clinical suspicion for painful AAOA, providers have numerous diagnostic modalities, including newer hybrid techniques, that can be used to solidify the diagnosis. When conservative efforts fail, C1-2 fusion is an effective and enduring treatment for most patients.

KEYWORDS: Atlantoaxial, Osteoarthritis, Treatment

INTRODUCTION

Sharp, shooting, or nagging pain in the suboccipital region may be the result of painful atlantoaxial osteoarthritis (AAOA) (16). In patients with AAOA, physical examination typically reveals marked restriction in axial rotation towards the affected side and localized unilateral tenderness in the C1-2 region (28). Neurological exam is typically normal (1). The association of AAOA with occipital neuralgia is well-established (8,12,28,30). Fortunately, most patients with this distinct clinical syndrome can be treated effectively with conservative modalities such as physical therapy (PT), massage, daily stretching exercises, transcutaneous electrical nerve stimulation, nonsteroidal anti-inflammatory drugs (NSAIDs), centrally acting pain medications such as gabapentin, and injection therapy. With C1-2 instability and/or severe, medically refractory pain from C1-2 facet arthrosis, surgical intervention may be necessary. Atlantoaxial fusion is

typically the treatment of choice (12). Numerous case reports and series have demonstrated excellent clinical outcomes after C1-2 fusion for severe arthritis in the atlantoaxial joint (10,13,15,18,27,29). In this manuscript, we present an illustrative case highlighting the workup and management. We also discuss proposed pathophysiology for the entity, diagnostic modalities, and surgical treatment.

CASE REPORT

The patient was a 67 year-old-male who presented with pain in the right suboccipital region and a grinding sensation at the skull base. The pain started abruptly nine months prior to presentation to our institution after he inadvertently grounded a golf club during a hard swing. The pain was localized to the right suboccipital area with occasional lancinating pain to the ipsilateral occipital region. He had limited range of motion and increased pain with right axial rotation of the neck. Pain

was also exacerbated by neck flexion. The patient had been treated with extensive conservative modalities including PT, cervical traction, NSAIDs, trigger point injections, activity modification, and massage therapy. The open mouth odontoid view x-rays and the coronal CT images showed right-sided C1-2 joint space obliteration and subchondral sclerosis (Figures 1 and 2). His magnetic resonance imaging (MRI) revealed increased peri-facet short T1 inversion recovery (STIR) signal (Figure 3). A CT-guided injection was performed to confirm the C1-2 pain generator. Figure 4 demonstrates an illustrative left-sided injection in another patient. The patient experienced 80% transient relief in his pain symptoms following the injection. The patient elected to proceed with surgical intervention. We performed a navigation-aided C1-C2 instrumented fusion utilizing Harms construct (Figure 5). There was no C2 neurectomy or direct decompression of the C2 nerve performed. The patient tolerated the procedure well. Three weeks after the operation, the patient reported he was pain-free. His pain relief persisted at the one year follow up and, and his CT scan demonstrated solid C1-2 arthrodesis. Figure 6 demonstrates fusion across the degenerated C1-2 joint.

DISCUSSION

Painful AAOA is a well-established and distinct clinical syndrome (14). The suboccipital pain may be severe and incapacitating for patients (1,2). Patients may also present

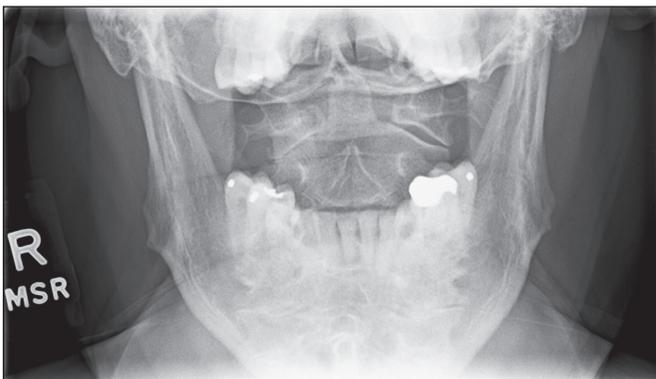


Figure 1: Open mouth odontoid view radiographs are a fast, simple test to detect narrowing of the C1-2 articulations. In this case, the positioning is suboptimal, but the asymmetric right sided narrowing is readily apparent (yellow arrow).

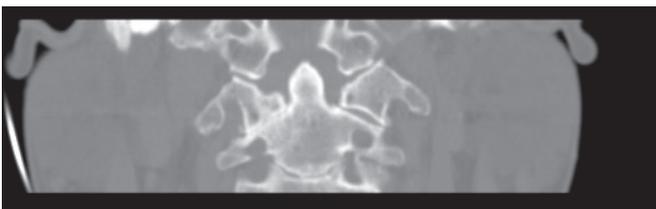


Figure 2: CT is far superior to radiographs and MRI in the evaluation of osseous anatomy and pathology. Here, a coronal CT image demonstrates asymmetric osteoarthritis of the right C1-2 articulation with loss of joint height, cortical irregularity, and subchondral sclerosis.

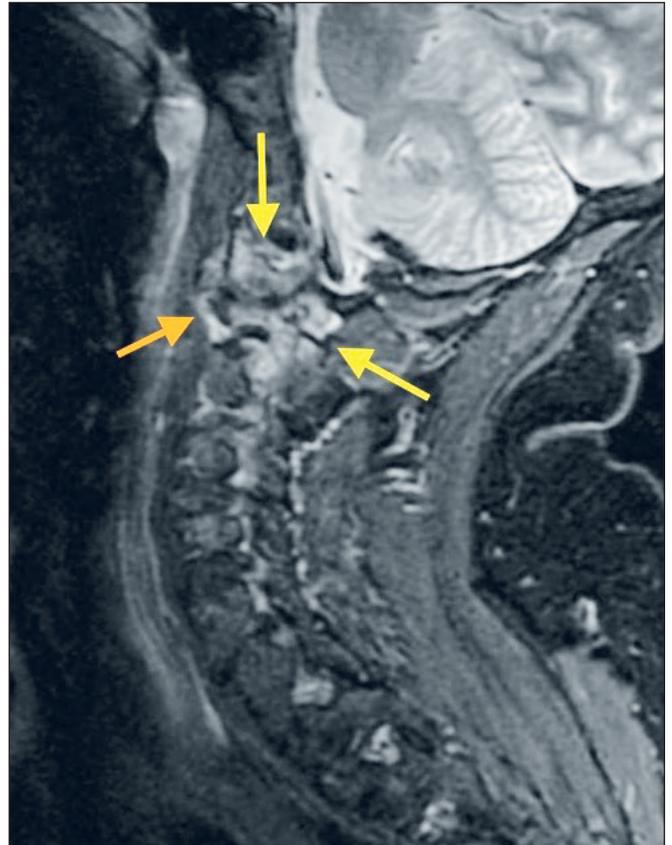


Figure 3: Sagittal STIR image shows hyperintensity in the right C1-2 joint and bone marrow (yellow arrows) reflecting reactive marrow edema. Hyperintensity in the local soft tissues (orange arrow) highlights the utility of MRI in characterizing active inflammation in all tissue types.

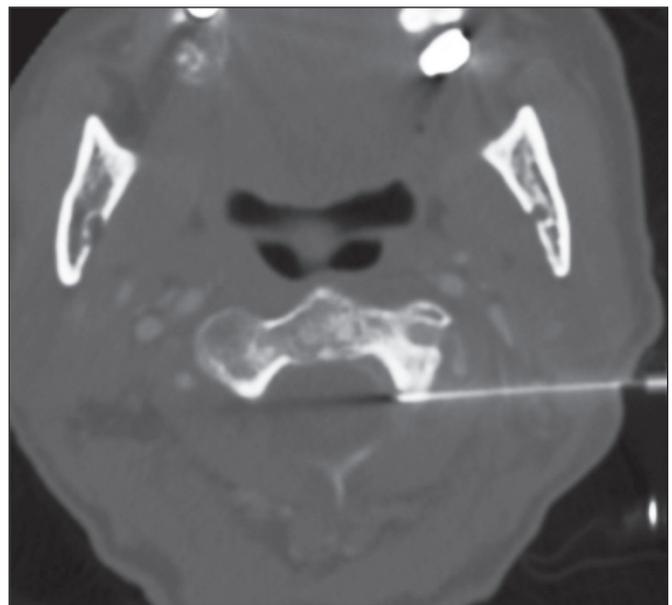


Figure 4: Intraprocedural CT image shows needle positioning for a left C2 nerve block in a different patient. In some cases, a rapid CT angiogram may be performed immediately prior to needle placement to confirm the location of the vertebral artery.



Figure 5: Lateral radiograph (A) shows fixation with C1 lateral mass and C2 pedicle screws.



Figure 6: The coronal CT image shows successful fusion across the degenerated joint.

with occipital neuralgia, an equally disabling neuropathic pain syndrome.

Pathophysiology

Cervical spondylosis is common, and the prevalence increases with age. This finding has been attributed to the increased mobility and large neutral zone, defined as the region where the joint moves easily with minimal applied force, of the C1-2 joints (3,5).

In a large retrospective study of 1,543 adult trauma patients who underwent a cervical spine CT scan, 39% of elderly patients were noted to have intraosseous cysts, calcific synovitis, or other arthritic findings affecting the C1-2 joints (4). Painful AAOA is known to be associated with occipital neuralgia (8,12,30). The superior articulating facet may compress the ganglion or irritate the greater occipital nerve which arises from the medial branch of the dorsal ramus of C2 nerve within the C1-2 interlaminar space (16).

Another theory is a C2 entrapment neuropathy between the hypertrophied C2 joint and surrounding soft tissues (20-22). Repetitive mild compressions or a limited number of more severe compressions of the entrapped C2 nerve may cause injury and segmental demyelination of the sensory axons. With demyelination, even micromovements in the joint contacting the injured nerve may lead to lancinating, paroxysmal, occipital neuralgia pain (16). A third theory involves venous congestion surrounding the hypertrophied, inflamed facet joint. The venous congestion may lead to edema, ischemia, cellular disruption, and demyelination (16).

Why Painful AAOA is Misdiagnosed

Although painful AAOA has distinct clinical features, the diagnosis is often missed or delayed. Patients may present initially with more mild symptoms which are attributed to myofascial pain or a soft tissue etiology. Another complicating factor is that the atlantoaxial articulations are often obscured on conventional radiographs by overlying maxilla and skull base. Elderly patients commonly have diffuse spondylosis, which distracts treating providers from the arthritic C1-2 joint. The prevalence of misdiagnosis is highlighted in one surgical series involving 42 patients treated with C1-2 fusions for AAOA. Twelve percent of patients in the series had undergone prior subaxial cervical fusion for a misdiagnosed pain generator (1).

The Radiological Assessment of AAOA

The characteristic appearance on radiographs and CT is asymmetric C1-2 facet osteoarthritis with loss of cartilage and subchondral sclerosis. On MRI, marrow edema is seen at the site of active inflammation, best visualized on STIR (short tau inversion recovery) or fat-saturated T2- weighted images, as these sequences remove marrow fat signal and highlight only water signal, which appears hyperintense. Fat saturated post contrast T1-weighted images also highlight the site of inflammation in the bone, joint, and surrounding soft tissues (6,7). In addition to morphologic evaluation with MRI, CT and radiographs, physiologic techniques such as bone scan (25,31), SPECT (single-photon emission computed tomography)/CT, and FDG-PET (positron emission tomogra-

phy) can be used to identify sites of active bone turnover and osteoblast activity.

Recently there has been great interest in the use of SPECT-CT for identifying pain generators in the lumbar and cervical spine. In this test, there is increased radiotracer uptake at the site of high metabolic activity (bone turnover), which can increase the sensitivity of CT for anatomic localization. Anatomy is better visualized with SPECT/CT than with bone scintigraphy. No studies show superiority of physiologic imaging techniques over morphologic techniques, and relatively few studies have assessed the use of nuclear medicine tests for diagnosis of cervical facet arthropathy. A retrospective study of 7 patients with neck pain assessed with SPECT CT, postoperative neck disability index, and visual analog scale scores showed that SPECT-CT can be a useful adjunct in the workup of neck pain secondary to facet arthropathy. In some cases, SPECT-CT may obviate diagnostic injections (26). A larger study of patients with axial back or neck pain, including facets at all spinal levels, showed that 48% of patients had hypermetabolism of facet joints on SPECT-CT (25).

When clinical features and imaging are inadequate for isolating the pain generator, diagnostic injections are useful for definitive diagnosis. C2 dorsal root ganglion block or C1-2 facet injection with anesthetic and steroid under CT guidance is a useful tool for both diagnosis and pain management. There are several case reports of successful treatment of cervicogenic headache with RF ablation (20,21), however systematic reviews and meta-analyses have been inconclusive due to lack of high quality randomized control trials (11,12). In one retrospective study with 32 patients, C1-2 intra-articular injections, 81% of patients experienced 50% or greater pain relief following the injections. Unfortunately, at 6 months the group's mean pain score was not statistically different from the mean preoperative pain score, implying the analgesia benefit is short-term (23). Other studies have also shown only short-term benefit with C1-2 injections for painful AAOA (19). In rare cases, one or more injections can alleviate pain sufficiently until the joint autofuses or stabilizes, and no further treatments are required.

The C2 nerve block procedure is most safely performed under CT guidance with use of a small amount of myelographic contrast to ensure that the needle does not contact the vertebral artery or enter the thecal sac. Ablation is followed by an additional nerve block to decrease postprocedural pain and inflammation. Complications from this procedure are uncommon, typically numbness and dysesthesia. A large study involving 136 fluoroscopic-guided, C1-2 injections in 72 patients reported minor adverse events up to 18.5% of patients. The most common procedural adverse events were vascular uptake of contrast (3.7%) and paresthesia (2.2%). The most common post-procedural adverse event was increased joint pain (5.9%). All intra- and post-procedural adverse events resolved within 3 months and no permanent sequelae were identified (2). No similar studies have been performed for CT guidance, with its superior anatomic resolution. When performed under CT-fluoroscopy, the procedure can

be performed even more rapidly and with limited radiation exposure to the patient. C1-2 injections are part of the multimodal treatment algorithm for C1-2 joint pain and can provide useful diagnostic information in selected AAOA cases.

Surgical Treatment

With extensive degenerative facet arthropathy or instability, C1-2 fusion may be necessary. Clinical series have demonstrated overwhelmingly positive clinical outcomes (10,13,15,18,27,29). A systematic review reported greater than 92% successful arthrodesis following C1-2 fusion for this indication (9). In one series with 35 patients and a mean follow up of 6.5 years, 89% of patients were pain free or had markedly improved pain symptoms after C1-2 fusion for symptomatic AAOA (12). In another large series involving 42 patients, the average preoperative Neck Disability Index (NDI) score was 26.88 ± 24.85 , which improved to 10.59 ± 14.88 at the 1-year follow-up ($p=0.004$). In the same study, severe disability based on NDI score decreased from 18% to 2% at 1 year and 0% at 2 years ($p=0.01$) (1).

Various surgical techniques for C1-2 fusion in this patient population have been described including Halifax clamps with wiring, Gillies's fusion, C1-2 transarticular facet screws and fixation using C1 lateral mass and C2 pedicle / pars screws (Harm's construct) (12,15,17,24). A recent meta-analysis has not shown superiority of any technique (9). The predominate technique utilized in more recent series is the Harm's construct, as was described earlier in the illustrative case (1). Successful treatment has been described without decompression of the C2 nerve or C2 neurectomy (24). Although surgeons may elect for C2 neurectomy to aid in hemostasis, expose the joint for arthrodesis, and avoid postoperative irritation of the C2 nerve by the C1 lateral mass screw (16), excellent clinical outcome can be achieved without sacrificing the C2 nerve.

CONCLUSION

Painful AAOA is a well-established clinical syndrome that can result in severe symptoms and disability. Although AAOA is not novel, diagnosis is often missed or delayed. Clinical suspicion based on presentation is key. Various diagnostic modalities, including hybrid imaging, can be used to confirm the diagnosis. When necessary, C1-2 fusion is a highly effective, definitive treatment for symptomatic AAOA.

AUTHORSHIP CONTRIBUTION

Study conception and design: MTN, WG, MKL

Data collection: MTN

Analysis and interpretation of results: MTN, WG, MKL

Draft manuscript preparation: MTN

Critical revision of the article: MTN, WG, MKL

Other (study supervision, fundings, materials, etc...): MTN, WG, MKL

All authors (MTN, WG, MKL) reviewed the results and approved the final version of the manuscript.

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