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CASE REPORT

C1–T2 decompression and fusion for C2 erosive pannus—a case report

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INTRODUCTION

The occipitocervical complex consists of the bony and ligamentous structures from the occiput to the C2–C3 disk space. This region provides a major contribution to the motion of the cervical spine and hence its stability is crucial [1]. Cervical instability can result from different processes; including trauma, tumors, degenerative changes, hypermobility connective tissue disorders, and inflammatory changes to the cervical spine. Inflammatory changes can result in pannus formation and erosion of joint surfaces as well as laxity of capsular structures. In the rheumatoid cervical spine, the loss of joint congruity and ligamentous incompetence from chronic inflammatory changes leads to atlantoaxial instability. This instability has often been associated with peri-odontoid pannus [2].

Peri-odontoid pannus of the cervical spine is a well-recognized complication of rheumatoid arthritis. However, a recent study demonstrated that only 28% of patients with atlanto-axial pannus found on cervical spine MRI had an associated diagnosis of rheumatoid arthritis, while the remaining patients had either CPPD (44.7%), a non-rheumatoid inflammatory rheumatic disease (7.8%), or no underlying systemic disease (19.5%) [3]. The same study also noted that patients with incidental atlanto-axial pannus on cervical spine MRI were unlikely to have previously unrecognized rheumatoid arthritis [3]. In spite of this, there is a paucity of literature discussing the operative management of a patient with peri-odontoid pannus of non-rheumatologic etiology. In the only published case report on this topic, Lagares et al. performed an occipitocervical fusion in one patient with a peri-odontoid pannus of non-rheumatologic etiology, following which the patient's pannus resolved and myelopathy significantly improved [4]. In this case report, we describe a patient with progressive cervical myelopathy in the setting of a compressive peri-odontoid pannus of non-rheumatoid etiology who was successfully treated with a C1–T2 posterior cervical decompression and fusion, thereby sparing the atlanto-occipital joint.

CASE PRESENTATION

An 80-year-old female was referred to our clinic with a several-month history of progressively worsening neck and upper extremity numbness and tingling, weakness in her bilateral upper and lower extremities, transient bilateral upper and lower extremity paralysis, problems with fine motor skills, and balance issues. She reported that the transient episodes of bilateral lower extremity paralysis occurred when she extended her neck to look up, and as a result, she tried to avoid doing this as the last episode caused her to fall. She reported no issues with bowel or bladder incontinence. On manual motor strength testing, she had 4/5 strength in her bilateral deltoids and biceps, 4−/5 strength in bilateral triceps, 4+/5 strength in bilateral wrist extensors, and 4/5 strength in bilateral finger flexors and hand intrinsics with some intrinsic muscle wasting. In her lower extremities, she demonstrated 4/5 strength in the bilateral iliopsoas, 4+/5 strength in bilateral quadriceps, 4/5 strength in bilateral tibialis anterior, and 4+/5 strength in bilateral EHL. When describing the physical examination, we have referred to the ASIA/ISCoS International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI). She was noted to have a mildly positive Romberg and a positive Hoffmann's on the right. She denied any constitutional symptoms of illness, recent weight loss, excessive fatigue, night sweats, or other symptoms that would raise concern for systemic illness or malignancy.

A review of the patient's radiographs, which included multiple views of the cervical spine, demonstrated marked spondylotic changes throughout the cervical spine (Fig. 1). She had evidence of anterolisthesis at C3–4 and C4–5. She was able to extend into a more lordotic posture, which measured roughly 13 degrees of lordosis. A computed tomography (CT) scan of the cervical spine demonstrated a lytic lesion in the odontoid with the erosion of the posterior cortex, most consistent with pannus formation, erosive changes of the superior articulations of the C1 lateral masses, multilevel spondylotic changes, and spondylolisthesis at C3–4 and C4–5 (Fig. 2). A review of the patient's cervical spine MRI further characterized the large cystic-appearing mass at the odontoid, which was identified to be consistent with pannus formation causing stenosis at the C1–2 level in addition to multilevel central and neuroforaminal stenosis from C2 to C7. The patient's images were also notable for cord compression and myelomalacia (Fig. 3). Images of the cervical spine were also reviewed with our orthopedic oncologists and neuroradiologists in a multidisciplinary conference due to the large lesion in the odontoid process along with the extensive soft tissue mass. After a full review by this multi-disciplinary team, the lesion was determined to be a benign cyst due to degeneration or an underlying inflammatory process, and as such a formal biopsy was not recommended.

Our rheumatology colleagues were consulted for their input given that the radiographic findings may be a sign of undiagnosed rheumatoid arthritis. Serial laboratory panels were

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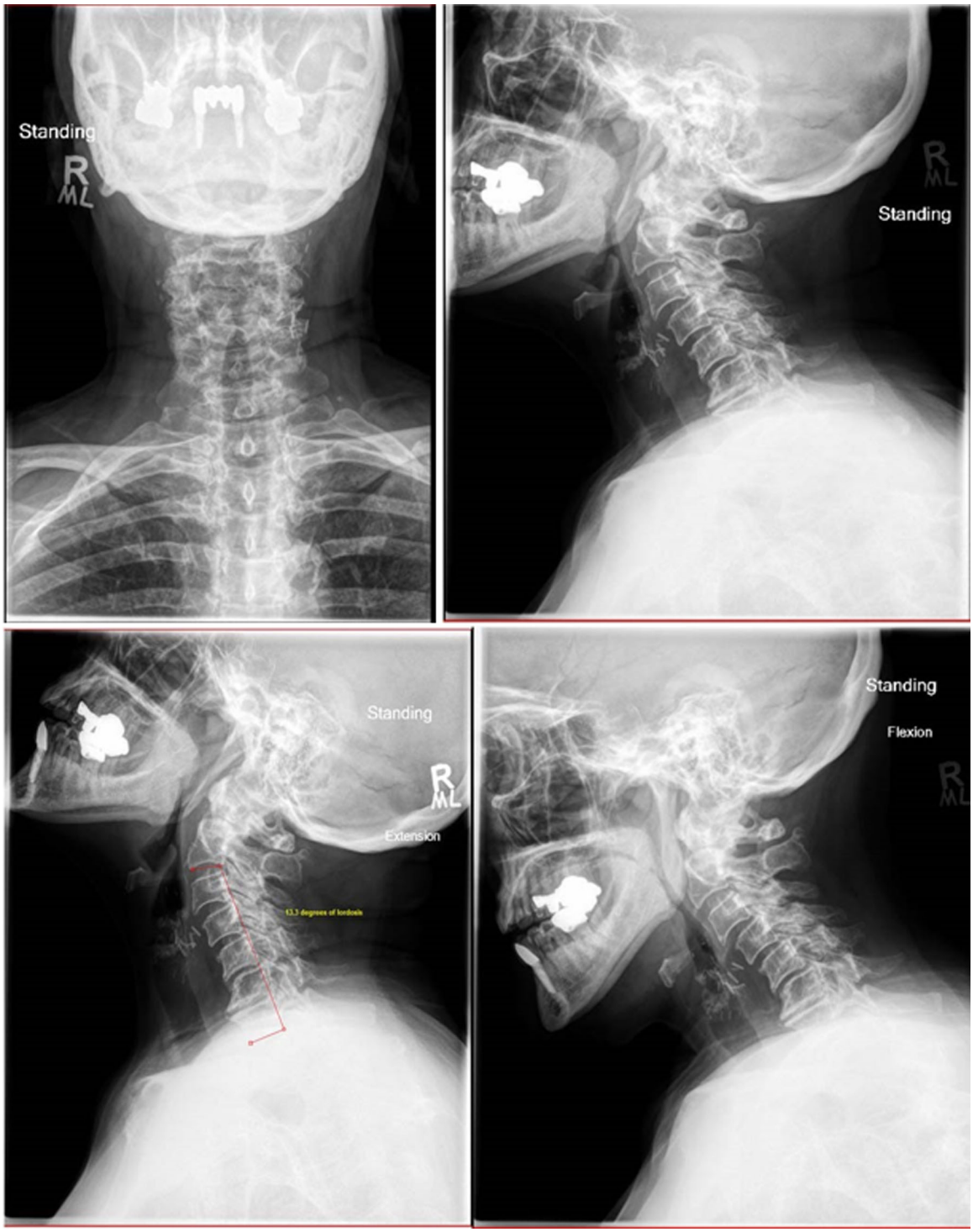

Fig. 1 Preoperative radiographs. Preoperative standing cervical spine radiographs, including AP (top left), lateral (top right), extension (bottom left), and flexion (bottom right) views.

drawn including rheumatoid factor (RF) and cyclic citrullinated peptide antibodies (CCP), both of which were within normal limits. We obtained X-rays of the hands, wrists, feet, and ankles as part of the evaluation for possible rheumatoid arthritis, but these did not demonstrate any other joints with significant erosive changes suggestive of rheumatoid arthritis. She was felt to not meet the criteria for the diagnosis of rheumatoid arthritis and therefore was not placed on any steroids or disease-modifying antirheumatic drugs (DMARDs). Additional consideration was given to the possibility that the patient's imaging findings represented an infectious process. However, the patient's lack of constitutional symptoms of illness and normal inflammatory markers assuaged these concerns. The patient was counseled that her severe myelopathic symptoms warranted urgent surgical intervention. The patient was agreeable to surgery but was unfortunately lost to follow-up for 6 months. She returned with the progression of her

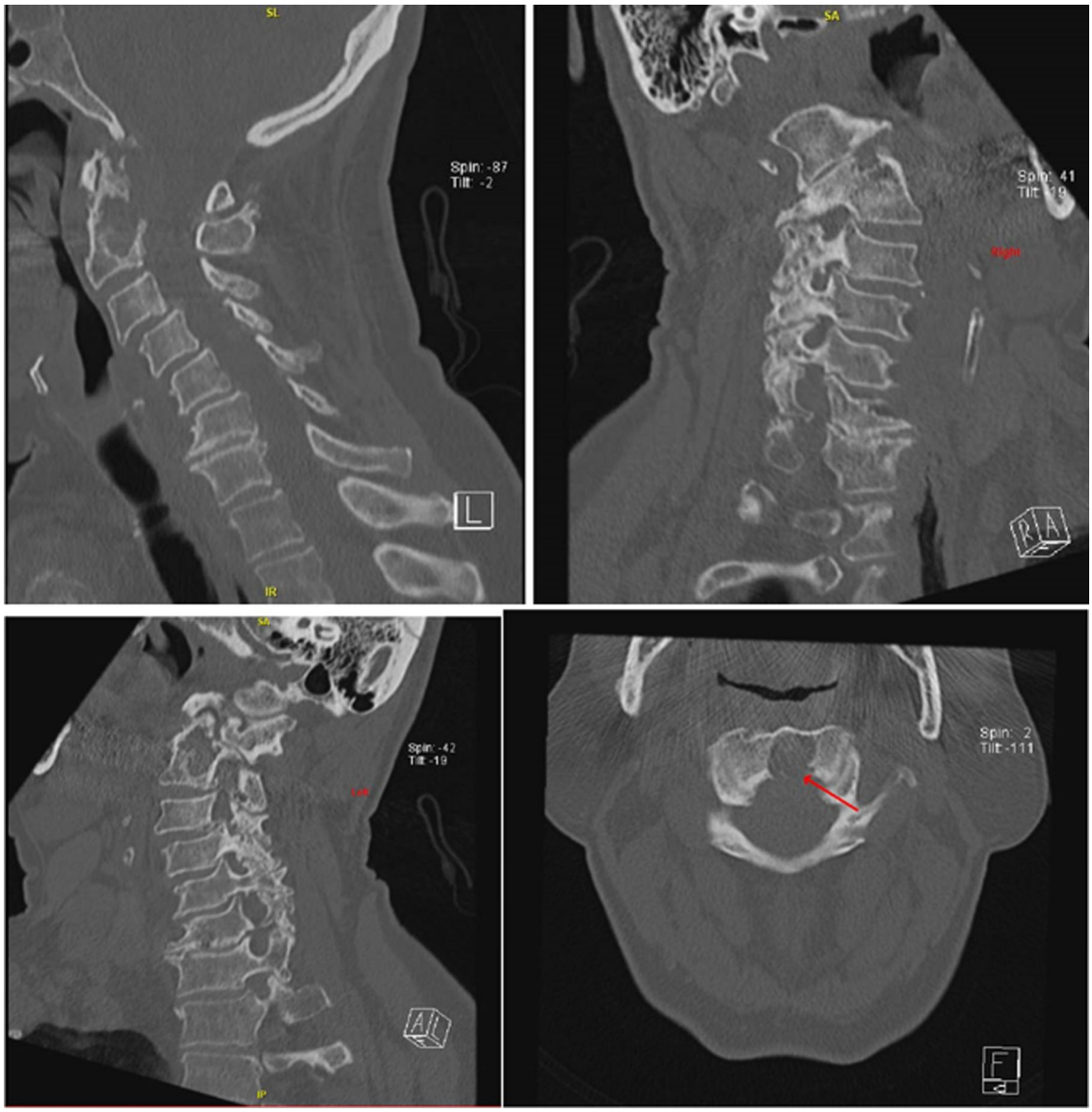

Fig. 2 Preoperative CT Scan. Preoperative CT scan demonstrating the C2 erosive pannus and diffuse subaxial spondylotic changes.

symptoms to the point that she avoided looking up as she knew this would cause her to fall uncontrollably. She also noted progressive issues with fine motor skills and dexterity over the preceding 6 months. Surgical intervention was discussed with the patient to involve multilevel decompression and fusion from a posterior approach with the goal to halt the further neurologic decline. Given the patient's extensive subaxial degenerative changes with anterolisthesis at multiple levels and overall kyphotic alignment, she was counseled regarding the need to extend her fusion throughout the subaxial spine and across the cervicothoracic junction to confer the greatest likelihood of acceptable postoperative sagittal alignment and improved patient-reported outcomes [5, 6].

The patient proceeded to the operating room the following day. After general anesthesia with endotracheal intubation, the patient was positioned prone in standard fashion on a Jackson table with Mayfield pinions. Prior to placing the patient prone, full neuromonitoring was initiated. Motor evoked potentials showed no changes during the supine to prone transition. Subperiosteal exposure was performed to expose the dorsal bony elements from C1–T2 and instrumentation were placed in standard fashion. We then placed lateral mass screws in the subaxial cervical spine, lateral mass screws at C1 using the Harms technique, pars screws at C2, and pedicle screws at T1 and T2. After all the instrumentation was placed, we proceeded to perform our decompressive laminectomy from C1–C4, with a dome laminectomy at C5 in standard fashion (Fig. 4). The patient's neck was then gently brought into extension with the Mayfield pinions prior to placement of our contoured 3.5 titanium rods bilaterally. Finally, we decorticated the lateral masses from C2–C7, the transverse processes and lamina at T1 and T2, and the C1–2 joint. Autograft and morselized cancellous allograft were placed into the fusion bed. The wound was closed in the standard fashion and an incisional wound VAC was used. Neuromonitoring showed no changes in motor evoked potentials or somatosensory evoked potentials throughout the duration of the case.

Post-operatively, the patient was placed in a Miami J cervical collar and was admitted to the orthopedic floor. Post-operative radiographs were obtained and demonstrated reduction of the previous anterolisthesis and acceptable alignment and

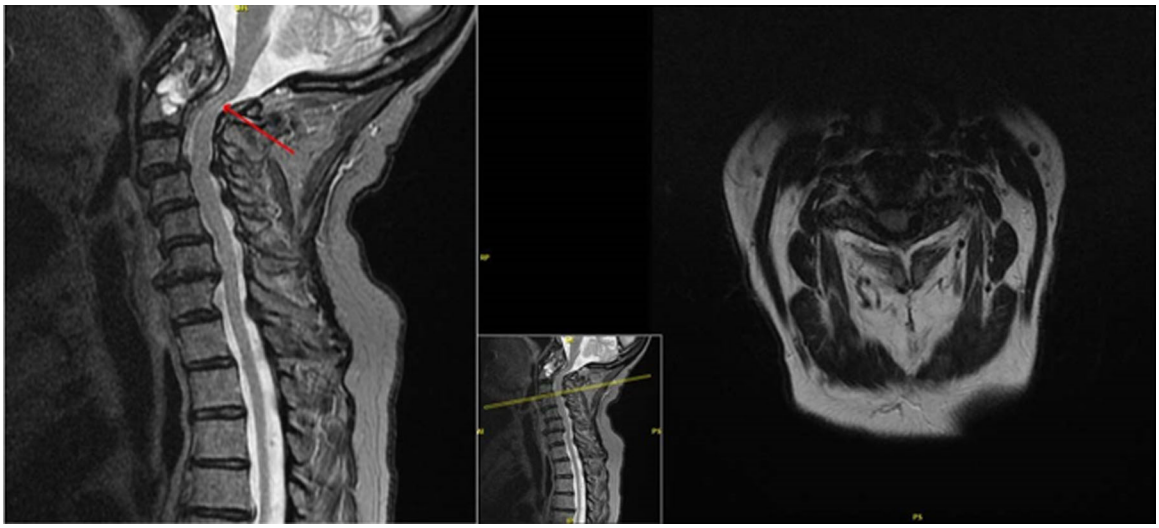

Fig. 3 Preoperative MRI. Preoperative mid-sagittal T2 (left) and axial T1 MRI sequences demonstrating the C2 erosive pannus, myelomalacia (red arrow), and diffuse subaxial spondylotic changes.

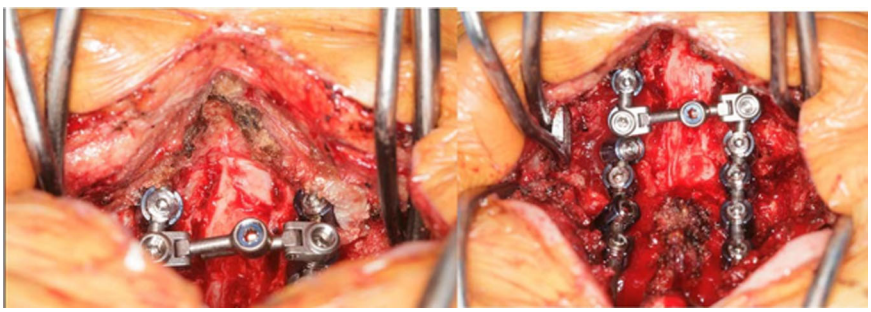

Fig. 4 Intraoperative Photographs. Intraoperative photo demonstrating wide decompression of the cervical spine from C1-4 with a C5 dome laminectomy.

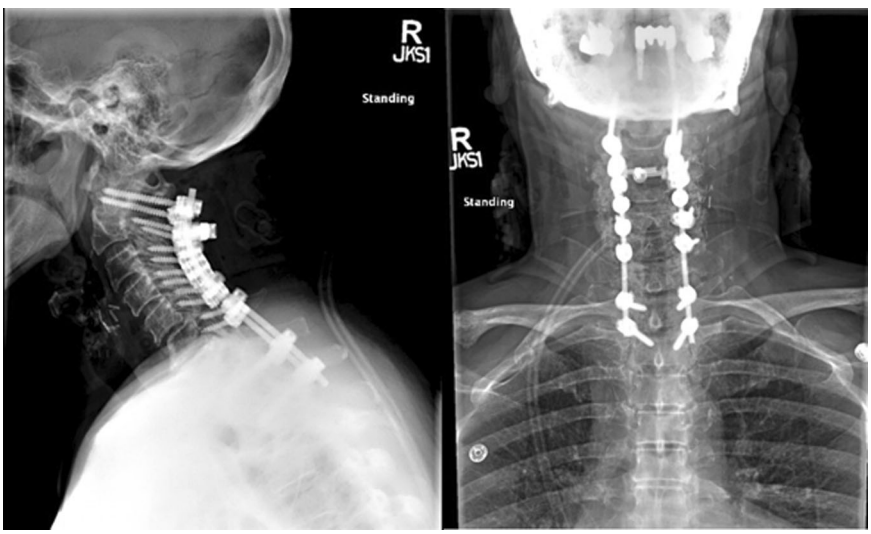

Fig. 5 Early Postoperative Radiographs. Postoperative radiographs were obtained prior to the patient's dismissal from the hospital.

hardware placement (Fig. 5). Mobilization with the help of nurses and PT was encouraged. The cervical collar was gradually weaned after the 6-week mark. Unfortunately, the patient did develop occipital neuralgia that lasted for 18 months. Intermittent injections of the occipital nerve provided some relief. The patient's persistent numbness and paresthesias improved with time. No episodes of severe transient weakness/paralysis or falls were reported postoperatively. At her 2-year postoperative visit, a repeat MRI scan was obtained. This demonstrated significant decompression of the spinal cord at the operative segments with no persistent cord compression or cord signal change. There was also a notable decrease of her degenerative

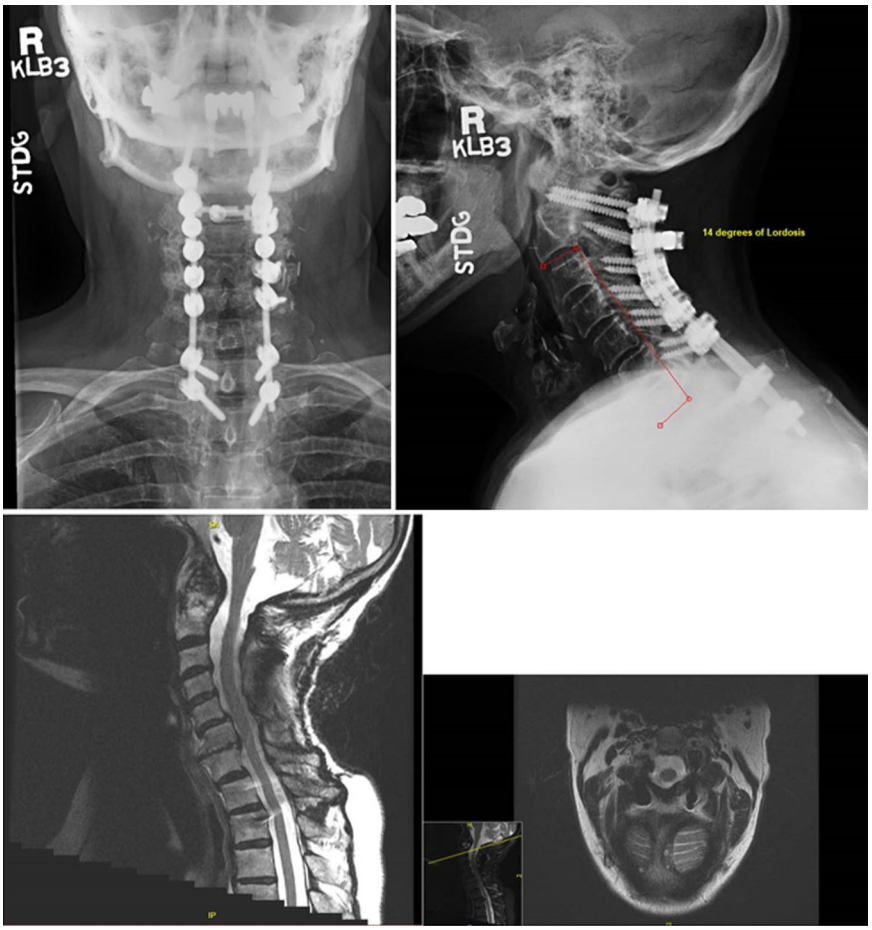

Fig. 6 Two Year Postoperative Radiographs. Two-year postoperative radiographs (top left and top right) and mid-sagittal (bottom left) and axial (bottom right) T2 MRI demonstrate wide patency of the spinal canal at the decompressed levels, complete regression of the peri-odontoid pannus, and maintained cervical alignment.

pannus at C2 (Fig. 6). Her motor strength grading had also improved with time, as most of her upper extremity myotomes graded 5/5 with the exception of finger flexors and interossei muscles, which demonstrated 4+/5 strength. She had full strength in all remaining myotomes in her lower extremities. Her incision was well healed with no signs or concerns for infection. She did report persistent occasional neck aches/stiffness and has noticed a loss of motion in her neck. She expressed her overall satisfaction with her clinical improvement and would repeat the surgical intervention.

DISCUSSION

Progressive cervical myelopathy caused by a compressive peri-odontoid pannus is a rare entity on its own. The most common scenario is that of a patient with underlying history of rheumatoid arthritis. However, case reports and articles in the literature have documented pannus formation in non-rheumatoid patients, resulting in progressive cervical myelopathy with atlanto-axial instability [3, 7–9]. Though histopathologic diagnosis is the gold standard for determining a soft tissue mass to be a pannus, magnetic resonance imaging (MRI) is able to reliably discriminate a pannus from other peri-odontoid soft tissue masses [10]. While specific imaging findings depend upon whether the pannus is hypervascular, hypovascular, or fibrous, a combination of T1-weighted, T2-weighted, and contrast-enhanced MRI sequences can be diagnostic for pannus formation when interpreted by an experienced radiologist [10]. The radiographic features of the cervical spine in our patient were indistinguishable from those seen in rheumatoid patients, and for this reason, rheumatologic evaluation was recommended. The peri-odontoid pannus formation in our patient is likely similar in the pathophysiology to that of an RA patient given their indistinguishable features on radiographic evaluation.

The inflammatory process in the cervical spine predominantly consists of T lymphocytes, macrophages, and plasma cells, and that the process has a predilection for synovial tissues such as the atlanto-occipital and atlanto-dens joints [11, 12]. The inflammatory process will eventually lead to weakening/laxity of the ligamentous structures which may then lead to instability and pannus formation [4, 13]. Given the inextricable association between instability and pannus formation in these patients, it is critical to provide extrinsic stability to the atlantoaxial joint in order to achieve resolution of the pannus. To this end, only one case report in the literature has discussed the successful nonoperative treatment of a peri-odontoid pannus by utilizing a rigid cervical collar for 6 weeks, and the natural history of these lesions is to progress without spontaneous resolution until stability is conferred [14]. In our case report, the patient had episodes of transient paralysis and weakness when flexing or extending her neck likely suggesting a dynamic instability. Because of the manipulation, one needs to perform during surgery, it is crucial to have neuromonitoring, especially in cases with underlying instability.

The occipitocervical complex consists of the bony and ligamentous structures from the occiput to the C2–C3 disk space.

This region provides a major contribution to the motion of the cervical spine [1]. The articulation between the occiput and C1 provides ~50% of flexion and extension and the articulation of C1–2 provides approximately 50% of cervical rotation. It is imperative to inform patients that fusion of the upper cervical spine will lead to severe limitations in cervical range of motion post-operatively. In our patient, loss of range of motion was her primary complaint at the final follow-up. Pre-operative discussion regarding this limitation and its implication on activities such as driving should be addressed.

Fusion of the C1–C2 joint by utilizing a C1 lateral mass screw usually requires manipulation or sacrifice of the C2 nerve root. Although sacrifice of the C2 nerve root in order to adequately visualize the C1 lateral mass as well as the C1–C2 joint has been well described in the literature [15–17], we chose to preserve the nerve root as we had a good working window for C1–2 fusion and were able to gently retract the nerve for our C1 lateral mass screws. It should also be noted that post-operative neuropathic pain does occur and may even have a higher prevalence with C2 nerve transection than C2 nerve preservation [18, 19]. Our patient did develop occipital neuralgia in the post-operative visit, though fortunately, it did improve with time.

At the 2 year visit a repeat MRI and radiographs of the cervical spine were obtained for further post-operative evaluation (Fig. 6). The standing radiographs demonstrated that the hardware was in a stable position and the patient had maintained her alignment. Clinically and radiographically the patient had achieved a solid fusion. The MRI of the cervical spine demonstrated resolution of her pannus and maintained decompression of her cervical spinal cord, with notable improvement in cord signal.

Improvement or resolution of peri-odontoid pannus after surgical stabilization has been described in the literature [4, 20, 21]. As mentioned earlier, instability is usually required to induce pannus formation. The instability of the cervical spine in combination with the progressive buildup of pannus formation can result in spinal cord compression and even sudden death [22]. The result of stabilization of the atlantoaxial joint will cease the excessive motion and, hence, regression of the pannus can be expected over time. Stabilizing the cervical spine via an occipitocervical or an atlanto-occipital fusion for a compressive C2 pannus has both been described. Fusion to the occiput, rather than C1, significantly restricts flexion and extension of the cervical spine. With adequate C1 lateral masses, we try to avoid a fusion to the occiput, but patients are informed pre-operatively that there is a possibility that the fusion may be extended to the occiput if screw fixation is deemed inadequate intra-operatively.

Lastly, the topic of whether to decompress directly vs indirectly has been debated in the literature. Historically a trans-oral approach was favored for direct removal of the odontoid pannus and decompression of the spinal cord [23]. However, more recently spine surgeons have largely trended towards a posterior approach to avoid the complications associated with an anterior procedure [24–26]. As previously mentioned, stabilization of the cervical spine, which ceases the excessive motion/instability of the atlantoaxial joint, can halt the progression and even lead to regression of the peri-odontoid mass. For this reason, some favor posterior cervical fusion without decompression, as the natural history is the regression of the peri-odontoid mass [24, 26, 27]. That being said, we performed a posterior decompression in this patient given her severe myelopathic findings and subaxial stenosis in the setting of myelomalacia. In patients with an isolated erosive C2 pannus with mild symptoms consideration for a fusion without decompression may be appropriate.

CONCLUSION

In this case report, we present a patient with severe progressive cervical myelopathy in the setting of a compressive C2 pannus, without an underlying history of rheumatoid arthritis. The patient demonstrated significant recovery with C1–T2 posterior instrumentation and decompression. The patient had improvement in imaging findings such as reduction in anterolisthesis, regression of the C2 pannus, and signal cord abnormality. Clinically the patient's gait and balance improved. She no longer experienced transient episodes of paralysis and there was a marked improvement in her upper and lower extremity strength. This case report strengthens the current literature that C2 pannus formation is not just a disease process that is seen in rheumatoid arthritis patients. We conclude that in carefully selected patients with C2 pannus formation causing compressive myelopathy, posterior decompression, and arthrodesis without extension to the occiput may be an appropriate surgical treatment.

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COMPETING INTERESTS

The authors declare no competing interests.

ADDITIONAL INFORMATION

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