Retro odontoid pseudo tumor with cervical medullary compression: A case report

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Introduction:
The disease entity of retro odontoid pseudo tumor may over time enlarge and cause cervical medullary compression leading to a progressive myelopathy from extrinsic compression.¹ This mass may also be known as an odontoid pannus or phantom tumor.² In rare instances there may be development of pannus-related cysts that may lead to further compression of the upper spinal cord and lower medulla, as was seen in our case. In the past prior authors have suggested treatment through a trans oral approach with direct removal and decompression.³ In recent literature, more surgeons have been trending towards upper cervical spinal fusion with or without laminectomy for indirect decompression and treatment of the pannus.⁴

Case Report:
A 97-year-old man presented to our neurosurgical service with an inability to ambulate, global paresis of upper and lower extremity, atrophy of the intrinsic hand muscles with long track signs noted in the upper extremity. After work-up was completed, the patient was found to have large odontoid pannus causing compression at the cranial cervical junction. We also noted a cyst associated with this pannus causing significant mass effect on the medulla leading to a T2 hyperintense signal at the medulla consistent with myelomalacia (figure 1). Patient had diffuse spondylosis of the cervical spine along with central stenosis at the 4-5 and 5-6 level. MRI of brain and CT angiogram were obtained and were unrevealing for other pathology. The posterior odontoid mass was found to be due to his rheumatoid arthritis.
Interventions:

After discussion with family, it was decided that the patient would undergo posterior cervical arthrodesis with instrumented fixation from C1-6 with decompressive laminectomy from C4-6 to treat associated stenosis. Intraoperative neuro monitoring was utilized along with fluoroscopy for localization and lateral mass screw placement. No signal changes were noted intraoperatively and patient was successfully extubated. Post-operative x ray is provided (figure 2). Patient was placed in our ICU for post-operative monitoring. Patient tolerated procedure well and his hospital course was uncomplicated. He was sent for a short course of inpatient rehabilitation.

Outcome:

Patient was followed up for two years. There was improvement in the patient’s clinical symptoms of motor weakness and he was eventually able to ambulate without assisted device. The patient tolerated the procedure without any associated surgical or medical complications. Radiographic evaluation at the two-year mark showed marked reduction in the pannus and associated cyst with resolution of T2
hyperintense signal within the medulla (figure 2). To the best of our knowledge there are no reports of such a large odontoid pannus with an associated cyst and medullary compression.

Figure 2: A. x-ray showing C1-6 posterior fusion. B. T2 sagittal MRI showing reduction in cyst size and resolution of signal within medulla. C. T2 MRI showing odontoid process with reduction in pannus size.

Discussion

Retro odontoid pseudo tumor or odontoid pannus can be seen with inflammatory disease entities such as rheumatoid or psoriatic arthritis, and in the non-inflammatory settings, for example chronic dialysis, post-traumatic pseudo arthrosis, and degenerative disease. The primary proposed mechanism is chronic atlanto-axial instability which leads to the development of this pannus. This produces an inflammatory process with fibrocartilaginous mass formation in the posterior odontoid space which over time can lead to direct spinal cord compression. However, there are a few instances where no atlantoaxial instability is noted on imaging and yet there still may be development of this mass lesion. In our case, the patient had an underlying inflammatory spondyloarthropathy, but was under no active treatment. In general, overall incidence is not known but a recent 11-year case series review found that incidence of retro odontoid pannus in patients with symptomatic atlanto-axial instability with associated
risk factors including rheumatoid arthritis, os-odontoideum, dens fracture and Morquio syndrome was 23.2%.

In the past, authors have recommended a direct form of decompression with removal of the odontoid pannus. Surgical treatments included anterior trans oral odontoidectomy and posterior transdural resection. Recently, it has been proposed to surgically fuse the posterior spine, primarily at the C1 to C2 junction. Authors have reported good radiographic and clinical outcomes that have obviated the need for a transoral anterior approach. In our case, fusion not only included C1 and C2, but was extended to the lateral masses of C6. Decompressive laminectomy was completed at the C4 to 6 levels to treat the associated central canal stenosis. Our surgical intervention showed good outcome with resolution of the pannus compression on the medulla and radiographic reduction of the mass. The use of C1-2 fusion is a known safe treatment for atlanto-axial instability and has also been extended as a treatment for painful arthritis. Some authors have recommended occipital cervical fusion, to treat associated occipital cervical instability and adjacent segment disease. This indirect treatment also lead to radiographic improvement with good functional outcome. There is also literature supporting the use of C1 laminectomy for indirect decompression and in some cases it lead to reduction in pannus size. This strategy may be useful in cases where patients may not tolerate prolonged surgeries with larger blood loss and lengthy anesthesia exposure. Concern with completion of C1 laminectomy without fusion may lead to further atlanto-axial instability, but a reduction in neurologic symptoms can be feasibly obtained without progressive instability in most cases.

Conclusion:

Retro odontoid pseudo tumors or odontoid pannus is a disease process characterized by formation of a posterior mass with possible cystic formation, which may lead to marked compression on vital neuronal structures at the cervico-cranial junction. Certain risk factors have been associated with an odontoid
pannus including rheumatoid arthritis, Os odontoneum, or malunion of odontoid fracture. The basis of this disease process is believed to be due to atlanto-axial instability. However, this may not be true for all cases, since it has been found that indirect techniques in the form of C1 laminectomy have helped reduce pannus size. In our case, we proposed cervical fusion and avoided a transoral approach due to our patient’s comorbidities and inability to tolerate this procedure. After our two-year follow-up, the pannus and associated cyst had significant reduction with resolution of cord signal change within the medulla. We support the use of posterior fixation for treatment of this disease process. Further investigation in the pathophysiology of this disease process to help create a more unified treatment approach is needed.
References:


