Odontoid pseudotumor and serial postfusion radiographic evaluation in a patient with a C1–2 mass

Daniel S. Yanni, MD,¹ Alexander Y. Halim, BA,² and Daniela Alexandru, MD¹

¹Department of Neurological Surgery, University of California Irvine School of Medicine, Irvine, California; and ²University of Virginia School of Medicine, Charlottesville, Virginia

Odontoid pseudotumor is a mass occurring around the odontoid process in the cervical spine and can cause significant neurological symptoms at the craniocervical junction due to compression of the spinal cord and cervicomedullary junction at this level. A literature review was performed to provide input on options for treatment and prognosis for this lesion. The literature search found 12 papers in which pseudotumor was treated with posterior decompression and fixation. Posterior decompression and fixation with serial imaging to monitor the size of the pseudotumor postsurgery is a safe and effective treatment option for odontoid pseudotumors.

<http://thejns.org/doi/abs/10.3171/2014.10.SPINE13987>

KEY WORDS pseudotumor; craniocervical junction; posterior fusion; cervical

ODONTOID pseudotumor is a mass occurring around the odontoid process in the cervical spine, and can cause significant neurological symptoms at the craniocervical junction due to compression of the spinal cord and cervicomedullary junction at this level.^{1,22,25} This in turn can lead to severe neurological deficit in patients, which can range from mild myelopathy to complete paralysis in severe cases.^{21,22,25} A retroodontoid mass can occur in patients due to a variety of conditions such as rheumatoid arthritis,^{13,15,16} long-term dialysis,^{17,22} mucinous degeneration of transverse ligament of the atlas,^{6,22} or migrated disc herniation.^{13,21,22} Moreover, retroodontoid pseudotumor can also occur idiopathically. It is postulated that degenerative changes and ligamentous calcification can cause immobility and instability of the lower cervical segments, thus increasing the mechanical stress on the atlantoaxial joint resulting in pseudotumor formation.^{5,19} In this paper we report the case of a 78-year-old man with a C1–2 mass, diagnosed after the patient presented with myelopathic and spastic quadriparetic symptoms. We provide a description of the treatment for this condition, including radiographic follow-up, as well as review the literature on the subject to provide options for treatment and prognosis for this lesion.

Case Report

History and Examination

We report the case of a 78-year-old active businessman

who presented to the emergency room with a 3-week history of significant difficulty with gait and fine motor movements. The patient reported having a 1-year history of progressively worsening symptoms of pain, numbness, and tingling initially in the lower extremities and progressing to the upper extremities. He also complained of worsening bilateral lower-extremity weakness and difficulty ambulating. The patient had a medical history of hypertension and prostate cancer, treated with radiation 10 years prior and with a normal prostate-specific antigen. The patient also had a smoking history (one-quarter pack per day for 15 years), but quit approximately 7 years prior to the presenting symptoms.

On physical examination, the patient had spastic weakness in the bilateral upper and lower extremity (3/5 to 4–/5), profound myelopathic reflexes with hyperreflexia present throughout bilateral upper and lower extremities, clonus in bilateral lower extremities, upgoing bilateral Babinski reflexes, and bilateral positive Hoffman reflex. He had intact sensation and intact rectal tone, with some impairment of proprioception. He underwent MRI of the cervical spine, which showed a C1–2 mass with severe compression of the spinal cord (Fig. 1). The initial differential diagnosis included the following: metastatic tumor (given the patient's history of prostate cancer and a smoking history); a craniocervical junction tumor such as meningioma; a rheumatoid pannus; or odontoid pseudotumor. The patient was started on Decadron, with mild initial improvement of his symptoms. Initial evaluation included a transoral biopsy

SUBMITTED November 2, 2013. **ACCEPTED** October 21, 2014.

INCLUDE WHEN CITING Published online March 13, 2015; DOI: 10.3171/2014.10.SPINE13987.

DISCLOSURE The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

FIG. 1. Sagittal MRI studies of the cervical spine. Admission T1-weighted image obtained with contrast (**A**) and a T2-weighted image (**B**) showing a large mass at the C1–2 level, compressing the spinal cord and mildly enhancing with contrast.

of the lesion; the transoral biopsy was performed due to the patient's smoking history and his history of prostate cancer. Biopsy results were nondiagnostic for malignancy, and only consisted of cartilage and soft connective tissue. Given these biopsy findings, a complete rheumatoid arthritis workup was performed by the rheumatology service. All rheumatological workup was negative for rheumatoid arthritis.

Operation

The patient underwent a suboccipital craniectomy, posterior cervical decompression, occipitocervical instrumentation, and fusion of his cervical spine within days of his presentation to the emergency room. Given the severity of central stenosis and foraminal stenosis due to herniated discs and multilevel degenerative spondylosis with intrinsic cord changes, decompression included a suboccipital craniectomy and C1–5 laminectomies with foraminotomies. Hardware placed included the following: occipital plate; C-1 lateral mass screw; C-2 pedicle screw; and C-3, C-4, and C-5 lateral mass screw placement for stabilization and fusion of the occiput to C-5 (Fig. 2). Intraoperative neuromonitoring revealed a significant improvement in amplitude and latency in all extremities immediately following completion of the C-1 laminectomy. Postoperative imaging revealed excellent placement and alignment of the hardware with wide decompression of the neural elements (Fig. 3). The CT scans and flexion-extension radiographic imaging obtained at 12 months postoperatively revealed excellent fusion from the occiput down to C-5 without evidence of instability.

Postoperative Course

The patient was transferred to the NeuroICU with immediate improvement in neurological examination after surgery. One year after surgery, the patient was at near

FIG. 2. Intraoperative photograph showing decompression and hardware placement.

normal status on neurological examination, with significant improvement in gait, fine motor skill, and strength as well as improvement of myelopathy. On serial radiological examination, the MRI of the cervical spine showed near resolution of the pseudotumor, without impingement on the spinal cord (Fig. 4).

Discussion

Odontoid pseudotumor can cause significant myelopathy in patients with rheumatoid arthritis, long-term dialysis, degeneration of transverse ligament of the atlas, migrated disc herniation, anterior subluxation of the atlantoaxial joint, spondylosis, and pseudarthrosis following fracture

FIG. 3. Postoperative anteroposterior (**A**) and lateral (**B**) radiographic cervical spine series obtained following the posterior cervical decompression and fusion.

FIG. 4. Postoperative T2-weighted MR images obtained at 3 months (A), 6 months (B), and 12 months (C) after posterior decompression and fusion showing progressive resolution of the mass.

of the odontoid process.^{14,22} The literature reports that resection of the mass and stabilization of the craniocervical junction are preferred options for treatment^{5,14} (Table 1).

Our patient did not have a history of trauma, fracture of the odontoid process, rheumatoid arthritis, or history of dialysis. The histopathological analysis of the biopsy specimen showed that the mass was nonneoplastic and was consistent with cartilage and soft connective tissue. Other authors have reported similar cases in the literature as well. Yamaguchi et al. reported a series of 3 cases of idiopathic pseudotumor, in which the patients underwent laminectomy and posterior fusion with complete resolution of symptoms and reduction or disappearance of the mass.²⁴ Additionally, our literature review gives several descriptions of the histopathological appearance of a pseudotumor specimen. Crockard et al. describes the mass associated with pseudotumor as degenerative fibrochondral tissue without evidence of inflammation,¹ similar to the histopathological description in the case we report. Sze et al. determined that the pseudotumor tissue formation was the direct result of chronic atlantoaxial subluxation and had the appearance of a granuloma.¹⁹

Pseudotumor can arise from different ligaments or parts of the ligaments comprising the atlantoaxial joint. Figure 5A illustrates the various ligaments in this region. The cruciate ligament passes from the atlas through the foramen magnum to attach on the posterior surface of the clivus in the cranial base (Fig. 5A). The apical ligament arises anterior to the tip of the odontoid, usually from a small coronal groove, and inserts into the anterior rim of the foramen magnum at the basion (Fig. 5B). The alar ligaments connect the sides of the odontoid to the tubercles on the medial side of the occipital condyle (Fig. 4A). The tectorial membrane is the most superficial layer toward the dura mater, and it is continuous with the posterior longitudinal ligament (Fig. 5B). Although these ligaments are interconnected, our literature review revealed that the most common site of occurrence of the pseudotumor was

from the band of the cruciate ligament²⁴ (Fig. 5C). In the case of our patient, it is difficult to identify which ligament is primarily involved due to the considerable size of the pseudotumor mass (Fig. 1).

Traditionally, 2 approaches have been used for the treatment of odontoid pseudotumor. Some surgeons advocate the direct anterior resection of the pseudotumor via a transoral or a transnasal approach.^{5,23} The most significant advantage of this procedure traditionally was the ability to analyze the tissue directly by histopathological analysis, to evaluate for a neoplastic process. However, this approach has been traditionally fraught with many complications including complicated postoperative management, an increased risk of infection, need for external fixation and/or fusion due to instability, delayed extubation due to the risk of palatal dehiscence, pharyngeal hemorrhage, and airway edema.^{12,24} However, with the current state of neuronavigation, obtaining a small tissue biopsy sample is easier and poses less risk to the patient. In our case we opted to obtain a minimally invasive transoral biopsy sample by using fluoroscopy to decrease the risk to the patient while obtaining a specimen to evaluate for malignancy.

Alternatively, some surgeons recommend posterior decompression and fusion with or without direct mass resection.¹⁹ The posterior approach with decompression and fusion carries fewer potential complications and risks to the patient than an anterior only or a combined approach. We obtained serial MRI evaluation at 3, 6, and 12 months postoperatively to ensure the reduction of the size of the mass, because any increased size or decline in neurological status would have prompted an anterior resection (Fig. 4). Grob and colleagues and Yamaguchi et al. used this approach in their patients to treat deformity of the atlantoaxial region with minimal complications and reduction or resolution of the pseudotumor.^{8,9,24} The extent of the fusion should be evaluated for each patient individually based on clinical, neurophysiological, and radiographic findings. For a small pseudotumor, a C1–2 fusion may fix the unsta-

TABLE 1. Literature review of cases of pseudotumor at the craniocervical junction along with treatment and follow-up

Authors & Year	Presenting Symptoms	Causes	Treatment	Results
Rosenberg et al., 1991	Intermittent paresthesias of the lt hand spreading progressively to the lt forearm, arm, & leg; decreased hearing in lt ear & recent 2-day history of numbness over the entire lt side of the body	Retroodontoid ruptured discs appearing as a mass	Suboccipital craniectomy w/ C-1, C-2, & partial C-3 laminectomies & mass drainage	Marked reduction in the size of the retroodontoid mass, w/o evidence of recurrence on MRI
Choe et al., 1993	Numbness of hands & feet & weakness of extremities, progressive difficulty in balancing, & lower occipital headache; positive Romberg sign & mild dysmetria	Synovial cyst of the dens causing spinal cord compression	Transoral resection of C-1 arch & dens, cyst removal, & upper cervical spine fusion	Recovery of strength & sensation in the patient's arms & legs
Cho et al., 2001	Hearing difficulties in the rt ear & occasional pain in the rt temporal bone & temporomandibular joint	Idiopathic inflammatory pseudotumor	60 mg prednisolone/day for 10 days & tapered over the following 4 days, & 2 subsequent RT treatments	Short-term symptomatic relief w/ recurrence of mass at 4-mo follow-up
Zorzon et al., 2001	Numbness, incoordination, & clumsiness in both hands, gait instability, & bladder & bowel dysfunction	Synovial cysts of the atlantodental articulation	C-1 hemilaminectomy & partial C-2 hemilaminectomy, complete evacuation of the cyst contents, & subtotal removal of the cyst wall	Symptomatic relief w/o recurrence of cyst
Jun et al., 2002	Spastic tetraparesis, loss of joint position sensation, & stiffness in the legs	Retroodontoid pseudotumor w/ diffuse idiopathic skeletal hyperostosis	Transoral removal of the extradural mass & dorsal atlantoaxial fusion	Marked decrease in spasticity & improved muscle strength
Sagiuchi et al., 2006	Spastic tetraparesis, 4/5 strength in all extremities, all deep tendon reflexes hyperactive, bilateral Babinski sign, dysesthesia in patient's hands & toes	Extradural cyst extending posteriorly to the dens & compressing the upper cervical cord, & atlantoaxial subluxation	Cyst: Philadelphia collar; atlantoaxial subluxation: C1–2 posterior fusion	Spontaneous regression of cyst & successful C1–2 posterior fusion for atlantoaxial subluxation
Ito et al., 2007	Occipital pain, numbness of both UEs, & mild weakness of the rt UE	Rheumatoid arthritis–negative atlantoaxial dislocation complicated by an extradural mass in the craniovertebral junction	C-1 laminectomy w/ posterior fixation of atlantoaxial dislocation	Motor weakness of the rt UE & sensory disturbance of both UEs improved, & 3-mo follow-up imaging showed reduced retroodontoid mass size
Takami et al., 2007	Progressive myelopathy manifesting as clumsy hand & gait disturbance after a fall	Retroodontoid pseudotumor	C-1 laminectomy & posterior C1–2 fixation w/ individual screw placement in C-1 & C-2	At 6 mos postop, no evidence of instability in the atlantoaxial joint & diminished retroodontoid pseudotumor
Cecchi et al., 2008	Progressive tetraparesis, ataxic gait, & cervical pain	Atlantoaxial degenerative articular cyst	Philadelphia collar & a short-term course of NSAIDs & corticosteroids	Almost complete regression of atlantoaxial cyst & symptomatic relief
Goel & Dange, 2008	Spastic quadriparesis & only able to walk w/ the aid of crutches	Rheumatoid retroodontoid pannus, fixed atlantoaxial dislocation, basilar invagination, & significant cord compression	Lateral mass reconstruction w/ distraction of the facets & impaction of a spiked metal spacer & bone graft w/in the joint	Complete disappearance of retroodontoid pannus & reduction of basilar invagination & atlantoaxial dislocation
Tanaka et al., 2010	Severe myelopathy w/ motor weakness of both UEs, sensory disturbance in all 4 extremities, clumsy hand movements, increased deep tendon reflex, & a positive Babinski reflex	Retroodontoid pseudotumor	Surgical decompression of posterior spinal canal by laminectomy of the atlas & the axis & posterior C1–2 transarticular fixation w/o mass resection	Immediate symptomatic relief & marked reduction in mass size & spinal compression

RT = radiotherapy; UE = upper extremity.

FIG. 5. Schematic drawings showing the anatomy of the atlantoaxial joint. **A:** Ligaments of the atlantoaxial joint. **B:** Sagittal view of the atlantoaxial joint in a normal spine. **C:** Sagittal view of the atlantoaxial joint in a patient with a pseudotumor. Copyright Daniela Alexandru. Published with permission.

ble segment and help reduce the mass. For larger masses, which involve multiple ligaments around the odontoid and produce craniocervical spinal cord compression, a larger surgery may be needed with extensive decompression and fusion including C-1 and the occiput.

Conclusions

Odontoid pseudotumor is a mass that surrounds the odontoid process and can cause significant neurological complications due to compression of the neural elements, including brainstem and upper cervical spinal cord. Managing a lesion such as this requires careful assessment and evaluation. From our literature review it appears that in the absence of a neoplastic process, a posterior cervical decompression and fusion can be safely performed to decompress and stabilize the patient's spine and prevent further injury (Table 1). Resection of the mass may be performed at this time, although our serial radiographic follow-up and the literature review indicate that the pseudotumor mass regresses over time once the decompression and stabilization procedure has been performed. This finding supports the previous literature (Crockard et al.⁵ and Sze et al.¹⁹) findings that chronic instability and degeneration may form the pseudotumor, and that stabilization of this joint can arrest or reverse the pseudotumor. Serial MR images can be used to monitor the reduction in size of the mass. Our patient demonstrated near resolution of his pseudotumor on radiological examination and a near normal neurological examination at 1 year postsurgery. It appears from our literature search that posterior decompression and fixation with serial imaging to monitor the size of the pseudotumor postsurgery is a safe and effective treatment option for odontoid pseudotumors.

Acknowledgment

The authors would like to acknowledge Christian Rafla for his assistance in the literature search.

References

1. Boden SD: Rheumatoid arthritis of the cervical spine. Surgical decision making based on predictors of paralysis and recovery. *Spine (Phila Pa 1976)* **19**:2275–2280, 1994
2. Cecchi PC, Peltz MT, Rizzo P, Musumeci A, Pinna G, Schwarz A: Conservative treatment of an atlantoaxial degenerative articular cyst: case report. *Spine J* **8**:687–690, 2008
3. Cho YS, Kim SM, Chung WH, Hong SH: Inflammatory pseudotumor involving the skull base and cervical spine. *J Laryngol Otol* **115**:580–584, 2001
4. Choe W, Walot I, Schlesinger C, Chambi I, Lin F: Synovial cyst of dens causing spinal cord compression. Case report. *Paraplegia* **31**:803–807, 1993
5. Crockard HA, Sett P, Geddes JF, Stevens JM, Kendall BE, Pringle JA: Damaged ligaments at the craniocervical junction presenting as an extradural tumour: a differential diagnosis in the elderly. *J Neurol Neurosurg Psychiatry* **54**:817–821, 1991
6. Fransen P, Pizzolato GP, Otten P, Reverdin A, Lagier R, de Tribolet N: Synovial cyst and degeneration of the transverse ligament: an unusual cause of high cervical myelopathy. Case report. *J Neurosurg* **86**:1027–1030, 1997
7. Goel A, Dange N: Immediate postoperative regression of retroodontoid pannus after lateral mass reconstruction in a patient with rheumatoid disease of the craniovertebral junction. Case report. *J Neurosurg Spine* **9**:273–276, 2008
8. Grob D: Posterior occipitocervical fusion in rheumatoid arthritis and other instabilities. *J Orthop Sci* **5**:82–87, 2000
9. Grob D, Schütz U, Plötz G: Occipitocervical fusion in patients with rheumatoid arthritis. *Clin Orthop Relat Res* **(366)**:46–53, 1999
10. Ito H, Neo M, Yoshida M, Fujibayashi S, Yoshitomi H, Nakamura T: Efficacy of computer-assisted pedicle screw insertion for cervical instability in RA patients. *Rheumatol Int* **27**:567–574, 2007
11. Jun BY, Yoon KJ, Crockard A: Retro-odontoid pseudotumor in diffuse idiopathic skeletal hyperostosis. *Spine (Phila Pa 1976)* **27**:E266–E270, 2002
12. Kingdom TT, Nockels RP, Kaplan MJ: Transoral-transpharyngeal approach to the craniocervical junction. *Otolaryngol Head Neck Surg* **113**:393–400, 1995
13. Kosaka E, Ishihara H, Osada R, Nakamura H, Kimura T: Periodontoid pannus migration into the spinal canal with re-

- duction of rheumatoid atlantoaxial subluxation: a case report. **J Orthop Sci** 7:703–706, 2002
14. Lansen TA, Kasoff SS, Tenner MS: Occipitocervical fusion for reduction of traumatic periodontoid hypertrophic cicatrix. Case report. **J Neurosurg** 73:466–470, 1990
 15. Oohori Y, Seichi A, Kawaguchi H, Tajiri Y, Oda H, Nakamura K: Retroodontoid pseudotumor resected by a high cervical lateral approach in a rheumatoid arthritis patient: a case report. **J Orthop Sci** 9:90–93, 2004
 16. Rosenberg WS, Rosenberg AE, Poletti CE: Cervical disc herniation presenting as a mass lesion posterior to the odontoid process. Report of two cases. **J Neurosurg** 75:954–959, 1991
 17. Rousselin B, Helenon O, Zingraff J, Delons S, Druke T, Bardin T, et al: Pseudotumor of the craniocervical junction during long-term hemodialysis. **Arthritis Rheum** 33:1567–1573, 1990
 18. Sagiuchi T, Shimizu S, Tanaka R, Tachibana S, Fujii K: Regression of an atlantoaxial degenerative articular cyst associated with subluxation during conservative treatment. Case report and review of the literature. **J Neurosurg Spine** 5:161–164, 2006
 19. Sze G, Brant-Zawadzki MN, Wilson CR, Norman D, Newton TH: Pseudotumor of the craniovertebral junction associated with chronic subluxation: MR imaging studies. **Radiology** 161:391–394, 1986
 20. Takami T, Goto T, Tsuyuguchi N, Nishikawa M, Ohata K: Posterior C1-2 Fixation with cancellous screw and rod system for retro-odontoid pseudotumor associated with chronic atlantoaxial subluxation. Technical note. **Neurol Med Chir (Tokyo)** 47:189–194, 2007
 21. Tanaka S, Nakada M, Hayashi Y, Mohri M, Hayashi Y, Uchiyama N, et al: Retro-odontoid pseudotumor without atlantoaxial subluxation. **J Clin Neurosci** 17:649–652, 2010
 22. Tojo S, Kawakami R, Yonenaga T, Hayashi D, Fukuda K: Factors influencing on retro-odontoid soft-tissue thickness: analysis by magnetic resonance imaging. **Spine (Phila Pa 1976)** 38:401–406, 2013
 23. Vougioukas VI, Hubbe U, Schipper J, Spetzger U: Navigated transoral approach to the cranial base and the craniocervical junction: technical note. **Neurosurgery** 52:247–251, 2003
 24. Yamaguchi I, Shibuya S, Arima N, Oka S, Kanda Y, Yamamoto T: Remarkable reduction or disappearance of retroodontoid pseudotumors after occipitocervical fusion. Report of three cases. **J Neurosurg Spine** 5:156–160, 2006
 25. Yonezawa I, Okuda T, Won J, Sakoda J, Nakahara D, Nojiri H, et al: Retrodental mass in rheumatoid arthritis. **J Spinal Disord Tech** 26:E65–E69, 2013
 26. Zorzon M, Skrap M, Diodato S, Nasuelli D, Lucci B: Cysts of the atlantoaxial joint: excellent long-term outcome after posterolateral surgical decompression. Report of two cases. **J Neurosurg** 95 (1 Suppl):111–114, 2001

Author Contributions

Conception and design: Yanni, Halim. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: Alexandru, Halim. Critically revising the article: Yanni, Halim. Reviewed submitted version of manuscript: all authors. Statistical analysis: all authors. Administrative/technical/material support: all authors. Study supervision: Yanni.

Correspondence

Daniel S. Yanni, Department of Neurosurgery, UC Irvine Medical Center, 200 S. Manchester Ave., Ste. 210, Orange, CA 92868. email: dyannimd@hotmail.com.