79 YEAR OLD WOMAN PRESENTS WITH BILATERAL HAND AND ARM NUMBNESS / DECREASED COORDINATION

> Emily Arciero, MS4 Michael Baldwin, MD



Clinical History

- 79 year old female with history significant for L1-L5 spinal fusion years prior, recently diagnosed with L5-S1 spondylolisthesis
 - Other PMHx- HTN, HLD, T2DM, PAD, depression
- Patient had been admitted to hospital for syncopal episode (May 2020)
 - Head CT without contrast showed severe central spinal stenosis/cord compression at atlantoaxial joint due to possible soft tissue mass
 - CT of cervical spine without contrast performed, corroborated initial head CT findings
- Subsequently referred to outpatient orthopaedic spine clinic, had first visit in June 2020



Presenting Symptoms

• Initial visit to orthopaedic spine clinic:

- Patient endorsed a 2 month history of bilateral hand numbress with loss of coordination
 - Numbness in hands radiating to mid-forearm
- Loss of balance, felt as though she was "drunk" when trying to walk
- No change in bowel/bladder function
- Symptoms progressively worsening
- Patient reported being in car accident about 2 months ago, started noticing symptoms after the event



Extra Information

Physical exam

- First visit (June 2020)
 - Significant ataxic gait
 - 3 beats of clonus bilaterally
 - Hoffmann negative bilaterally
 - Strength 5/5, reflexes 2+ bilaterally in upper and lower extremities
- Second visit (July 2020)
 - Hyperreflexia of bilateral upper extremities
 - Inversion of brachioradialis reflex
- Labs
 - Rheumatoid factor negative in 2018



CT Scan C Spine w/o Contrast

















Moderate degenerative atlantodental arthropathy

Mild to moderate atlantoaxial arthrosis







MRI C Spine w/o Contrast





Heterogeneous, overall hypointense, welldemarcated retro-odontoid mass compressing spinal cord

No T2 hyperintense signal surrounding spinal cord at C1/C2 level

T2 hyperintensity within spinal cord

R

Sagittal T2

C5

TI

Axial T2



MRI C Spine w/ and w/o Contrast









Diagnosis?



Dx = Retro-Odontoid Pseudotumor

Retro-odontoid pseudotumor (ROP)

- Defined as soft tissue proliferative changes at the atlantoaxial junction surrounding the region of the transverse ligament
- Clinical presentation
 - Often asymptomatic
 - Acute inflammation in these masses can manifest as neck pain/headache
 - As a chronic process, mass effect on cervical spine can manifest as myelopathy, including sensory and motor deficits



DDx Retro-Odontoid Pseudotumor

The following entities may cause retro-odontoid pseudotumor:

- Chondrocalcinosis
 - Calcific deposits in transverse ligament- tend to be linear and diffuse
 - Associated pyrophosphate arthropathy → narrowing of intervertebral discs, bony sclerosis, osteophyte formation, often with bony erosions and subchondral cysts
- Hemodialysis-associated amyloidosis
 - Bony erosion and cystic change
- Pigmented villonodular synovitis
 - Heterogeneously low signal intensity
 - Rarely forms osteolytic bone lesions
 - Blooming on GRE sequences secondary to hemosiderin content
- Chronic odontoid fracture
 - Poor healing \rightarrow altered/increased movement \rightarrow formation of fibrous soft tissue mass around fracture site
- Gout
 - Tophi may be faintly calcified
 - MRI appearance of tophus can be indistinguishable from calcium hydroxyapatite crystal deposit
- Pannus (RA)- can be hypervascular, hypovascular, or fibrous
- Osteoarthritis

*although ROP has been shown to be related to mechanical instability, prior studies have not shown a single specific cause for soft tissue proliferation

RADIOLOGY

HEAL

DDx Retro-Odontoid Pseudotumor

Other retro-odontoid entities:

Retro-odontoid synovial cyst

- Simple fluid signal intensity cystic structure (although occasionally can have a complex appearance due to neovascularization and hemorrhage)
- Epidural lipomatosis
 - High T1 signal, fat suppression on T1 FS, low T2 signal
- Epidural hematoma
 - MRI appearance of hemorrhage varies based on acuity
 - High T1 signal, no fat suppression on T1 FS, low T2 signal
 - >75% found dorsally within spinal canal
- Ossification of posterior longitudinal ligament
 - Isointense to cortex and usually extends caudal to craniocervical junction
 - Unlike in pseudotumor- varied signal intensity, centered at posterior aspect of dens



Surgical Intervention

- C1-C3 posterior decompression and C1-C4 instrumented posterior spinal fusion
- Resulted in significant interval decrease in prominence of soft tissue pannus along the posterior aspect of the dens, along with no cord compression at the C1 level





RADIOLOGY

Post-Op XR C Spine

Bilateral lateral mass screws at C1





Post-Op MRI C Spine w/o Contrast (6 mo f/u)







tissue prominence along posterior aspect of dens



Restoration of T2 hyperintensity surroundin spinal cord at C1-C3



Table 1	Imaging features of various causes of retro-odontoid pseudotumor						
Etiology		T1 signal	T2 signal	STIR signal	Ossification	Erosions	Histopathology
Rheumatoid arthritis							
Hypervascular pannus		High	High	High	No	Yes	Granulation tissue with large amount of inflammatory cells, angioblasts, vessels, and edema [35, 38]
Hypovascular pannus		Low	High, mixed	Low			Connective tissue with poorly vascularized collagen fibers [35, 38]
Fibrous pannus		Low	Low	Low			Nonvascularized fibrous connective tissue with few cells [35, 38]
Osteoarthritis		Low	Low	Low	No	Rarely	Dense fibrous tissue proliferation with immature bone formation [72]
Retro-o	odontoid synovial cyst	Low	High	High	No	No	Degenerative ligamentous changes with inflammatory reaction [22, 67]
	dialysis-associated loidosis	Low to intermediate	Low to intermediate	Low	No	Yes	Extracellular deposition of the fibrous protein β2-microglobulin [59, 60]
CPPD		Low	Variable, heterogenous	Low	Yes	Yes	Inflammatory cells with positively birefringent rhomboid crystals (calcium pyrophosphate) [73]
PVNS		Low to intermediate	Low to intermediate	High	No	Yes	Mononuclear histiocytes mixed with multinucleated giant cells; interspersed hemosiderin deposition [62]
Ossification of Posterior Longitudinal Ligament		Low	Variable	Low	Yes	No	Formation of ossific-calcific components in the PLL [71]
Gout		Low	Variable, heterogenous	High	Yes, faintly calcified	Yes	Deposition of monosodium urate crystals [74]
Fracture		Low	High, mixed	High	Yes	Maybe	Callus formation, remodeling of fracture, reactive fibrous changes [65]
Epidural lipomatosis		High	Low	Low	No	No	Mature adipose tissue [68]
Epidural hematoma		High	Low	Low	No	No	Blood clot [69, 70]

Anatomy

- Normal soft-tissue anatomy of retroodontoid region:
 - Cruciate ligament (transverse ligament and longitudinal band)
 - Posterior longitudinal ligament
 - Thecal sac

 \bullet

Given the synovial articulation b/w the transverse ligament and odontoid, there is a potential space that can be affected by inflammatory and reparative processes → resulting in proliferative soft tissue



UCONN

HEALTH

RADIOLOGY

Pathophysiology

Repetitive cycle of ligamentous injury and repair likely drive development, initiated by transverse ligament damage from inflammation, minor trauma, degenerative changes, or altered biomechanics

- *Regardless of the etiology, spontaneous regression of ROP is commonly observed following surgical fusion (as in this case)
 - This further supports the theory that XS motion at the craniocervical junction may be a major contributing factor in the development of abnormal soft tissue





Radiographic Features

Soft tissue thickening at the atlantoaxial junction around the expected location of the transverse atlantal ligament, posterior to the dens

- CT
 - Mineralization within the pseudotumor
 - Bony erosion may be present in RA, CPPD, gout, pigmented villonodular synovitis
- MRI- signal changes vary by etiology → no reliable imaging features that distinguish RA-related pannus from non-RA retro-odontoid pseudotumor
 - T1- usually hypointense
 - T1 precontrast- identification of hemorrhage, fibrous tissue, mineralization
 - T2 post contrast- to characterize vascularity of pseudotumor
 - T2- variable, often heterogeneous
 - This sequence can be used to identify edema within odont of CONN pseudotumor, or C spine

RADIOLOGY

Discussion/Conclusion

- Retro-odontoid pseudotumor is an uncommon entity with varied and overlapping imaging appearances described on MRI in patients with and without RA
 - ROP is a diverse entity commonly mislabeled as pannus
 - In the setting of RA, retro-odontoid soft tissue proliferation is most commonly referred to as pannus, although histological reports are rare and do not always confirm inflammation
- Pannus-like lesions have been reported in patients with severe degenerative disease in the cervical spine
 - This patient had extensive multilevel degenerative changes
 - No prior diagnosis of RA
- Imaging findings in this case (low T1 signal (nonenhancing) and low T2 signal) consistent with soft tissue proliferation seen in osteoarthritis (dense fibrous tissue with immature bone formation), or a fibrous pannus (nonvascular fibrous connective tissue with few cells)
- Regardless of etiology, ROP tend to regress after stabilization of the C spine, which was observed in this patient

RADIOLOGY

ΕA

References

- Shi J, Ermann J, Weissman BN, Smith SE, Mandell JC. Thinking beyond pannus: a review of retro-odontoid pseudotumor due to rheumatoid and non-rheumatoid etiologies. Skeletal Radiol. 2019 Oct;48(10):1511-1523. doi: 10.1007/s00256-019-03187z. Epub 2019 Mar 13. PMID: 30868232.
- Kushchayev SV, Glushko T, Jarraya M, Schuleri KH, Preul MC, Brooks ML, Teytelboym OM. ABCs of the degenerative spine. Insights Imaging. 2018 Apr;9(2):253-274. doi: 10.1007/s13244-017-0584-z. Epub 2018 Mar 22. PMID: 29569215; PMCID: PMC5893484.
- Joyce AA, Williams JN, Shi J, Mandell JC, Isaac Z, Ermann J. Atlanto-axial Pannus in Patients with and without Rheumatoid Arthritis. J Rheumatol. 2019 Nov;46(11):1431-1437. doi: 10.3899/jrheum.181429. Epub 2019 Apr 1. PMID: 30936276.
- Park JH, Lee E, Lee JW, Kang Y, Ahn JM, Yeom JS, Kang HS. Postoperative Regression of Retro-odontoid Pseudotumor After Atlantoaxial Posterior Fixation: 11 Years of Experience in Patients With Atlantoaxial Instability. Spine (Phila Pa 1976). 2017 Dec 1;42(23):1763-1771. doi: 10.1097/BRS.000000000002222. PMID: <u>28459776</u>.
- Pierce JL, Donahue JH, Nacey NC, Quirk CR, Perry MT, Faulconer N, Falkowski GA, Maldonado MD, Shaeffer CA, Shen FH. Spinal Hematomas: What a Radiologist Needs to Know. Radiographics. 2018 Sep-Oct;38(5):1516-1535. doi: 10.1148/rg.2018180099. PMID: 30207937.
- <u>https://radiopaedia.org/articles/retro-odontoid-pseudotumour?lang=us</u>

