Strange Case of Seronegative RA: Initial Presentation with Cervical Spine Pannus and Erosion

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Introduction

Rheumatoid arthritis (RA) is a chronic autoimmune disease with articular and extra-articular manifestations. It typically affects small to medium joints, but it can affect the spine C1-C2 in prolonged disease (>10 years). Atypical presentations are important to recognize.

- Inflammation -> Pannus = synovial lining cells that can destroy bone and cartilage[1]
- Cervical pannus can be secondary to RA, spine surgery/trauma, steroids, idiopathic

Case

- CC: 64-year-old woman with hx hereditary hemochromatosis, presenting with neck pain of 2 months in duration, with stiffness and limited range of motion.

- HPI: Episodic pain in the neck that came as flares, sometimes associated with headaches, balance disturbances, and brief paresthesias.
- Initial flare improved with physical therapy, anti-inflammatory medications (ketorolac, nabumetone), & muscle relaxants (cyclobenzaprine), but then later recurred.
- No previous spine surgeries, trauma, steroid use, nor evidence of pseudogout.
- Physical exam: mild bilateral wrist tenderness with no synovitis on exam.
- ESR, CRP wnl; ANA, RF, CCP, HLA-B27 negative
- Hand X-rays: carpal crowding, joint space narrowing, & early carpal destruction. Narrowing of the scaphoid, trapezoid, trapezium joint space with a paucity of productive bony changes raising the possibility of an inflammatory arthropathy.

Figures

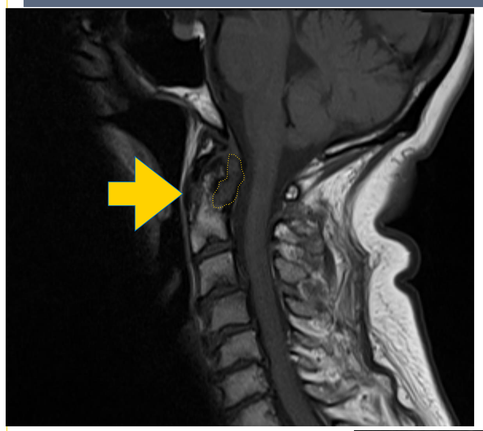
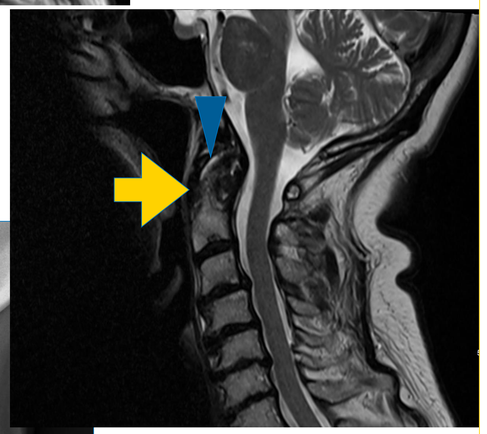


Figure 1: C-spine MRI (top left) T1 and (middle right) T2 show atlantoaxial pannus formation (yellow dotted shape) with moderate edema within C1-C2 (blue triangle) and erosion of the dens (yellow arrows).



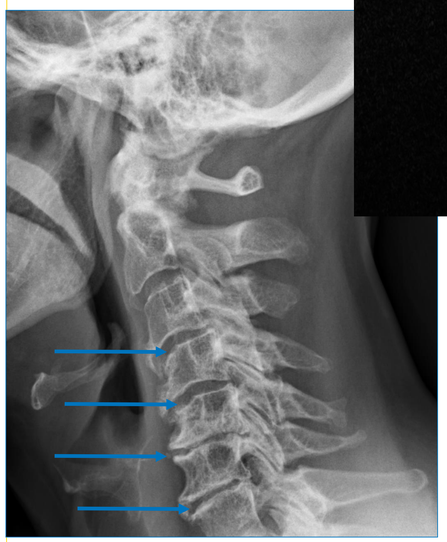


Figure 2: (left) C-spine X-ray: decreased C3-C7 disc spaces (blue arrows), disc disease with spurring, & facet arthropathy. Overall: marked degenerative discogenic & arthritic changes.

Working Dx: Seronegative RA with C-spine involvement as the presenting feature.

Discussion

- Unusual presentation of RA with early pannus formation and carpal crowding, which typically occur after years.
- Prior cervical pannus cases -> RA diagnosis
  - 80yM with normal ESR/CRP/RF but low +anti-CCP; 63yF +ESR/+CRP but **-RF/-anti-CCP** [2]
  - 64y patient with +RF/+CCP [3]
  - 35yM with +RF/+anti-CCP [4]
- Prior cervical pannus cases with seronegative RA
  - In *Joyce et al*, 29/105 (27.6%) confirmed cervical pannus cases had RA (28/29 had known RA). RA patients were younger than non-RA patients (median age 63 yrs, IQR 49–71 yrs versus 79 yrs, IQR 72–85 yrs; $p < 0.0001$), more often female (86% vs 55%, $p = 0.0032$), less likely to have prior cervical spine surgery (0% vs 18%, $p = 0.016$). RA patients' median disease duration 19 years (IQR 6–69 yrs) and 73% (19/26 patients with available data) were seropositive, **thus 27% (7/26) of the cervical pannus patients with RA were seronegative** [5]

Conclusions

- C-spine involvement may be the presenting symptom in seronegative RA, as opposed to what was previously believed, that it only occurs in late disease.
- Crucial early identification, before atlantoaxial subluxations/life-threatening spine or brainstem compressions
- Highlights the challenge and importance of diagnosing RA based on atypical presentations.

References

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