

# Teaching NeuroImage: Crowned Dens Syndrome

## An Acute Attack of Calcium Pyrophosphate Deposition Disease Mimicking Acute Meningitis

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**Table** Laboratory Data on Admission and at Discharge 10 Days Later

Variable	On admission	At discharge	Normal range
<b>Blood</b>			
White blood cell count ( $\times 10^9/L$ )	30.5	10.75	4.8–10.8
Neutrophils ( $\times 10^9/L$ )	25.73	8.98	1.50–6.50
Lymphocytes ( $\times 10^9/L$ )	1.00	1.15	1.20–3.40
Monocytes ( $\times 10^9/L$ )	3.76	0.59	0.30–0.60
C-reactive protein (mg/dL)	26.4	3.42	<0.5
Erythrocyte sedimentation rate (mm/h)	65	23	1–20
Glucose (mg/dL)	141	—	70–110
Rheumatoid factor (KIU/L)	Negative	—	
Anticyclic citrullinated peptide antibody (RU/mL)	Negative	—	
<b>CSF</b>			
Red blood cell count (per $\mu L$ )	Absent	—	
White blood cell count (per $\mu L$ )	2	—	
Protein (mg/dL)	54	—	10–45
Glucose (mg/dL)	99	—	40–70
Gram's stain	No bacteria seen	—	
Viral and bacterial multiplex PCR assay	Negative	—	
CSF cultures	Negative	—	

An 88-year-old man presented with acute onset of severe neck pain, meningismus, headache, and fever (up to 38°C). Blood tests showed raised inflammatory markers, but CSF analysis was not suggestive of CNS infection (Table). Autoimmune antibodies were negative. Cervical spine CT showed calcifications of the transverse ligament of the atlantoaxial joint with inflammatory changes revealed by cervical spine MRI (Figure). Clinical presentation and biochemical and radiologic findings were all consistent with crowned dens syndrome.<sup>1,2</sup> Symptoms and inflammatory markers promptly decreased after a brief course of anti-inflammatory treatment. Recognition of this syndrome is important to differentiate it from other infectious or autoimmune diseases and avoid unnecessary treatment.

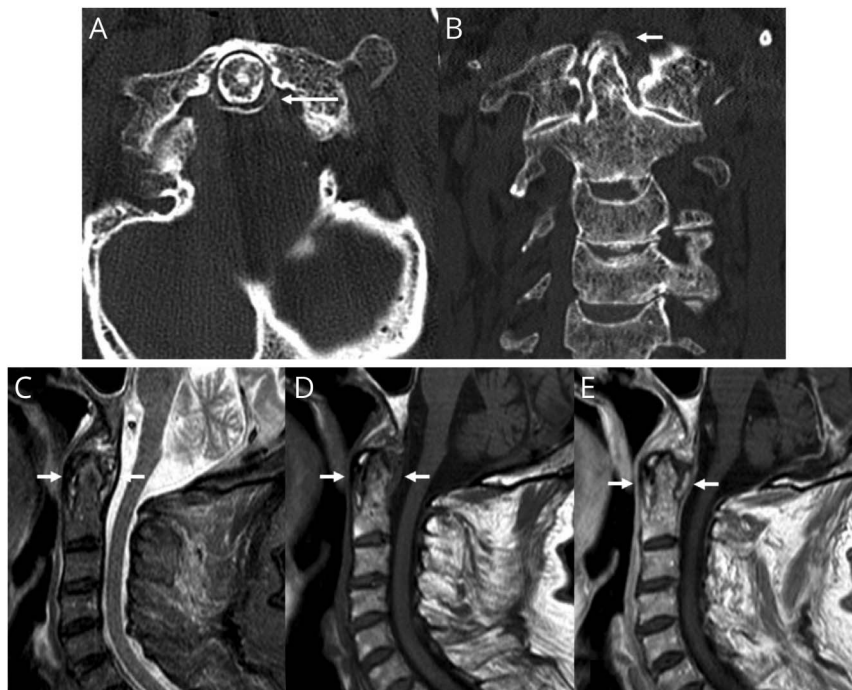
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**Figure** Imaging Features of an Acute Attack of Calcium Pyrophosphate Deposition Disease in the Atlantoaxial Joint



(A) Axial CT image of the atlantoaxial joint shows curvilinear calcification of the transverse ligament (long arrow). Coronal CT image (B) demonstrates crown-shaped calcium deposits (short arrow) around the odontoid process. Spine MRI shows inflammatory tissue (arrows) surrounding the odontoid dens characterized by high signal on sagittal fat-suppressed T2-weighted image (C), low signal on sagittal T1-weighted image (D), and enhancement on postcontrast sagittal T1-weighted image (E).

## Disclosure

The authors report no relevant disclosures. Go to [Neurology.org/N](http://Neurology.org/N) for full disclosures.

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## Appendix (continued)

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## Appendix (continued)

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## Appendix (continued)

Name	Location	Contribution
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## References

1. Bouvet JP, Parc JML, Michalski B, Benlahrache C, Auquier L. Acute neck pain due to calcifications surrounding the odontoid process: the crowned dens syndrome. *Arthritis Rheum.* 1985;28(12):1417-1420.
2. Ledingham D, Cappelen-Smith C, Cordato D. Crowned dens syndrome. *Pract Neurol.* 2018;18(1):57-59.

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