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## Teaching Neuroimage: Crowned Dens Syndrome, an Acute Attack of Calcium Pyrophosphate Deposition Disease Mimicking Acute Meningitis

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A 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 atlanto-axial joint with inflammatory changes revealed by cervical-spine-MRI (figure). Clinical presentation, biochemical and radiological 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.

**Table:** Laboratory data on admission and at discharge 10 days later

Variable	On admission	At discharge	Normal range
<b>Blood</b>			
White-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	–	
Anti-cyclic citrullinated peptide antibody (RU/ml)	Negative	–	
<b>Cerebrospinal fluid</b>			
Red-cell count (per $\mu l$ )	Absent	–	
White-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	–	

## References

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2. Ledingham D, Cappelen-Smith C, Cordato D. Crowned dens syndrome. *Pract Neurol*. 2018;18:57–59.

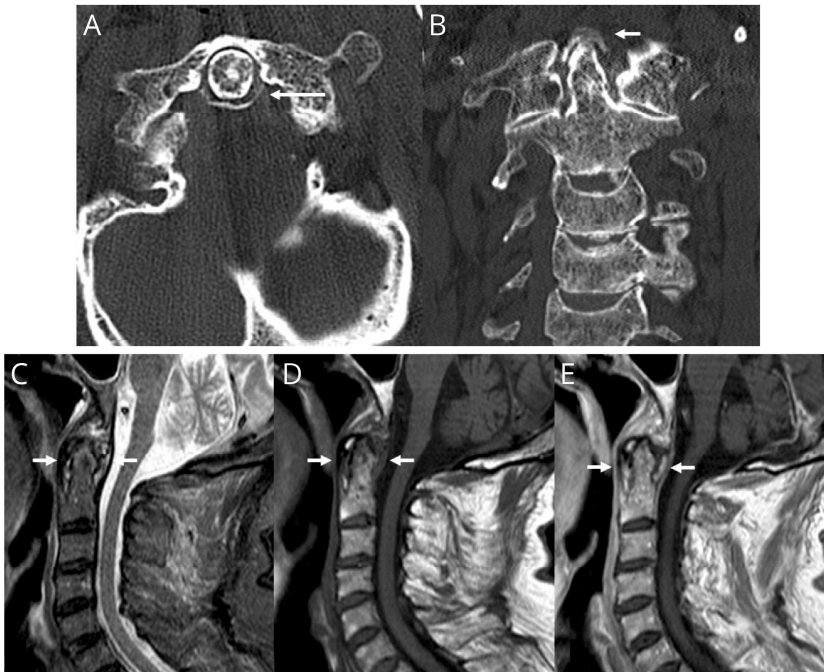
**Ethics approval:** the study was performed in accordance with the ethical standards statement.

**Informed consent:** the patient provided written informed consent.

## Figure

Title: Imaging Features of an Acute Attack of Calcium Pyrophosphate Deposition Disease (CPPD) in the Atlantoaxial Joint.

Legend: (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 (arrow) 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 post-contrast sagittal T1-weighted images (E).



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