

Teaching NeuroImage: Crowned Dens Syndrome

An Acute Attack of Calcium Pyrophosphate Deposition Disease Mimicking Acute Meningitis

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Neurology® 2022;99:442-444. doi:10.1212/WNL.0000000000200949

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Table Laboratory Data on Admission and at Discharge 10 Days Later

Variable	On admission	At discharge	Normal range
Blood			
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	—	
CSF			
Red blood cell count (per μL)	Absent	—	
White blood cell count (per μL)	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.^{1,2} 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.

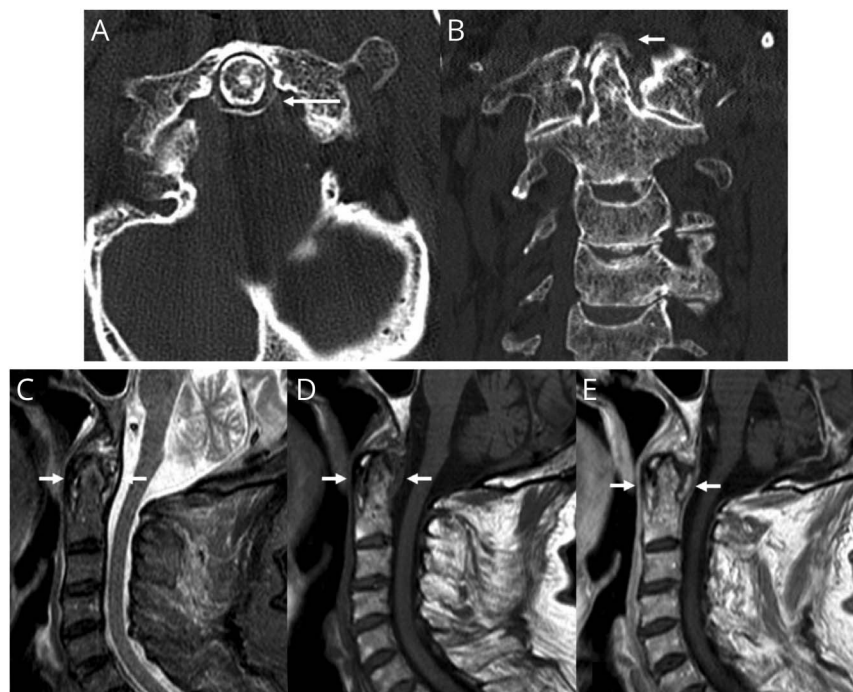
Study Funding

No targeted funding reported.

From the Department of Pathophysiology and Transplantation (A.F., G.C., M.C.S.), University of Milan; Neurodegenerative Diseases Unit (A.F., A.A., A.M.P., G.G.F., S.F., E.S., D.G., M.C.S., T.C.), and Neuroradiology Unit (G.C., G.P.), Fondazione IRCCS Ca' Granda, Ospedale Maggiore Policlinico, Milan; and Department of Biomedical (S.F., E.S., D.G.), Surgical and Dental Sciences, University of Milan, Italy.

Go to Neurology.org/N for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

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 for full disclosures.

Publication History

Received by *Neurology* March 8, 2022. Accepted in final form May 20, 2022. Submitted and externally peer reviewed. The handling editor was Roy Strowd III, MD, Med, MS.

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Alessandro Francia, MD	Department of Pathophysiology and Transplantation, University of Milan; Neurodegenerative Diseases Unit, Fondazione IRCCS Ca' Granda, Ospedale Maggiore Policlinico, Milan, Italy	Drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data
Giorgio Conte, MD	Department of Pathophysiology and Transplantation, University of Milan; Neuroradiology Unit, Fondazione IRCCS Ca' Granda, Ospedale Maggiore Policlinico, Milan, Italy	Drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; and analysis or interpretation of data

Appendix (continued)

Name	Location	Contribution
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Appendix (continued)

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Appendix (continued)

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Neurology 2022;99;442-444 Published Online before print July 5, 2022

DOI 10.1212/WNL.0000000000200949

This information is current as of July 5, 2022

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