

# Imaging of Atlantoaxial Calcium Pyrophosphate Deposition Disease With Unusual Onset as Drop Attack

Valerio D'Agostino<sup>1</sup>, MD, Università degli Studi di Napoli Federico II Scuola di Medicina e Chirurgia, Diagnostic and Interventional Radiology, Naples; Bianca Vanzi<sup>2</sup>, MD, Alma Mater Studiorum Università di Bologna, Diagnostic and Interventional Imaging, Bologna; Riccardo Ghermandi, MD, Paolo Spinnato<sup>3</sup>, MD, IRCCS Istituto Ortopedico Rizzoli, Bologna, Italy. Address correspondence to Dr. V. D'Agostino, Diagnostic and Interventional Radiology - IRCCS Istituto Ortopedico Rizzoli, Via G.C., Pupilli n. 1, 40136, Bologna, Italy. Email: valerio.dagostino123@gmail.com. The authors declare no conflicts of interest relevant to this article. Ethics approval from the authors' institutions was waived, according to the policy for case reports. Informed consent was obtained from the patient.

Calcium pyrophosphate deposition disease (CPPD) affects mostly appendicular joints.<sup>1</sup> Symptoms can be similar to those of gout, metastatic bone disease, and spinal tumors.<sup>2</sup> When it involves the first cervical vertebrae, CPPD can cause crowned dens syndrome; the classic presenting triad is headache, fever, and morning cervical pain. Dual-energy computed tomography (CT) scanning identifies erosive bone changes and composition of abnormal calcifications, thus considered the gold standard for imaging at the C1-C2 level.<sup>3</sup> Magnetic resonance imaging evaluates radiculopathy, cord compression, or cervical myelopathy.<sup>4</sup>

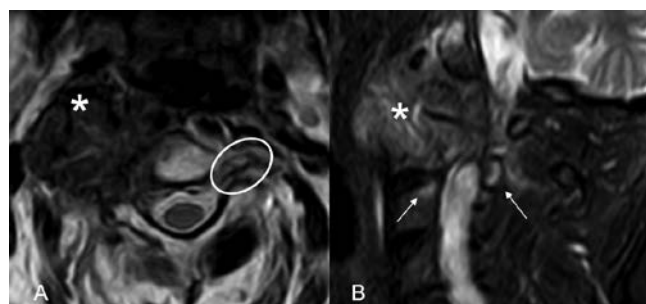
A 72-year-old woman without neurologic comorbidities suffered a drop attack, and complained of pain after right rotation of the neck. Laboratory tests showed hyperuricemia; other results were unremarkable. Cervical spine magnetic resonance imaging showed a voluminous mass occupying the right lateral

recess and intervertebral foramen, encroaching the C2-C3 right root nerve (Figure 1). The mass was characterized by low-signal intensity in both T1-weighted (W) and T2W sequences, with a mild hyperintensity on short-tau inversion recovery sequence, along with C2-C3 vertebral endplate edema. No myelopathy was detected. Given the size of the mass, a biopsy was performed. Pre-procedure CT scan revealed its calcific nature, polycyclic margins, bone erosive changes, and calcification of alar and transverse ligaments of the atlas and anterior atlanto-occipital membrane (Figure 2). Pathologists' polarized light revealed CPPD crystals.

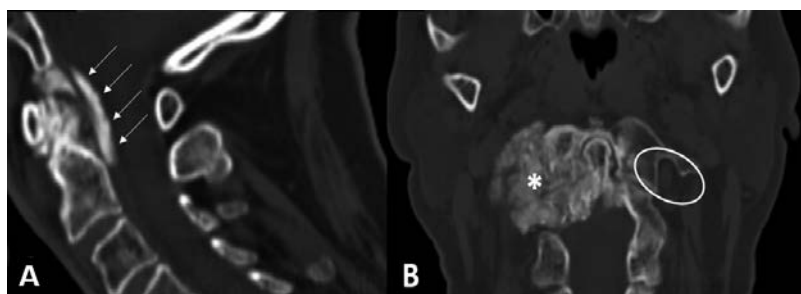
CPPD crystals can form a voluminous tumor-like mass that can compress vascular structure, as in our case, where the bulky mass probably provoked the drop attack, causing stenosis of right vertebral artery. This rare presentation could pose a challenging differential diagnosis with tumoral calcinosis, although the recognition of crowned dens appearance should strongly suggest CPPD.<sup>5</sup>

## REFERENCES

1. Masuda I, Ishikawa K. Clinical features of pseudogout attack. A survey of 50 cases. *Clin Orthop Relat Res* 1988;229:173-81.
2. Magid SK, Gray GE, Anand A. Spinal cord compression by tophi in a patient with chronic polyarthritis: case report and literature review. *Arthritis Rheum* 1981;24:1431-4.
3. Daniels DL, Williams AL, Haughton VM. Computed tomography of the articulations and ligaments at the occipito-atlantoaxial region. *Radiology* 1983;146:709-716.
4. Parker GD, Jones NR, Scott G. Cervical myelopathy in C.P.P.D. deposition disease. *Australas Radiol* 1990;34:82-5.
5. Sissons HA, Steiner GC, Bonar F, et al. Tumoral calcium pyrophosphate deposition disease. *Skeletal Radiol* 1989;18:79-87.



**Figure 1.** MRI sequences. (A) T2W DRIVE (Philips) on axial plane and (B) T2W FSE on sagittal plane. The bulky laterocervical mass (asterisk) shows a homogeneous hypointensity on T2W images. (A) T2W 3D DRIVE helped to assess the root-nerve compression in the conjugation foramen; the left is completely normal (white oval). (B) T2W highlights the bone edema on C3 vertebral superior endplate and on C2 posterior spinous process (white arrows). FSE: fast spin echo; T2W: T2-weighted.



**Figure 2.** CT axial and MPR of cervical spine. CT shows a bulky laterocervical mass (asterisk) with calcium-like density without bone erosions that occupies almost totally the C2-C3 conjugation foramen and the lateral recess. MPR-reconstructed (A) sagittal scan visualizes the massive calcification of the transverse ligament (white arrows); and (B) coronal scan depicts the involvement of C2-C3 intertransversarii right foramen and a probable obstruction of vessels in it, while the left is clear and vertebral artery is patent (white oval). CT: computed tomography; MPR: multiplanar reconstruction scan.