Acute Pain in the Neck: Don't Miss the Crown!

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Figure 1. Axial computed tomography (CT) of the atlantoaxial joint. There is curvilinear calcification of the transverse ligament of the atlas (arrow), consistent with crowned dens syndrome.

An 84-year-old woman presented with a one day history of severe and disabling occipital and neck pain. On admission, she was feverish, disoriented to place showing prominent neck rigidity. She was particularly agitated, whenever the range of her neck movements was examined. Otherwise, her neurological examination, including fundoscopy, was unremarkable. Her medical background included hypertension, diabetes mellitus type 2, and osteoporosis. Routine laboratory examination revealed heightened indices of systemic inflammation. Due to the combination of fever, neck stiffness, and altered mental status, we performed a lumbar puncture which showed no evidence of central nervous system infection.

Computed tomography of the cervical spine demonstrated extensive calcifications surrounding the atlantoaxial joint creating the appearance of an halo, or crown, posterior to the odontoid process (see Figure 1). Based on the clinical data and this imaging finding, crowned dens syndrome was

diagnosed and short-term treatment with naproxen (1000 mg/d) resulted in complete resolution of the patient's symptoms, sustained over a period of 6-month follow-up.

Crowned dens syndrome is a clinoradiological condition characterized by relapses of occipital pain and neck stiffness related to the composition of calcium pyrophosphate in the periodontoid ligaments between the dens and the atlas.^{1,2} Evidence of an ongoing inflammatory process in the form of fever and elevated inflammatory serum markers commonly coexists. This syndrome may perfectly masquerade as a meningoencephalitis, as was the case in our patient. Its prompt recognition is essential in order to avoid unnecessary diagnostic and therapeutic interventions in this context.^{1,2}

Authors' Note

The article has not been submitted for publication elsewhere. All authors have contributed substantively to the conception, design, or analysis and interpretation of the data, have contributed substantively to the drafting of the manuscript or critical revision for important intellectual content, have given final approval of the version to be published, and agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. On behalf of the authors, the corresponding author states that we acted in accordance with the ethical standards of the 1964 Declaration of Helsinki.

Declaration of Conflicting Interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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