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CROWNED DENS SYNDROME: REPORT OF THREE CASES AND A REVIEW OF THE LITERATURE

Akihiko Oka, MD, PHD,* Koichi Okazaki, MD,† Ayumu Takeno, MD,* Satoshi Kumanomido, MD,* Ryusaku Kusunoki, MD,* Shuichi Sato, MD, PHD,‡ Shunji Ishihara, MD, PHD,‡ Yoshikazu Kinoshita, MD, PHD,‡ and Masayoshi Nishina, MD, PHD*

*Department of Emergency and Critical Care Medicine, Faculty of Medicine, Shimane University, Enya-cho, Izumo-City, Shimane Prefecture, Japan, †Fourth Department of Internal Medicine, Faculty of Medicine, Shimane University, Enya-cho, Izumo-City, Shimane Prefecture, Japan, and ‡Second Department of Internal Medicine, Faculty of Medicine, Shimane University, Enya-cho, Izumo-City, Shimane Prefecture, Japan

Reprint Address: Akihiko Oka, MD, PHD, Department of Emergency and Critical Care Medicine, Faculty of Medicine, Shimane University, Enya-cho, Izumo-City, Shimane Prefecture 693-8501, Japan

□ **Abstract—Background: Patients with crowned dens syndrome (CDS), which is pseudogout of the atlantoaxial junction induced by "crown-like" calcifications around the dens, present with symptoms of severe neck pain, rigidity, and high fever. CDS patients are often misdiagnosed as having meningitis or polymyalgia rheumatica, leading to potentially unnecessary invasive procedures for diagnosis and treatment. Case Report: We report 3 patients with CDS who had characteristic findings on computed tomography (CT), all of whom quickly recovered with nonsteroidal anti-inflammatory drug (NSAID) administration. In addition, we reviewed 72 published cases, including our patients. CDS typically occurs in elderly people (mean age 71.4 years). Common symptoms include neck pain (100%), neck rigidity (98%), and fever (80.4%), and most show elevated inflammatory markers (88.3%) on serum laboratory tests. Neck pain on rotation is a characteristic and helpful symptom in the diagnosis. The most useful modality is CT (97.1%), showing linear calcium deposits around the dens, mostly in the transverse ligament of atlas (TLA). CT number is especially helpful to distinguish a normal TLA (35–110 HU) from a calcified one (202–258 HU) in our cases. The most effective treatment is NSAID administration (85%), which usually leads to marked resolution of symptoms within days or weeks. Why Should an Emergency Physician Be Aware of This?: Due to acute and severe symptoms, CDS patients often present to an emergency department. To avoid unnecessary invasive procedures for diagnosis and treatment, CDS should be considered in the differential diagnosis of febrile neck pain.** © **2015 Elsevier Inc.**

□ **Keywords—pseudogout; neck pain; calcification; computed tomography**

INTRODUCTION

Neck pain is a common reason for presentation to an emergency department (ED) and is experienced by up to 71% of people in their lifetimes (1). Crowned dens syndrome (CDS) is a rare cause of neck pain with fever due to pseudogout of the atlantoaxial junction induced by "crown-like" calcifications around the dens (2). Because the symptoms mimic meningitis, temporal arteritis, or polymyalgia rheumatica (PMR), many CDS patients have invasive procedures for diagnosis (lumbar puncture, temporal artery biopsy) and unnecessary treatments (antibiotics, steroids) (3–5). We report three cases of CDS with characteristic findings on computed tomography (CT), all of which quickly recovered with nonsteroidal antiinflammatory drug (NSAID) administration.

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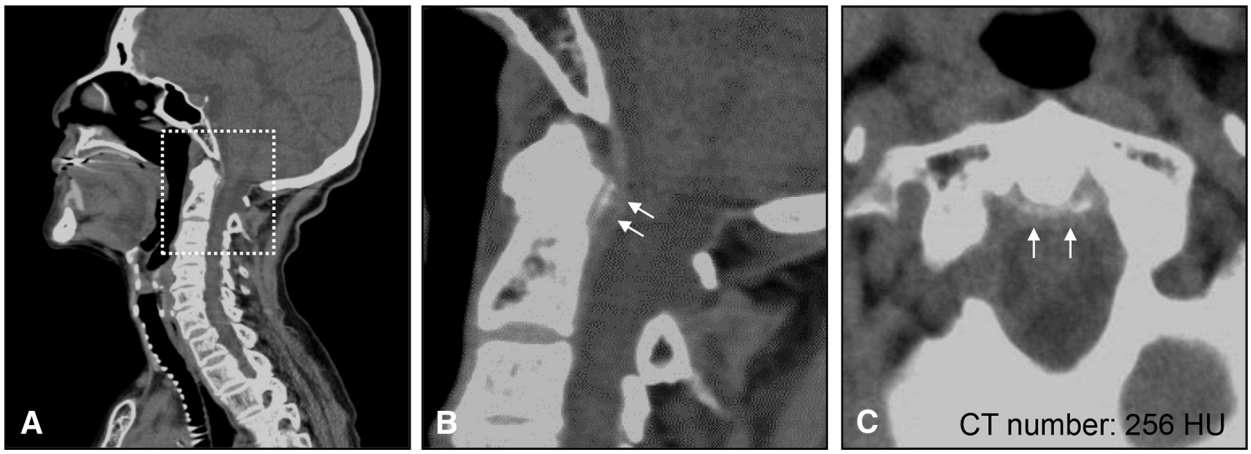

Figure 1. (Case 1) Computed tomography (CT) images of a 93-year-old woman with crowned dens syndrome (window level, 30 HU; window width, 250 HU). (A) Sagittal CT image showing calcifications in the transverse ligament of atlas (TLA). (B) Magnified view of the small square in (A). Image showing calcifications in the TLA (arrow). (C) Axial CT image showing calcifications surrounding the odontoid process (arrow). CT calcifications measure 256 HU (region of interest area: 1.8 mm^2).

CASE REPORT

Case 1

A 93-year-old woman presented to our ED with a 2-day history of high fever and severe neck pain radiating to the right shoulder. She presented 1 week previously to her primary care physician with pharyngalgia and a mild fever, and was treated with antibiotics without improvement. In our department, she had a fever (37.7°C) and severe neck rigidity. Laboratory findings showed an elevated C-reactive protein (CRP, 1.37 mg/dL; normal below 0.2). Our initial diagnosis was infectious meningitis, but cervical CT revealed linear calcifications around the dens (Figure 1), suggestive of CDS. We treated her with a suppository of diclofenac 25 mg, resulting in marked resolution of her neck pain and rigidity within 1 h.

Case 2

An 83-year-old woman presented with a 1-week history of pharyngalgia and a 2-day history of fever and neck pain. On examination, she had a high fever (38.5°C) and neck rigidity. Laboratory findings showed elevated CRP (16.49 mg/dL), and CT revealed linear calcifications posterior to the dens (Figure 2), resulting in a diagnosis of CDS. Administration of oral loxoprofen 60 mg led to resolution of her symptoms within 3 days.

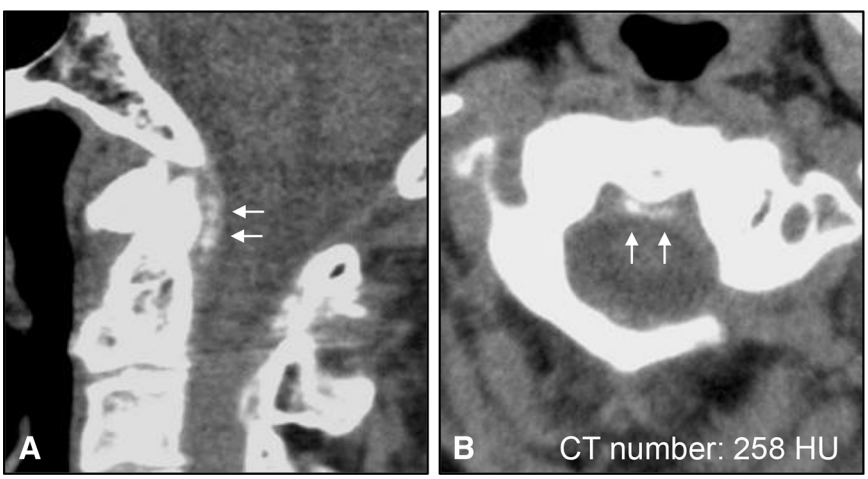

Figure 2. (Case 2) Computed tomography (CT) images of an 83-year-old woman with crowned dens syndrome (window level, 30 HU; window width, 250 HU). (A) Sagittal CT image showing calcifications in the transverse ligament of atlas. (B) Axial CT image showing calcifications surrounding the odontoid process (arrow). CT calcifications measure 258 HU (region of interest area: 1.8 mm^2).