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IMAGING HIGHLIGHT

A rare case of crowned dens syndrome masquerading as meningitis

[1]Dong-geon Lee *MD*, [1]Seung Won Choi *MD PhD*, [2]Sunggun Lee *MD PhD*, [3]Min Wook So *MD PhD*, [1]Doo-Ho Lim *MD PhD*

[1]Division of Rheumatology, Department of Internal Medicine, Ulsan University Hospital, University of Ulsan College of Medicine, Ulsan; [2]Division of Rheumatology, Department of Internal Medicine, Haeundae Paik Hospital, Inje University College of Medicine, Busan; [3]Division of Rheumatology, Department of Internal Medicine, Pusan National University Yangsan Hospital, Yangsan, South Korea

INTRODUCTION

Crowned dens syndrome (CDS) is a clinicoradiographic entity defined by an acute onset of cervico-occipital pain, fever, nuchal rigidity, and radiological calcifications of the cruciform ligament around the odontoid process.[1] These calcium deposits, mostly calcium pyrophosphate dihydrate (CPPD) crystals or hydroxyapatite crystals, can remain asymptomatic or be responsible for clinical manifestations such as chronic cervical pain or spinal cord compression.[2] CDS is a rare condition; however, it is frequently misdiagnosed.[3] Here, we report a rare case of CDS masquerading as meningitis to highlight CDS as an important differential diagnosis in patients presenting with acute neck pain and fever.

CASE REPORT

An 87-year-old woman visited our hospital complaining of a 4-day history of non-traumatic severe occipital headache and low-grade fever of 37.8°C. She had a history of hypertension, for which she was taking amlodipine 5 mg daily. In addition, she had been diagnosed with cancer of the left breast 4 years ago, which had been in complete remission for 3.5 years after left mastectomy and adjuvant chemotherapy were performed. She was a never-smoker and social drinker; however, she had stopped drinking 4 years ago.

On admission, her blood pressure was 153/85 mmHg, heart rate was 95 beats/min, body temperature was 37.8°C, and respiratory rate was 20 breaths/min. During her physical examination, neck palpation revealed mild tenderness; however, she complained of severe pain and neck stiffness on rotation in any direction, resulting in limited range of neck motion. Initial routine laboratory tests revealed markedly elevated white blood cell count (12,120/mm^3), erythrocyte sedimentation rate (87 mm/h), and C-reactive protein level (13.91 mg/dL), but not serum uric acid level (3.5 mg/dL) (Table 1). Blood culture showed negative results, and cerebrospinal fluid (CSF) analysis revealed a slightly elevated white blood cell count of 5/μL and protein level of 60.4 mg/dL, whereas CSF gram stain/culture, acid-fast bacilli stain/culture, anti-herpes simplex virus, and varicella-zoster virus antibodies were all negative. Gadolinium-enhanced brain magnetic resonance imaging (MRI) showed neither abnormal enhancing nor metastatic lesion of breast cancer in the brain or meninges.

The patient was initially diagnosed with meningitis and treated with antibiotic agents (acyclovir 10 mg/kg intravenously thrice daily, ampicillin 2 g intravenously six times a day, ceftriaxone 2 g intravenously twice a day, and vancomycin 15 mg/kg intravenously twice a day) and acetaminophen 650 mg twice a day for 3 days. However, her symptoms failed to improve. Additional computed tomography (CT) of the neck revealed a remarkable crown-like calcification surrounding the odontoid process (Figure 1). Gadolinium-enhanced cervical MRI showed a calcified mass with low signal intensity on T2-weighted images and diffuse enhancement around the odontoid process, suggesting inflammatory changes (Figure 2). On the basis of the clinical and imaging findings, a diagnosis of CDS was established.

The patient was treated with a non-steroidal

Address correspondence to: Doo-Ho Lim, MD, PhD, Division of Rheumatology, Department of Internal Medicine, Ulsan University Hospital, University of Ulsan College of Medicine 877 Bangeojinsunhwan-doro, Dong-gu, Ulsan, South Korea 44033. Tel; +82-52-250-8970; e-mail:dlaengh@hanmail.net

Table 1: Initial routine laboratory and cerebrospinal fluid data

	Value	Normal range
Blood sample		
White blood cell (/mm^3)	12,120	4,000–10,000
Hemoglobin (g/dL)	12.6	14–18
Platelet (/mm^3)	345,000	140,000–400,000
Erythrocyte sedimentation rate (mm/h)	87	0–20
Total bilirubin (mg/dL)	0.5	0.1–1.2
Aspartate aminotransferase (IU/L)	19	0–40
Alanine aminotransferase (IU/L)	12	0–40
Alkaline phosphatase (IU/L)	58	25–100
Total protein (g/dL)	7.0	6.4–8.3
Albumin (g/dL)	4.1	3.5–5.0
Blood urea nitrogen (mg/dL)	12.8	8–20
Creatinine (mg/dL)	0.76	0.6–1.5
Uric acid (mg/dL)	3.5	2.1–7.4
Glucose (mg/dL)	110	70–110
C-reactive protein (mg/dL)	13.91	0–0.5
Cerebrospinal fluid		
White blood cell (/mm^3)	5	0–3
Red blood cell (/mm^3)	3	0–10
Protein (mg/dL)	60.4	15–45
Glucose (mg/dL)	73	50–80
Lactate dehydrogenase (IU/L)	12	8–50
Adenosine deaminase (IU/L)	0.5	0–8

anti-inflammatory drug (NSAID) (celecoxib 200 mg twice a day) and a low-dose steroid (prednisolone 5 mg twice a day). Her symptoms and laboratory data dramatically improved within 4 days. At 5 months after discharge, she was free of symptoms, with no evidence of recurrence.

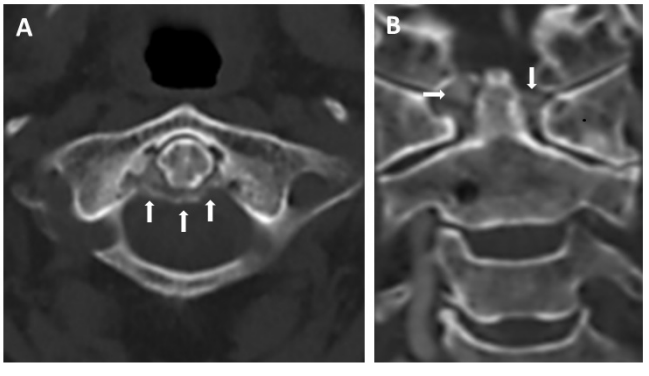

Figure 1. Computed tomography of the neck shows curvilinear calcifications of the transverse ligament of the atlas (Panel A, arrows) and crown-shaped calcium deposits surrounding the odontoid process (Panel B, arrows).

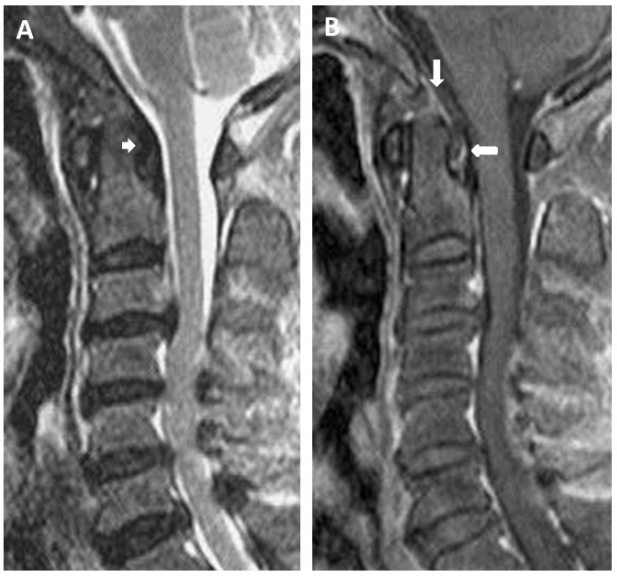

Figure 2. Magnetic resonance imaging findings around the odontoid process. (A) T2-weighted sagittal image shows a low-signal intensity calcified mass behind the dens (arrow). (B) T1-weighted fat suppression image with gadolinium-enhancement reveal diffuse enhancement around the odontoid process (arrows), suggesting inflammatory change.

DISCUSSION

CDS, first described and presented by Bouvet *et al*. in 1985[4], is defined as a combination of acute cervical pain with fever and microcrystalline deposition surrounding the top and sides of the odontoid process in a crown- or halo-like distribution. The microcrystalline deposition mostly consists of CPPD crystals, and typically, CPPD crystal deposition disease manifests as acute episodic mono-/oligo-arthritis involving a large joint (including the knees, wrists, and ankles), namely "pseudogout" or chronic arthropathy manifesting as mild joint pain and stiffness of the knees, wrists, elbows, and shoulders.[5] Similar to CPPD deposition around the peripheral joint, CPPD deposition around the odontoid process shows variable clinical manifestations from an asymptomatic state to chronic cervical pain.[2]

In clinical practice, the diagnosis of CDS is often difficult, owing to its atypical symptoms including temporal headache, jaw claudication, shoulder pain, and radiculomyelopathy. Moreover, the acute presentation of neck pain, headache, and fever in CDS frequently warrants a search for infectious or other inflammatory conditions such as abscess, meningitis, cervical spondylitis, metastatic tumors, giant cell arteritis, or polymyalgia rheumatic.[1] This could entail invasive procedures including lumbar punctures, temporal artery biopsies, and even surgical exploration of high cervical spine tissues before the correct diagnosis is established.[6-8] Therefore, CDS was considered as a differential diagnosis for the possible etiology of fever, headache, and spontaneous neck pain. In the present case, a lumbar puncture was performed, and the results were ambiguous, as observed in several previous studies[9,10]; hence, meningitis (slight elevation of white blood cell count and protein in CSF) could not be excluded. Brain MRI, performed to exclude the metastasis of possibly recurrent breast cancer, did not reveal any metastatic findings.

CT focusing on C1 and C2 is the gold standard imaging modality for CDS, because it is the technique of choice for detection of CPPD crystal deposition adjacent to the odontoid process.[3] While the typical, tiny, half-ringed, crown-like form of calcification behind the odontoid process is described as the most important feature of CDS, CPPD deposition may be observed posterior (50%), posterolateral (27.5%), circular (12.5%), anterior (5%), or lateral (5%) to the odontoid process.[11] Another useful aspect of CT is its ability to rule out the differential diagnoses of CDS, such as unrecognized odontoid fracture. Periodontoid calcification is not evident on plain radiographs in most patients.[11] Cervical MRI is generally unhelpful in the diagnosis of CDS; however, it may help exclude important differential diagnoses such as infectious spondylitis, discitis, myelopathy, and rare cases of malignancy.[12]

NSAIDs are the first-line treatment for CDS. NSAID treatment often brings about an improvement of symptoms and inflammatory markers. Corticosteroids, alone or in combination with NSAIDs, are also effective. In refractory cases, colchicine has been effectively used.[1-3] Frey *et al*.[13] reported that a patient with CDS showed incomplete pain improvement with both NSAIDs and systemic corticosteroids but went into remission with corticosteroid injection at C1-C2. Our patient was treated with an NSAIDs and a corticosteroid, and her symptoms and laboratory data dramatically improved within 4 days.

CDS is thought to have a good prognosis, with acute relapses treated episodically, resulting in symptomatic relief.[3] To date, there have been few studies on the natural course or potential factors responsible for re-attacks or neurological compromise. However, several cases have been reported of histologically proven massive periodontoid CPPD lesions causing atlantoaxial instability, spinal cord compression, or myelopathy requiring cervical decompression.[14,15]

In conclusion, CDS, also known as acute pseudogout of the neck, is a rare cause of cervical pain and is characterized by microcrystalline deposition. CDS is clinically accompanied by severe neck pain and stiffness, often in conjunction with pyrexia and elevated inflammatory markers. Hence, it is often misdiagnosed as meningitis, as in our case. This case suggests that CDS should be considered as the differential diagnosis for the possible etiology of fever, headache, and spontaneous neck pain of unknown origin.

Keywords: Crowned dens syndrome, pseudogout, meningitis

DISCLOSURE

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REFERENCES

1. Aouba A, Vuillemin-Bodaghi V, Mutschler C, De Bandt M. Crowned dens syndrome misdiagnosed as polymyalgia rheumatica, giant cell arteritis, meningitis or spondylitis: an analysis of eight cases. *Rheumatology (Oxford)* 2004; 43:1508-12.
2. Constantin A, Bouteiller G. Acute neck pain and fever as the first manifestation of chondrocalcinosis with calcification of the transverse ligament of the atlas. Five case-reports with a literature review. *Rev Rhum Engl Ed* 1998; 65: 583-5.
3. Siau K, Lee M, Laversuch CJ. Acute pseudogout of the neck--the crowned dens syndrome: 2 case reports and review of the literature. *Rheumatol Int* 2011; 31: 85-8.
4. Bouvet JP, le Parc JM, Michalski B, Benlahrache C, Auquier L. Acute neck pain due to calcifications surrounding the odontoid process: the crowned dens syndrome. *Arthritis Rheum* 1985; 28: 1417-20.
5. Wu DW, Reginato AJ, Torriani M, Robinson DR, Reginato AM. The crowned dens syndrome as a cause of neck pain: report of two new cases and review of the literature. *Arthritis Rheum* 2005; 53: 133-7.
6. Ishikawa K, Furuya T, Noda K, Okuma Y. Crowned dens syndrome mimicking meningitis. *Intern Med* 2010; 49: 2023.
7. Sekijima Y, Yoshida T, Ikeda S. CPPD crystal deposition disease of the cervical spine: a common cause of acute neck pain encountered in the neurology department. *J Neurol Sci* 2010; 296: 79-82.
8. Ring D, Vaccaro AR, Scuderi G, Pathria MN, Garfin SR. Acute calcific retropharyngeal tendinitis. Clinical presentation and pathological characterization. *J Bone Joint Surg Am* 1994; 76: 1636-42.
9. Inoue A, Kohno K, Ninomiya S, *et al*. Usefulness of cervical computed tomography and magnetic resonance imaging for rapid diagnosis of crowned dens syndrome: A case report and review of the literature. *Int J Surg Case Rep* 2017; 30: 50-4.
10. Takahashi T, Minakata Y, Tamura M, Takasu T, Murakami M. A rare case of crowned dens syndrome mimicking aseptic meningitis. *Case Rep Neurol* 2013; 5: 40-6.
11. Goto S, Umehara J, Aizawa T, Kokubun S. Crowned dens syndrome. *J Bone Joint Surg Am* 2007; 89: 2732-6.
12. Scutellari PN, Galeotti R, Leprotti S, Ridolfi M, Franciosi R, Antinolfi G. The crowned dens syndrome. Evaluation with CT imaging. *Radiol Med* 2007; 112: 195-207.
13. Frey ME, Dery FJ, Jr., Cifu DX. C1-2 steroid injection for crowned dens syndrome. *PM R* 2009; 1: 379-82.
14. Doita M, Shimomura T, Maeno K, Nishida K, Fujioka H, Kurosaka M. Calcium pyrophosphate dihydrate deposition in the transverse ligament of the atlas: an unusual cause of cervical myelopathy. *Skeletal Radiol* 2007; 36: 699-702.
15. Sethi KS, Garg A, Sharma MC, Ahmad FU, Sharma BS. Cervicomedullary compression secondary to massive calcium pyrophosphate crystal deposition in the atlantoaxial joint with intradural extension and vertebral artery encasement. *Surg Neurol* 2007; 67: 200-3.