Crowned Dens Syndrome Masquerading as Meningitis

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Abstract

The classic symptoms of meningismus, including fever, neck stiffness, and headache, should automatically trigger a prime differential of meningitis, but a close masquerader, albeit rare, is crowned dens syndrome. Herein, we report the case of a 71-year-old woman with clinical features of meningismus with elevated inflammatory biomarkers. However, computed tomography of the cervical spine revealed the presence of calcium deposits encircling the dens. Hence, an alternate diagnosis of crowned dens syndrome was considered. This was confirmed by the presence of similar pathology in other joints and the dramatic resolution of symptoms and inflammatory markers with the administration of nonsteroidal anti-inflammatory drugs.

Introduction

Crowned dens syndrome (CDS) is a clinico-radiological syndrome characterized by the presence of calcification in the ligament encircling the odontoid process. It presents with a variable duration of fever, meningism, and an inflammatory syndrome [1]. These micromcrystalline deposits most often consist of calcium pyrophosphate dehydrate (CPPD) crystals or hydroxyapatite crystals. Patients can be either asymptomatic or symptomatic, presenting with a clinical picture similar to meningitis. CDS is commoner in the elderly as the mean age at the time of onset was found to be seventy-three years in men and sixty-two years in women, according to Goto et al. [2]. Herein, we report the case of an older woman with osteoarthritis (OA) who presented with acute attacks of CDS. We have attempted to examine the complex relationship between OA and the pathogenesis of CDS.

Case Presentation

A 71-year-old woman with a history significant for systemic hypertension, hyperlipidemia, and OA presented with fever, flu-like symptoms, and arthralgia for two weeks. While cough and runny-nose resolved, she continued to have persistent fever, malaise, and weakness. These were severe enough to cause a partial limitation in her activities of daily living. She also noticed pain in her left shoulder, left knee, mid-back, and neck that was associated with neck stiffness, which made her preferentially turn her head to the right side. The patient denied experiencing headaches, photophobia, phonophobia, nausea, vomiting, visual impairment, and upper or lower extremity weakness. There was no history of intravenous drug use, recent travel, coronavirus disease 2019 (COVID-19) contact, or surgery.

On physical examination, she was alert and oriented to place, time, and person. Her heart rate was regular but fluctuated between 95 and 110 beats/min, and her blood pressure was 200/109 mmHg. A temperature of 101.7 F was recorded. Neurologic examination was unremarkable, except for mild neck stiffness when the neck was passively rotated from side to side, which was worse on the left side. Kernig and Brudzinski’s signs were negative. She was unable to raise her left upper and lower limbs because of severe aching pain in her left shoulder and knee, but the power in the right upper and lower extremities was 5/5. She noticed pain in her left shoulder, left knee, mid-back, and neck that was associated with neck stiffness, which made her preferentially turn her head to the right side. The patient denied experiencing headaches, photophobia, phonophobia, nausea, vomiting, visual impairment, and upper or lower extremity weakness. There was no history of intravenous drug use, recent travel, coronavirus disease 2019 (COVID-19) contact, or surgery.

Blood tests revealed leukocytosis (23.75 × 10^9 cells/L); mildly reduced hemoglobin level (11.3 g/L); and elevated erythrocyte sedimentation rate (ESR: 125 mm/h), C-reactive protein (CRP: 480.66 ng/dL), and rheumatoid factor (17 IU/mL). The cyclic citrullinated peptide, thyroid-stimulating hormone, and anti-nuclear antibody levels were within the normal range. She had normal serum creatinine, low potassium, and low magnesium. She had elevated liver enzymes: alkaline phosphatase (257 IU/L), alanine transaminase (39 IU/L), and aspartate transaminase (40 IU/L). A rapid COVID-19 polymerase chain reaction test was negative. Creatinine kinase levels were normal, and blood cultures were negative. Computed tomography (CT) of the cervical spine (C-spine) showed calcification of the alar and transverse ligaments (Figures 1 and 2). However, brain CT revealed no acute intracranial abnormality. Hence, magnetic resonance imaging of the brain was performed, which revealed no acute intracranial hemorrhage, infarction, or abnormal intracranial contrast enhancement. A left knee joint radiograph showed chondrocalcinosis of the knee joint (Figure 3).
CT of the lumbosacral spine showed left sacroiliitis. Urinalysis did not show any evidence of infection, likewise the chest radiograph.

**FIGURE 1:** Cervical spine CT scan showing calcifications of the transverse and alar ligament (crowned dens syndrome)
FIGURE 2: Cervical spine CT scan showing calcifications of the alar ligament (crowned dens syndrome)

FIGURE 3: X-ray of the left knee joint showing chondrocalcinosis on the
inflammatory markers within four days of commencement of NSAIDs. These patients respond dramatically to NSAIDs, systemic corticosteroid treatment, or colchicine treatment. Although no medication exists that causes lysis and removal of calcium pyrophosphate (CPP) crystals from the joints in patients with CDS, these patients respond dramatically to NSAIDs, systemic corticosteroid treatment, or colchicine treatment. Although no medication exists that causes lysis and removal of calcium pyrophosphate (CPP) crystals from the joints in patients with CDS, these patients respond dramatically to NSAIDs, systemic corticosteroid treatment, or colchicine treatment. Although no medication exists that causes lysis and removal of calcium pyrophosphate (CPP) crystals from the joints in patients with CDS, these patients respond dramatically to NSAIDs, systemic corticosteroid treatment, or colchicine treatment. 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Conclusions
A high index of suspicion, a thorough clinical examination, a functionally precise analysis of the CT of the C1-2 spine, and adjunctive X-ray images of the large joints in patients with severely elevated inflammatory markers and meningismus may obviate certain investigations and rule out differential diagnoses. Given that calcium deposits in the neck joints can also occur in other joints of the body, it may be necessary to look at the X-ray of large joints to further support the diagnosis of CDS. The resolution of these features with nonsteroidal antiinflammatory drugs confirmed the diagnosis of CDS.

Additional Information

Disclosures

Human subjects: Consent was obtained by all participants in this study. Conflicts of interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following: Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work. Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. Other relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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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. 2004, 43:1508-1512. 10.1093/rheumatology/keh370
2. Goto S, Umezawa J, Alzaara T, Kokubun S: Crowned dens syndrome. J Bone Joint Surg Am. 2007, 89:2732-2736. 10.2106/JBJS.F.01322
3. Oka A, Okazaki K, Takeno A, et al.: Crowned dens syndrome: report of three cases and a review of the literature. J Emerg Med. 2015, 49:9-15. 10.1016/j.jemermed.2015.02.005
4. Bouvet JP, le Parc JM, Michalski B, Benlahrache C, Asquier L: Acute neck pain due to calcifications surrounding the odontoid process: the crowned dens syndrome. Arthritis Rheum. 1985, 28:1417-1420. 10.1002/art.1780281215
5. Godfrin-Valnet M, Godfrin G, Godard J, et al.: Eighteen cases of crowned dens syndrome: presentation and diagnosis. Neurochirurgie. 2013, 59:115-120. 10.1016/j.neuchi.2013.03.003
6. Abhishek A: Calcium pyrophosphate deposition disease - beyond gout. Nat Rev Rheumatol. 2018, 14:592-602. 10.1038/s41584-018-0007-5
7. McCarthy GM, Dunne A: Calcium crystal deposition diseases - beyond gout. Nat Rev Rheumatol. 2018, 14:592-602. 10.1038/s41584-018-0007-5
8. Malak A, Kanneganti TD: Inflammasome activation and assembly at a glance. J Cell Sci. 2017, 150:3955-3963. 10.1242/jcs.207365
9. Le Goff P, Pennec Y, Youinou P: Articular chondrocalcinosis revealed by acute cervical symptoms simulating meningitis (author's transl). Sem Hop. 1980, 56:1515-1518.
10. Uhm M, Dewar C, Spouge D, Blocka K: Crowned dens syndrome: a rare cause of acute neck pain. Clin Rheumatol. 2013, 32:711-714. 10.1007/s10067-013-2179-5
11. François S, Guaydier-Souquieres G, Marcelli C: Acute sacroiliitis as a manifestation of calcium pyrophosphate dihydrate crystal deposition disease. A report of two cases. Rev Rhum Engl Ed. 1997, 64:508-512.
12. Ottaviani S, Brunier L, Sibilia J, et al.: Efficacy of anakinra in calcium pyrophosphate crystal-induced arthritis: a report of 16 cases and review of the literature. Joint Bone Spine. 2013, 80:178-182. 10.1016/j.jbspin.2012.07.018