CLINICAL VIGNETTE

Crowned Dens Syndrome: An Atypical Cause of Neck Pain

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Case

A 75-year-old female presented to the emergency department with severe neck, upper back, and bilateral shoulder pain. Her symptoms started suddenly a few days prior. She described the pain as a shooting spasm with an associated burning sensation extending to her scalp. Her strength and mobility were limited due to the pain, which made it difficult to brush her hair or turn her head from side to side. The review of systems was otherwise negative, including absence of fevers, rashes, vision changes, or temporal pain. About five weeks prior, she developed pain and swelling of her right elbow at the medial epicondyle. She presented to urgent care and was diagnosed with pseudogout based on imaging. An arthrocentesis was not done. Her elbow pain and swelling resolved spontaneously. Her past medical history included chronic myeloid leukemia, gastroesophageal reflux disease, irritable bowel syndrome, hyperlipidemia, right hip chondrocalcinosis and osteopenia with a prior hip fracture. Past surgical history and family history were noncontributory. Her medications included dasatinib, atorvastatin, and cholecalciferol. She has a nonsteroidal anti-inflammatory drug (NSAID) allergy. The patient is a former smoker, with no significant alcohol or illicit drug use.

At presentation, vital signs were remarkable for only temperature was 99.5 °F. There was marked tenderness to palpation in the painful areas. There was no skin rash. Neurologic exam was limited due to severe pain with movement and she was unable to rotate her head. Admission labs were remarkable for C-reactive protein (CRP) 2.5 (<0.8 mg/dL) and erythrocyte sedimentation rate (ESR) 80 (<= 25 mm/hr). Both of these tests were normal the week prior. There was no leukocytosis or electrolyte abnormalities. She was started on acetaminophen, gabapentin, lidocaine patch, methocarbamol, and tramadol as needed. There was a lower suspicion for giant cell arteritis or meningitis given the lack of headache, vision changes, fever, photophobia, or phonophobia. Cervical spine radiographs showed severe spondylosis, worse than prior. X-rays of both shoulders had findings compatible with rotator cuff pathology. The patient underwent magnetic resonance imaging (MRI) of the cervical spine with and without contrast which showed that the degenerative changes of the cervical spine had only mildly progressed compared to years prior.

Her symptoms did not improve on hospital day 1 after starting multiple analgesic medications. Rheumatology was consulted given concern for polymyalgia rheumatica. Colchicine 0.6mg twice daily was started while awaiting a computed tomography (CT) scan given suspicion for crowned dens syndrome. CT scan of the neck with contrast revealed calcification around the odontoid process (Figure 1). Additional labs to evaluate specific rheumatologic disorders all negative. On hospital day 2, CRP and ESR peaked at 10.1 and 87, respectively. She was discharged on hospital day 3 given the improvement in pain control, and the decreasing inflammatory markers. Her symptoms had nearly resolved at follow-up eight days later. Inflammatory markers, parathyroid hormone, and Vitamin D levels were normal when checked one month later.

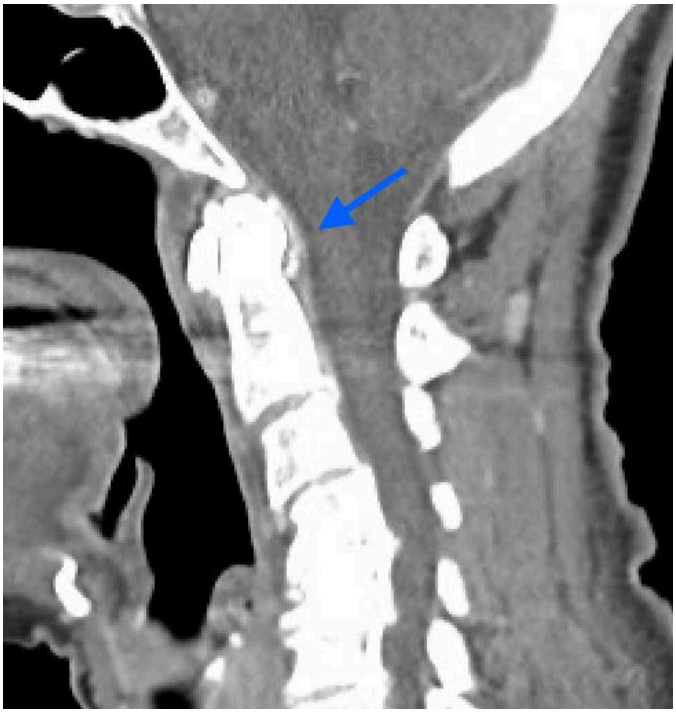

Figure 1.

Introduction

Crowned dens syndrome (CDS) is characterized by acute neck pain with radiographic findings of calcium deposition in the ligamentous structures around the odontoid process, also known as the dens. This can have the appearance of a crown on imaging. Although it can be due to deposition of calcium hydroxyapatite or calcium pyrophosphate, the clinical syndrome is more commonly associated with calcium pyrophosphate.[1] It is considered a less common presentation of calcium pyrophos-

phate deposition disease (CPPD), also known as pseudogout. Hypomagnesemia, hypophosphatasia, hyperparathyroidism, and hemochromatosis are metabolic conditions that are risk factors for CPPD.[2]

Diagnosis / Epidemiology

CDS is thought to be underdiagnosed although it is a rare disease. Diagnosis is based on the clinical presentation of acute neck pain and stiffness with restricted cervical range of motion, elevated inflammatory markers, calcification around the odontoid process on a CT scan, as well as exclusion of other causes. The differential diagnosis for CDS includes bacterial meningitis, polymyalgia rheumatica, giant cell arteritis, and cervical spondylosis.[3] CDS is most common in adults over age 65.[2] Isono *et al*, reviewed 72 cases of CDS in Japan and found a slight female predominance. This study reported patients with CDS had a higher rate of cervical rotation restriction versus flexion or extension limitation.[4] Clinical manifestations may include fever. Typical lab abnormalities include elevated ESR and CRP.[1] A CT scan is superior to plain radiographs and MRI for making the diagnosis.[2] One case report on crowned dens syndrome by Inoue *et al* noted hyperintensity of the soft tissue around the dens on a T2-weighted MRI with fat suppression.[5] However, MRI is not considered the gold standard for diagnosis. One important diagnostic consideration is asymptomatic calcification, incidentally found on imaging performed for other reasons. Chang *et al* examined the frequency of CPPD on CT scans done on 513 patients presenting with an acute trauma. The general prevalence of atlantoaxial CPPD was 12.5% in general. It increased to 34% in patients > 60 years and 49% in patients > 80 years.[6] Given the possibility of asymptomatic calcification, infectious causes should not be ruled out based on isolated imaging findings.[7]

Treatment / Prognosis

Although the incidence and prevalence of CDS are likely underestimated, it remains a rare presentation of CPPD. Common clinical symptoms include acute neck pain and stiffness with a median time of 8 days between symptom onset and resolution depending on the time of diagnosis after initial presentation.[4] However, there is a case report of a patient diagnosed with CDS after presenting with quadriparesis. The patient was found to have a mass with mineralization consistent with CPPD which caused cervicomedullary compression and required surgery.[8] Increased awareness of this diagnosis may avoid unnecessary testing and treatments such as lumbar puncture, temporal artery biopsy, and prolonged courses of steroids. At the same time, the diagnosis should not be made on imaging alone as there can be asymptomatic calcification. Treatment involves NSAIDs, steroids, or colchicine.[9] The specific medication and duration of treatment can vary depending on the patient's risk factors and response to treatment.

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