

CASE REPORT

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What a pain in the neck: Crowned dens syndrome as an uncommon cause of neck pain

Alexander M Pandelidis, Alan W Dow

ABSTRACT

Introduction: Crowned Dens Syndrome is an uncommon manifestation of calcium pyrophosphate deposition disease caused by crystal deposition and associated inflammation in the C1 and C2 area of the spine. Crowded Dens Syndrome typically causes posterior neck pain and limitation in range of motion. This report describes a case of this unusual and challenging diagnosis and its importance in the differential diagnosis of neck pain. **Case Report:** An 80-year-old woman presented as a transfer from another hospital for evaluation for possible epidural abscess. She noted several weeks of progressive posterior cervical neck pain. The pain was worse with passive motion, especially flexion, and she had decreased range of motion in all directions. She had elevated inflammatory markers, and neck computed tomography (CT) scan was non-diagnostic. Broad-spectrum antibiotics for treatment of a possible abscess or meningitis were initiated without improvement. After arrival at our hospital, a dual-energy CT scan concentrated at the C1–C2 area was performed. This imaging revealed multiple calcific deposits surrounding the odontoid with associated soft tissue thickening consistent with Crowned Dens Syndrome due to calcium pyrophosphate

deposition disease. She improved with steroid treatment. **Conclusion:** Crowned Dens Syndrome presents similarly to infectious, malignant, and other inflammatory causes of neck pain. Because specific imaging is often necessary to make the diagnosis, clinical awareness and suspicion are essential for identifying this condition and prescribing the correct therapy.

Keywords: Calcium pyrophosphate deposition disease, Crowned dens, Neck pain

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INTRODUCTION

Subacute neck pain with elevated inflammatory markers has a large differential with several life-threatening considerations including malignancy, infection, and inflammatory causes [1]. Diagnosis can prove challenging, but a correct diagnosis is essential because of the severe and progressive nature of some of the causes and the wide variation in treatment approaches indicated depending on the cause. We report a case of subacute neck pain which had initially been treated as a possible epidural abscess. Specific CT imaging revealed Crowned Dens Syndrome, an uncommon manifestation of calcium pyrophosphate deposition disease (CPPD). In one study, 15.9% of patients, not necessarily with CPPD, who underwent CT scans had evidence of this condition in the dens area, though only 12.5% of these patients

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were symptomatic [2]. This suggests that Crowned Dens Syndrome may be more common than suspected. Regardless, this condition should be on every clinician's differential for subacute or chronic neck pain with elevated inflammatory markers so that the appropriate treatment can be initiated.

CASE REPORT

An 80-year-old woman was transferred from an outside hospital for evaluation of persistent posterior neck pain which had not improved with initial management. On our initial evaluation, she described about two weeks of increasing posterior and lateral neck pain which worsened with any head movement. She was unable to find anything that relieved the pain and felt most comfortable with her neck fully flexed. She noted a mild posterior headache in addition to the neck pain. She denied fever, blurry vision, photophobia, cough or cold symptoms, dyspnea, abdominal pain, diarrhea, or dysuria.

Her past medical history included gastric cancer, originally treated with partial gastrectomy that was now metastatic to lungs, spleen, and liver. In addition, her medical history included end stage renal disease, hypertension, diabetes, hypothyroidism, and gastroesophageal reflux.

At the outside hospital, a CT scan showed diffuse prevertebral inflammation most prominent at C1–C4 with abnormal edema and enhancement in the odontoid process and the anterior arch of C1. Due to concern for a possible epidural abscess, she was started on antibiotics. Symptoms did not improve and a repeat CT was unchanged. She was transferred to our facility for further evaluation after four days of treatment.

On presentation to our facility, her temperature was 36.9°C, heart rate 105 bpm, blood pressure 140/78, and respiratory rate 16 bpm with an oxygen saturation of 95% on room air. The patient was pleasant, conversant, and in no acute distress, but she maintained her neck in flexion during the interview. Her cranial nerves were intact, and she was alert and oriented to person, place, and time. She had 5/5 strength and intact sensation bilaterally in the upper and lower extremities. She had no pain when moving her upper or lower extremities. On examination of the neck, there was no erythema or edema noted. She was moderately tender to palpation on the posterior and lateral portions of her neck. Her active range of motion was limited with rotation, flexion, and extension. There was no nuchal rigidity.

She had an elevated white blood cell count of 12.7×10^9 cells/L (normal range, 3.9×10^9 – 11.7×10^9 cell/L), a normal erythrocyte sedimentation rate of 14 mm in one hour (normal range, 0–20 mm in one hour), and an elevated C-reactive protein of 6.7 mg/dL (normal range, 0–0.5 mg/dL). Blood and urine cultures were drawn which remained negative for five days.

The patient was continued on her preceding antibiotic regimen of vancomycin and cefepime until cultures were negative for 48 hours. Scheduled acetaminophen treatment did not improve her symptoms. Due to persistent inflammatory neck pain with no known cause, rheumatology was consulted. They recommended a dual-energy CT of the cervical spine which showed multiple calcific deposits surrounding the odontoid with associated soft tissue thickening (Figure 1) consistent with Crowned Dens Syndrome.

The patient was begun on a 10-day course of 25 mg of prednisone daily. Plain films showed erosions in the knee and hand consistent with CPPD. The patient's neck pain and mobility improved rapidly, and she was discharged the next day. Nineteen days later, she was admitted for bacteremia, and her neck pain had recurred. She improved with a higher dose of prednisone and was discharged to complete a longer steroid taper.

DISCUSSION

The differential diagnosis of posterior neck pain has some important considerations [1]. Neoplasm, infection, and primary spinal cord or nerve disease are all serious conditions which can occur. A more diffuse picture of inflammation, such as additional pain in the shoulder or jaw, might be consistent with a more diffuse inflammatory disease like polymyalgia rheumatica and giant cell arteritis [3]. Crowned Dens Syndrome may be less common syndrome than these conditions but is important for practitioners to consider and diagnose. Other published case reports demonstrate the challenges of diagnosing this syndrome including unneeded lumbar punctures being done to rule out meningitis and steroid courses given due to suspicion of giant cell arteritis and polymyalgia rheumatica [4, 5].

Crowned Dens Syndrome occurs due to excessive cartilage pyrophosphate production leading to calcium pyrophosphate crystal deposition around the atlantoaxial joint. The classic triad associated with this condition is headache, fever, and cervical morning stiffness [4]. Most

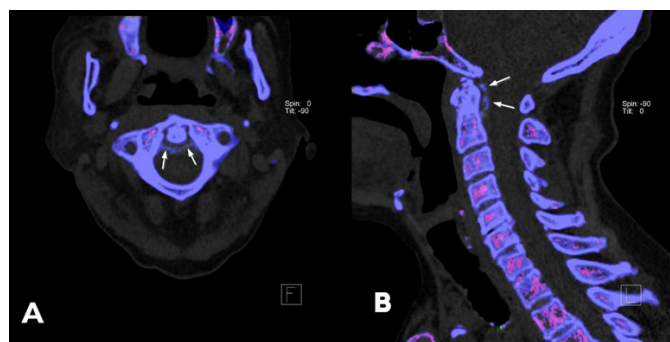


Figure 1: Dual-energy computed tomography (CT) scan at the level of C1–C2 transverse (A) and sagittal (B) showing calcium pyrophosphate crystal deposition surrounding the odontoid process.

patients with Crowned Dens Syndrome have a known history of CPPD with symptoms commonly affecting the knee, wrist, shoulder, or metacarpophalangeal joints [5, 6]. Plain film X-rays are usually adequate for diagnosis of peripheral calcium pyrophosphate deposition, but the overlap of structures within the caudal cervical spine area makes CT imaging focused on C1/C2 the gold standard of diagnosis [5]. Patients with CPPD calcification in peripheral joints have a higher prevalence of odontoid calcification than the general population [7]. While this patient had no known history of pseudogout, plain films of her knee and hand showed findings consistent with CPPD. In addition and in contrast to this case, most acute attacks of CPPD occur after trauma to the area or during acute illness [6] though, potentially, her ongoing cancer may have created an inflammatory state.

As such, clinicians need to be alert to this diagnosis in order to obtain the right imaging to confirm the condition. Both steroids and non-steroidal anti-inflammatory drugs are appropriate [6] if the diagnosis is made.

CONCLUSION

Crowned Dens Syndrome is an uncommon, but important cause of posterior neck pain and should be considered while evaluating patients with this complaint in both the inpatient and outpatient setting. Doing so can lead to quick, successful treatment and unnecessary and invasive procedures can be avoided.

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Author Contributions

Alexander M Pandelidis – Conception of the work, Design of the work, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, 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

Alan W Dow – Conception of the work, Design of the work, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, 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

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Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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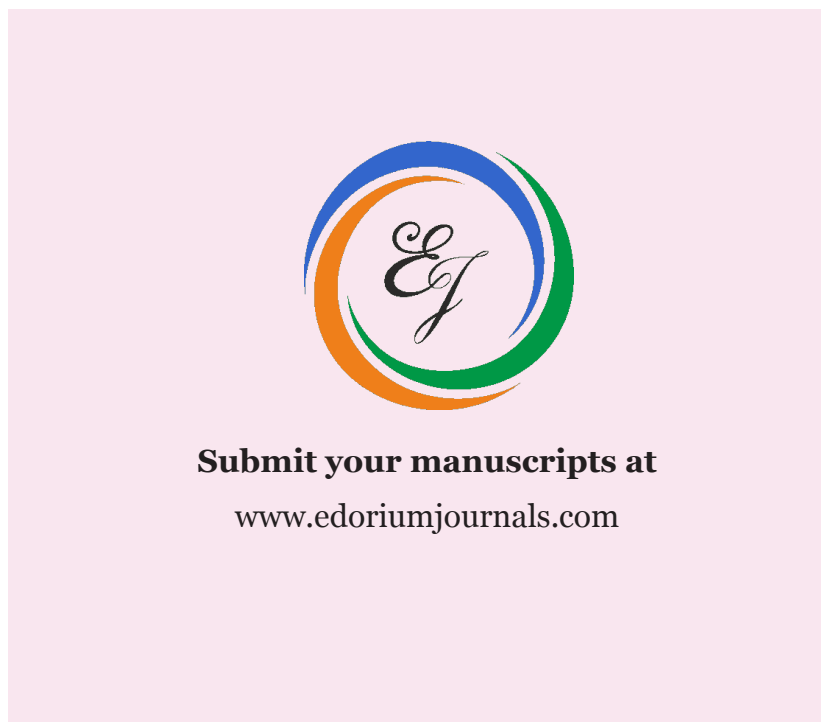
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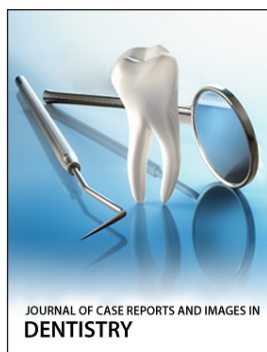
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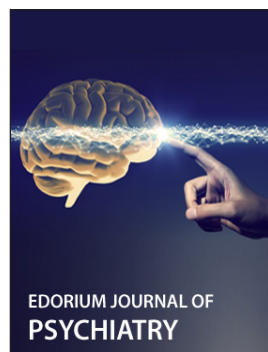
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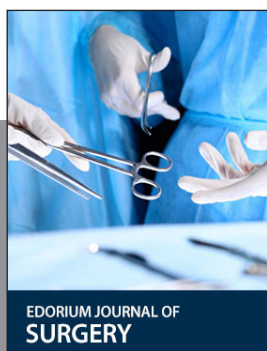
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