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Crowned dens syndrome : a case report followed by an update on differential diagnosis and treatment

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Introduction

Crowned dens syndrome (CDS) is a rare clinical presentation of chondrocalcinosis or calcium pyrophosphate crystal deposition (CPPD) disease, characterized by fever, neck pain and stiffness, associated with increased biological markers of inflammation. Diagnosis is made mainly via imaging, and the best modality remains the magnetic resonance imaging (MRI) of the neck showing calcification and calcium pyrophosphate crystals deposition around the odontoid process (1).

The aim of this report is to increase awareness of this rare and ill-known clinical tableau and to review its complications and the latest modalities of its diagnosis and treatment.

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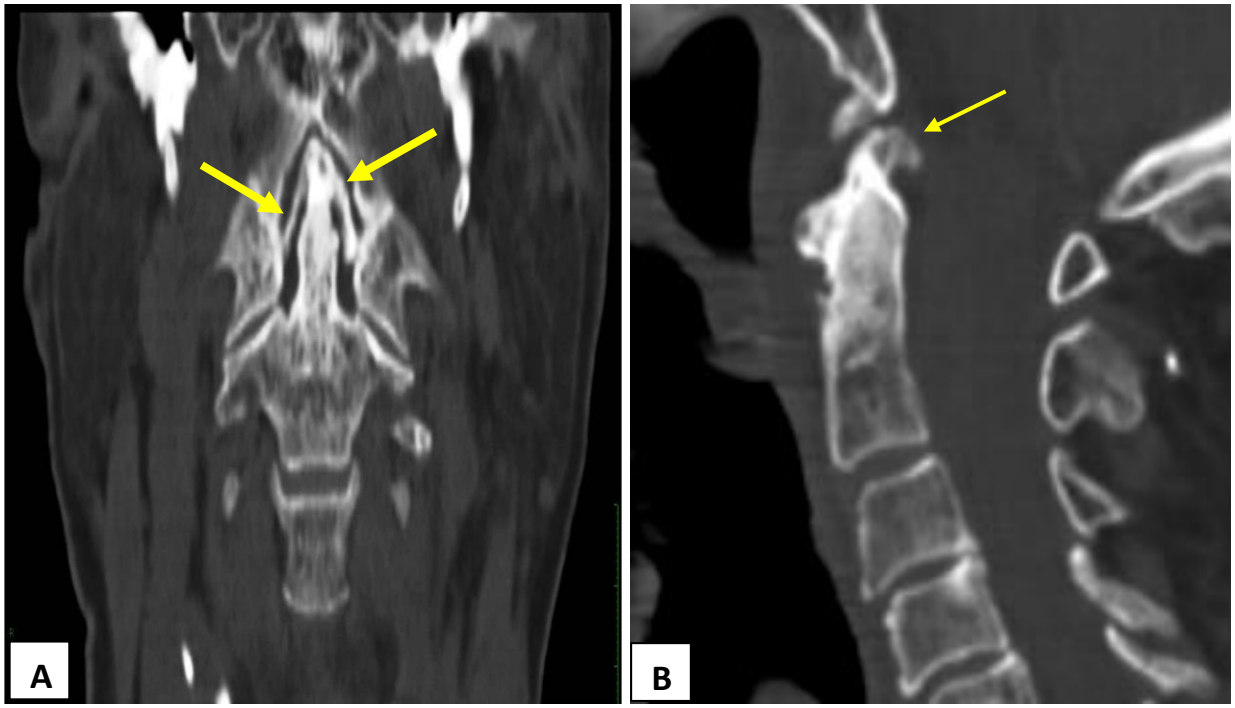
Clinical case:

An 83-year-old female patient presented to the emergency department (ED) of the Middle East Institute of Health (MEIH) with severe acute onset neck stiffness and pain of 10/10 intensity, radiating to the submandibular area and reaching the occipital part of the head, associated with nausea and 2 episodes of chills one day prior to presentation. Patient denied any recent history of trauma or surgery of the head and neck. Physical exam at that time was only remarkable for marked limitation of neck motion, and positive Kernig and Brudzinski signs. No skin changes, facial sinuses tenderness, cervical lymphadenopathies or neck swellings were noted. Vital signs at ED were within normal range except for a low grade fever. Laboratory work-up upon presentation showed hyperleukocytosis (WBC = 14000/mm3) with left shift and a C-reactive protein (CRP) of 189 mg/L (0-5 mg/L). Chest X-Ray only showed degenerative osteoarthritis of the thoracic spinal vertebrae. CT-scan of the brain without IV contrast done urgently showed no acute changes. As Clinical and biological features were suggestive of acute meningitis, cultures were taken including blood, urine and CSF fluid from lumbar puncture. The patient was admitted to the hospital for IV antibiotic treatment with a large spectrum carbapenem and close observation. On next 2 days, the patient continued to develop low grade fever with a fluctuating pattern, and levofloxacin and amikacin shots were added for maximal bacterial coverage. Urine and blood culture yielded positive for Escherichia Coli only resistant to penicillin. CSF fluid analysis and culture was unremarkable. So, the patient continued to receive IV antibiotics for urinary tract infection and bacteremia. And although the cultures were sensitive to the antibiotics given, the patient continued to suffer from fever, headache, neck stiffness with minimal improvement and response to IV acetaminophen. Laboratory markers continued to increase and CRP reached 325. The patient then underwent a series of imaging and laboratory tests to determine the cause of fever, including CT-scan of the chest, abdomen and pelvis with IV contrast, widal, wright, Rose Bengal, stool analysis, stool culture, clostridium difficile toxins, transthoracic and transesophageal cardiac ultrasound that were all negative. Finally, neck MRI was done to rule out spondylodiscitis or a cervical abscess and showed evidence of osteoarthritis on multiple cervical spinal levels, calcification of the odontoid process of C2, and thickening of the synovium and posterior ligaments (Fig C – D). Those results were compatible with a CT-scan of the neck then done and was in favor of a corona of calcification around the odontoid process suggesting a crowned dens syndrome (Fig A – B). The patient was put on colchicine therapy, and started to improve slowly, then corticosteroids were added and yielded a faster improvement. The C-reactive protein finally started to drop until reaching 23 mg/L upon the discharge. The patient was finally discharged on NSAIDs and colchicine. And after 1 month, the patient was seen in an outpatient setting where she reported dramatic improvement of her symptoms.

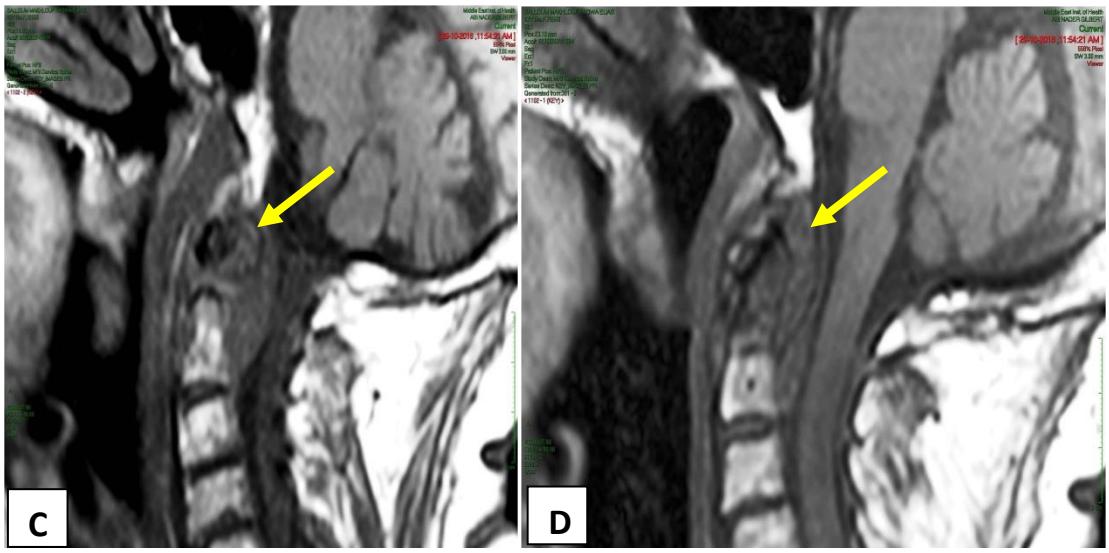
Discussion:

Crowned dens syndrome remains widely an under-recognized entity due to its misleading clinical presentation [1]. The clinical and biological tableau of neck stiffness, headache, and fever associated with elevated inflammatory markers are all suggestive of an acute infection or inflammation such as acute meningitis, osteomyelitis (2), cervical abcess or odontogenic infection (3), giant cell arteritis or polymyalgia rheumatica…etc. As it was her first episode of acute febrile headache, our patient was considered to have acute meningitis upon admission and was treated as such. Nevertheless, the persistence of symptoms besides well adapted antibiotherapy, led the medical team in charge of the patient to consider alternative diagnoses. The patient underwent a work-up for fever of unknown



A) Coronal CT reformation showing a continuous calcification of anterior dento-axial ligaments on both sides of the dens (Arrows)

B) Sagittal CT reformation showing a homogeneous dense tip of the dens due to postinflammatory sclerosis. Large calcifications of and around the apical ligament above the dense (Arrow)



C) T1 weighted sagittal MR image showing an important soft tissue swelling (arrows) posterior to the dens. The tip of the dens shows loss of the normal high signal intensity is vertebrae and is hypointense (due to the chronic inflammatory changes to calcification, as shown on the CT images)

D) T2 weighted sagittal MR image showing a small rim of fluid (arrows) posterior to the tip of the dens, part of the inflammatory changes.

origin, which lead after nine days of hospital stays to the diagnosis of crowned dens syndrome. This delay has been found in many situation in the literature (4) as is the case in a retrospective study where 9 out of 18 cases of crowned dens syndrome were misdiagnosed at their first presentation, leading to additional unnecessary workup [5].

On the other hand, many studies confirm that periodontoid calcification is an aging-related process and that calcification, more frequently observed on head CT in patients with peripheral joints osteoarthritis, may be totally asymptomatic eventhough it predisposes to neck pain and CDS in the future (6,7). It may be also associated with hyperuricemia (8).

Although head and cervical spine CTscan remains the first radiologic tests to evaluate a patient with febrile head and/or neck pain and before practicing the lumbar puncture, it is interesting to note an added value for MRI in the diagnosis and the response to treatment in crowned dens syndrome because the soft tissue around the odontoid process appears hyperintense on magnetic resonance T2-weighted imaging with fat suppression and disappears 2 weeks after treatment with corticosteroids or non-steroidal anti-inflammatory drugs. This advantage was first mentioned in the paper of Inoue A and al. in 2017 (9). In our case, both CT and MRI was performed and showed the typical aspects described hereby.

Moreover Japanese team tried to elaborate a semi-quantitative score based on calcium deposition on the transverse ligament of atlas (TLA) which was well correlated with CRP level, pain score and the treatment effects; in this paper the authors recommend that a low dosage (15-30mg) of corticosteroids be used as first-line drugs rather than conventional NSAID therapy and this steroid dose should be gradually tapered as soon as possible (10). Our patient responded quickly to first line treatment by colchicines and NSAIDs; a short term course of steroids was used at hospital and quickly tapered and stopped within 2 weeks.

If the patient with CDS or gouty arthritis is resistant to first and second line therapy with NSAID, colchicine and steroids, treatment with Anakinra (recombinant interleukin 1 receptor antagonist) may be considered; one retrospective study showed a complete response with a good tolerance in both diseases but a need for a longer duration treatment for patients with crowned dens syndrome (11).

A well known but a rare complication of CDS is the cervical myelopathy due to direct compression by the large calcium pyrophosphate mass at the C1–C2 level causing progressive neurologic deterioration and necessitating surgical decompression and stabilization (12).

Another rare complication is the chronic relapsing form of the CDS that predisposes the patient to steroids side effects and another therapeutic option is to be considered (13). Nevertheless, other chronic diseases such as polymyalgia rheumatic and giant cell arteritis should be ruled out (1).

Conclusion:

Crown dens syndrome is a rare manifestation of chondrocalcinosis that is ill recognized by clinicians. In a febrile patient with inflammatory axial neck pain and decreased cervical range of motion, a full inclusive clinical examination and an early careful imaging studies associated with high index of suspicion are needed to avoid many unnecessary and invasive investigations and to shorten the length of hospital stay.

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