

CASE REPORT

Hiding In Plain Sight - An Incidental Finding Of Crowned Dens Syndrome

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ABSTRACT

Crowned dens syndrome (CDS) is a rare condition in which there is an abnormal deposition of calcium pyrophosphate dihydrate (CPPD) or hydroxyapatite crystals in the ligaments surrounding the odontoid process forming a crown or “halo-like” configuration. It is often misdiagnosed. In this case, we had a patient that had been admitted in view of trauma, however on an incidental finding from the CT scan showed the diagnosis of “Crowned Dens Syndrome”. The aim of this report is to raise awareness to the existence of this condition thus preventing submitting patients to inaccurate treatment and unnecessary investigations.

INTRODUCTION

Crowned dens syndrome (CDS) is a rare condition in which there is an abnormal deposition of calcium pyrophosphate dihydrate (CPPD) or hydroxyapatite crystals in the cruciform and alar ligaments surrounding the odontoid process forming a crown or “halo-like” configuration (1). It was first described by Bouvet et al. in 1985 as a cause of acute neck pain. Crowned dens syndrome can present clinically with acute cervical pain, neck stiffness and increased inflammatory markers.

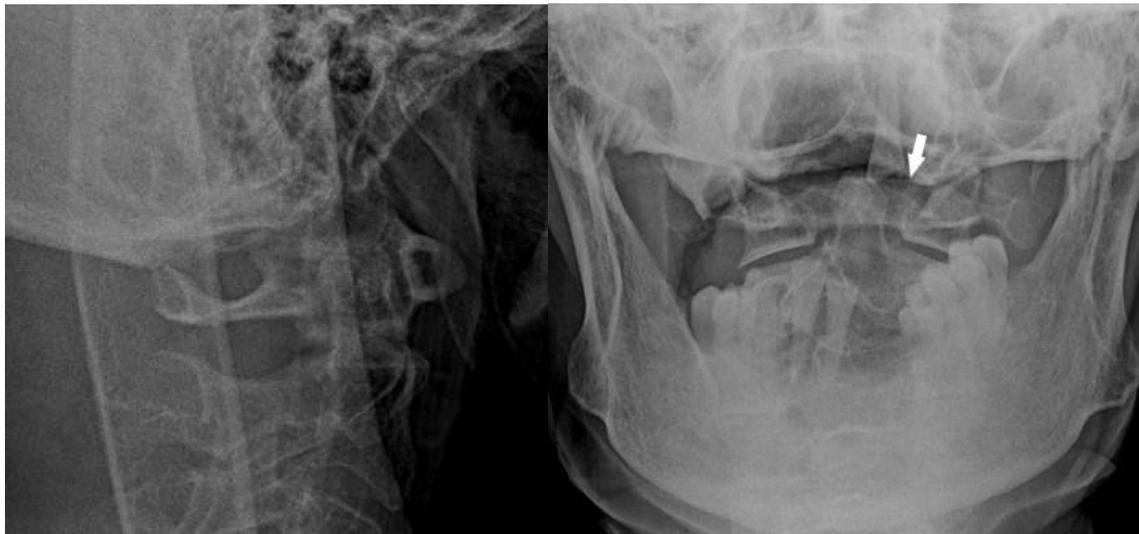
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CPPD crystal is produced by hypertrophic chondrocytes contained in hyaline cartilage. When this occurs at the ligaments surrounding cervicoaxial joint and causes pain and stiffness, it is diagnosed as Crowned Dens Syndrome (2). CDS is often misdiagnosed as polymyalgia rheumatica or meningitis. The aim of this report is to raise awareness to the existence of this condition thus preventing submitting patients to inaccurate treatment and unnecessary investigations.

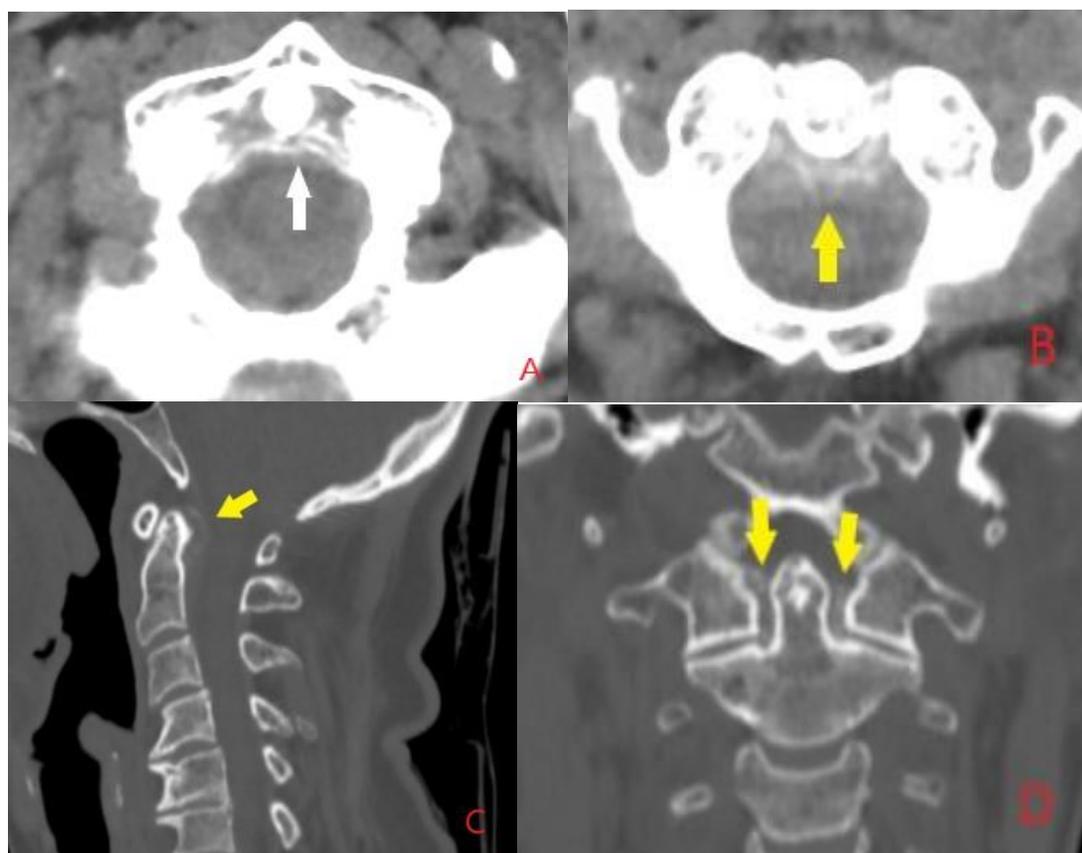
Case

A 68-years old lady presented to our emergency department post motor-vehicle accident with a forehead laceration wound and neck pain. The patient sustained a direct impact to her forehead by a tree branch while riding her motorcycle. She has hypertension treated with a single oral anti-hypertensive (Amlodipine), and chronic obstructive airway disease treated with Salbutamol inhaler on a PRN basis.

On physical examination, there was midline posterior cervical tenderness corresponding to the C2 vertebrae with paravertebral tenderness and no neurological deficit. Plain radiograph showed subtle radiopacity around the odontoid process (Figures 1). Computed tomography revealed a fracture of right transverse process of C1 and calcification surrounding the odontoid process (Figures 2). Her C-reactive protein was 33.6 mg/dL, white cell count $11.4 \times 10^9/L$, ESR 33 mm/hr. On further investigation she denied any past history of neck pain or stiffness prior to the trauma. The patient was placed on a cervical orthosis and given analgesia.



Figures 1: Lateral swimmer view and open mouth view x-ray of the neck. The open mouth view shows radiopacity lesions around the odontoid process (arrow).



Figures 2: **(A & B)**: CT transverse view - “halo-like” calcification seen around the odontoid process (arrow), **(C)**: CT median sagittal view (arrow) **(D)**: CT coronal view - calcification seen around the odontoid process

DISCUSSION

Crowned Dens Syndrome (CDS) is an uncommon inflammatory condition where the patient demonstrates cervical pain and stiffness due to calcium pyrophosphate dehydrate (CPPD) crystals or hydroxyapatite at the apical and alar ligaments (2).

Many cases have been reported, the syndrome typically effects those aged over 60 years and its association with hypomagnesaemia and hyperparathyroidism (3). Pseudogout usually manifest as polyarticular arthritis, usually affecting the peripheral joints; knee, wrist, shoulder and metacarpophalangeal joints (4,5). These attacks are more frequent after local trauma, during acute illnesses and postoperatively. Some may not have any symptoms and are only diagnosed incidentally from imaging.

CDS is also recognized as pseudogout of the atlantoaxial junction due to CPPD, its pathophysiology is still unclear (2,5). The calcification or chondrocalcinosis can occur in few areas including cruciform ligament, alar, transverse and apical ligament (2).

As of now, there is no standard diagnostic criteria for CDS. However, CT scan of the upper cervical region is the gold standard to detect calcification in transverse, apical and alar ligament. Performing plain radiograph systemically over other joints like knee or wrist when there is no specific symptom might be helpful while suspecting crystal deposition other than atlantoaxial joint in the CDS patients' group (2). In most of the cases show elevated inflammatory markers on serum laboratory tests which can help with the diagnosis beside imaging.

CDS differentials are vast. Acute or subacute presentations can be misdiagnosed as meningitis, polymyalgia rheumatica, cervical discitis, giant cell arteritis, epidural abscess, rheumatoid arthritis, osteomyelitis, retropharyngeal abscess or a metastatic tumour, and chronic relapsing presentations may be misdiagnosed as cervicogenic neck pain or occipital neuralgia. Rarely, it can progressively erode the atlantoaxial ligaments and the dens leading to atlantoaxial instability, cervical spinal cord compression and progressive quadriparesis (4).

The first line treatment is a short course of a non-steroidal anti-inflammatory drug (NSAIDs). Patients who cannot tolerate NSAIDs or for whom they do not work low dose corticosteroids are an option (3). Surgery is rarely required. Baysal et al. demonstrated that 1 of 17 patients progressed to myelopathy and was treated by decompression surgery (2).

CONCLUSION

Though rare in prevalence, the physicians should consider CDS in the deferential diagnosis of elderly with febrile neck pain. Failure to recognize it may result in inappropriate invasive investigations such as lumbar puncture and subsequent treatment with parenteral broad-spectrum antimicrobial therapy or prolonged external neck immobilization.

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