

## CLINICAL VIGNETTE

# The Clinical Spectrum of Crowned Dens Syndrome: An Analysis of Seven Cases

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### Abstract

The crowned dens syndrome is an unusual manifestation of calcium pyrophosphate dihydrate crystal deposition disease and represents a rare cause of neck pain. We report seven cases of crowned dens syndrome, assessing clinical and radiological features, associated conditions, therapy, and outcome. The acute presentation of crowned dens syndrome raised suspicion for infectious meningitis and cranial giant cell arteritis in two cases. Cervical cord compression was the initial presentation of periodontoid calcium pyrophosphate deposition disease in two other cases. A prior history or concurrent, self-limited, peripheral arthritis was clinically or radiologically apparent. Computed tomography scans performed better than magnetic resonance imaging in assessing calcifications of the dens area. We found a unique association with diffuse idiopathic skeletal hyperostosis, Eagle's syndrome, and Paget's disease of bone. Prompt initiation of therapy including isolated or combinations of non-steroidal anti-inflammatory drugs, corticosteroids, and colchicine yielded a dramatic response.

### Introduction

Crowned dens syndrome (CDS) was first described in 1985 by Bouvet et al<sup>1</sup> and represents a rare cause of severe neck pain in older adults. This radioclinical syndrome is defined by the association of periodontoid calcifications in a crown-like configuration and periodic attacks of febrile neck pain and stiffness.<sup>2</sup> This report illustrates the clinical and radiological features of seven cases of patients with CDS.

### Discussion

Crowned dens syndrome (CDS), also known as acute pseudogout of the neck, is an unusual manifestation of calcium pyrophosphate dihydrate crystal deposition disease (CPPD) and is related to the microcrystalline deposition of calcium pyrophosphate crystals in a periodontoid distribution.<sup>1</sup> CDS usually occurs in patients over 60 years of age and has a female predominance.<sup>3</sup>

CDS is characterized by periodic attacks of acute, severe neck, and shoulder girdle pain and stiffness mainly while attempting to rotate the head from side to side.<sup>3,4</sup> Fever, headaches, and raised inflammatory markers are frequently present.<sup>2,4</sup> As illustrated in the cases we will present in this report, a prior

history or concurrent, self-limited, peripheral arthritis may be clinically or radiologically apparent.<sup>5</sup> CPPD deposition can be precipitated by an underlying metabolic condition like hyperparathyroidism. We found a unique association with diffuse idiopathic skeletal hyperostosis, Eagle's syndrome, and Paget's disease of bone.

The acute presentation of CDS frequently raises suspicion for infectious meningitis, epidural abscess, polymyalgia rheumatica, giant cell arteritis, and/or metastatic malignancy.<sup>5</sup> A thorough investigation, such as we describe in cases 5 and 7, is necessary to make the correct diagnosis of CPPD.

Cervical CPPD is common in patients with peripheral CPPD, often remaining asymptomatic.<sup>6-8</sup> In a retrospective study conducted by Salaffi et al,<sup>6</sup> in 25 of 49 patients (51%) affected by peripheral CPPD, computed tomography (CT) scan of the cervico-occipital junction showed periodontoid calcified deposits. Only nine of the cervical CPPD cases had neck symptoms.

CT scanning performs better than magnetic resonance imaging (MRI) in assessing calcifications of the dens area and, therefore, considered the gold standard for diagnosis.<sup>6</sup> Plain radiograph is notoriously insensitive in detecting periodontoid opacification. CPPD crystal deposits in and around C1-C2 can lead to osseous abnormalities of the odontoid process, such as subchondral cysts, erosions, and even fractures of the odontoid.<sup>6,9</sup>

Cases 1 and 2 were particularly interesting, as cervicomedullary compression was the initial presentation of periodontoid CPPD. To date, few cases of cervical CPPD causing atlantoaxial instability, spinal cord compression, and myelopathy have been reported.<sup>10-12</sup> Similar to our index patient, most lesions were treated by surgical decompression via the transoral route with good post-surgical outcomes.<sup>13</sup>

Treatment first involves non-steroidal anti-inflammatory drugs, which are highly effective within days. Systemic corticosteroids, C1-2 steroid injection, or colchicine have also been used with good clinical efficacy.<sup>1,2,4,5,14</sup>

### *Case 1*

A 64-year-old man was admitted to the intensive care unit for respiratory failure and quadriplegia. He had a history of thyroid cancer treated with surgical resection, currently on thyroid replacement, newly-diagnosed primary hyperparathyroidism, and diffuse idiopathic skeletal hyperostosis. On examination, he could not lift all four extremities against gravity. There was no clear sensory level.

MRI of the cervical spine revealed low signal intensity mass on T2-weighted image posterior to the C2 vertebral body leading to severe canal compromise and myelopathic signal changes within the cord [Figure 1A-C]. On non-enhanced CT imaging, the mass had amorphous mineralization compatible with CPPD [Figure 1D].

The patient underwent C1 through 5 laminectomies with posterior instrumented fusion. Four months after the initial presentation, his quadriplegia completely resolved.

### *Case 2*

An 84-year-old man was seen by neurosurgery for severe neck pain and cord compression on imaging. He had been well until 2 years earlier, when he developed neck pain and stiffness that was initially intermittent but became constant. His pain was localized to the left side of the neck with radiation to the left parietal area and left forehead. He denied numbness or weakness in the arms or legs, swallowing or breathing difficulties. He had a history of generalized osteoarthritis, and prior C3-7 laminectomy. On examination, there was tenderness on palpation of the cervical spine with limited range of motion in all planes.

MRI scans showed a large nonenhancing soft tissue mass involving the C1-2 articulation with a cystic component extending into the canal on the left side, causing mild cord compression. There was no high signal seen within the cervical spinal cord at this level [Figure 2A-D]. A CT scan demonstrated amorphous calcifications within the soft tissue mass and mild erosive changes of the dens, most compatible with cervical deposition of calcium pyrophosphate crystals [Figure 2E, F]. Bilateral hand and knee radiographs showed chondrocalcinosis and findings typical of calcium pyrophosphate arthropathy.

Given the absence of signs and symptoms of a progressive myelopathy, non-operative treatment with low-dose prednisone and topical diclofenac was initiated. The neck pain and stiffness significantly improved.

### *Case 3*

A 71-year-old man was evaluated for a 5-year history of episodic upper cervical pain and stiffness. He had a history of documented crystal pseudogout affecting the wrists and knees. Other medical problems included stage IV chronic kidney disease, secondary hyperparathyroidism and coronary artery disease. He had tenderness and restriction of neck movement, especially on any attempt to rotate the head from side to side. His erythrocyte sedimentation rate (ESR) was 90 mm/h, and C-reactive protein was 12 mg/dL (range, 0.0-1.0 mg/dL).

Cervical MRI revealed soft tissue thickening at the C1-2 level extending slightly within the spinal canal without compression of the spinal cord [Figure 3A]. A subsequent CT showed a pannus with crown-like mineralization surrounding and slightly eroding the dens. On dual energy images, the calcification had blue signal similar to the cortex of the skull base and cervical spine, consistent with calcium pyrophosphate crystals [Figure 3B-D]. Skeletal radiographs revealed chondrocalcinosis at bilateral hips, knees, wrists and pubic symphysis. Treatment with prednisone 5 mg daily and colchicine 0.6 mg twice daily was followed by rapid improvement in neck symptoms and reduction in the frequency of flare-ups.

### *Case 4*

A 54-year-old man was seen with diffuse chondrocalcinosis. He had a history of a chronic arthropathy manifesting as mild joint pain and stiffness of knees, wrists, metacarpophalangeal joints, and shoulders. He reported intermittent neck pain associated with marked stiffness. Physical examination elicited tenderness throughout the posterior cervical spine and severe loss of neck and shoulder movement.

Plain radiographs showed multilevel cervical spondylosis, and 1st through 3rd metacarpophalangeal predominant arthropathy with chondrocalcinosis of knees, bilateral glenohumeral, and acromioclavicular joints. Laboratory workup including ferritin level, parathyroid hormone, calcium, magnesium, and alkaline phosphatase was normal. A cervical CT revealed odontoid calcifications affecting the transverse ligament of the atlas in a 'crown-like' appearance, compatible with CPPD [Figure 4A, B].

A trial of nonsteroidal anti-inflammatory drugs was unsuccessful because of gastric intolerance. He was started on short cycles of low-dose prednisone with satisfactory control of his neck symptoms.

### *Case 5*

A 92-year-old man presented to the emergency department with complaints of severe headaches and neck pain for 5 days. Initial clinical examination found cervical stiffness with Brudzinski's sign. Laboratory tests showed white blood cells 14 400/mm<sup>3</sup>, ESR 71 mm/hr, and C-reactive protein 18 mg/dL. Infectious meningitis was suspected; however, a lumbar puncture was normal.

Skeletal radiographs revealed extensive chondrocalcinosis of shoulders, knees, and wrists. A cervical CT scan of C1/C2 allowed the diagnosis of crowned dens syndrome showing small mineralization behind the odontoid process in addition to subtle posterior longitudinal ligament mineralization at C2-C3 and C3-C4 [Figure 4C].

His headaches, neck pain, and stiffness dramatically improved 3 days later with low-dose prednisone and with normalization of biological parameters in 14 days.

### Case 6

An 86-year-old man was hospitalized with upper cervical thoracic pain that had profoundly worsened over the last week. The pain was severe, nonradicular, nonmyelopathic, and unrelated to time of day, positioning, or activity. He had a history of tophaceous gout, Paget's disease of bone, generalized osteoarthritis, and severe osteoporosis. Examination showed greatly limited cervical extension and flexion, particularly lateral rotation.

CT examination of the cervical spine revealed extensive multilevel degenerative changes, ossification of the stylohyoid ligament bilaterally, and chondrocalcinosis present at the C1-2 level compatible with CPPD [Figure 4D].

Treatment with prednisone 10 mg daily and low dose colchicine resulted in rapid symptom improvement. The patient died 1 month later from unrelated cryoglobulinemic vasculitis.

### Case 7

An 85-year-old woman developed intermittent right-sided headache, which became daily after a couple of weeks. The head pain was felt most intensely in the right temporal-occipital area. There was no jaw or tongue claudication, visual disturbances, diffuse myalgias, or constitutional symptoms. She had a history of crystal-proven pseudogout of the ankle, C1 vertebral body ring fracture from a previous fall, osteoporosis, and diabetes mellitus. The neurological and general examinations were normal, except for a reduced range of motion of the neck and slight tenderness over the right temporal artery.

She was referred to a rheumatologist for suspicion of cranial giant cell arteritis. Her ESR was 55 mm/h, and C-reactive protein was normal. She received steroid therapy in anticipation of the temporal biopsy. Giant cell arteritis was not confirmed on histological examination of the right temporal artery.

A cervical CT showed atlantoaxial instability with widening of the predental space, cranial settling with occipital condyles essentially articulating with C2 lateral masses due to separate C1 lateral masses as a consequence of the previous C1 fracture and an extensive retrodental partially calcified soft tissue mass leading to marked disintegration of dens and mild spinal canal stenosis consistent with CPPD [Figure 4E-G]. The neurosurgeon opted against surgical intervention because of absence of neurological signs and cervical myelopathy.

The patient was prescribed colchicine 0.6 mg daily. After a week, her headache improved. After 2 months, colchicine was stopped as she was completely asymptomatic.

### Conclusion

Clinicians should be aware of CDS as a significant cause of acute febrile neck pain in older adults. Often asymptomatic, CDS is likely under recognized, as many cases go undiagnosed. The prompt initiation of therapy usually yields a dramatic response. Long-term follow-up is necessary, as neck CPPD can

sometimes lead to cord compression with significant neurological sequelae.

### Table and Figures

**Table 1.**

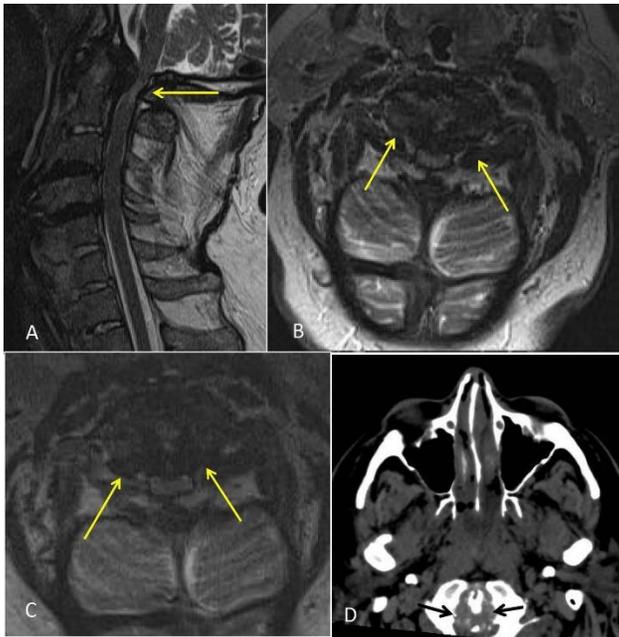
TABLE 1. Clinical and Imaging Features in Seven Patients With Crowned Dens Syndrome (CDS)				
Case	1	2	3	4
Age (yrs)/ Sex	64/M	84/M	71/M	54/M
Symptoms at presentation	Respiratory failure, quadriparesis	Neck pain, stiffness	Episodic neck pain, stiffness	Intermittent neck pain
Elevated ESR/CRP	NC	NC	Yes/Yes	NC
First evoked diagnosis	Cervical cord compression	Cervical cord compression	Chronic neck pain	Chronic neck pain
Previous / Concurrent peripheral arthritis	No	No	Yes	Yes
X-rays peripheral joints	None	Chondrocalcinosis	Chondrocalcinosis	Chondrocalcinosis
Cervical CT imaging for CDS	Calcifications	Calcification, erosion of dens	Calcification, erosion of dens	Calcifications
Treatment	Laminectomy	Prednisone	Prednisone, colchicine	NSAIDs, Prednisone
Outcome	Resolution of quadriparesis	Improvement in neck pain	Reduction in flare-up frequency	Improved in neck pain

NC, Not check; NSAID, non-steroidal anti-inflammatory drug

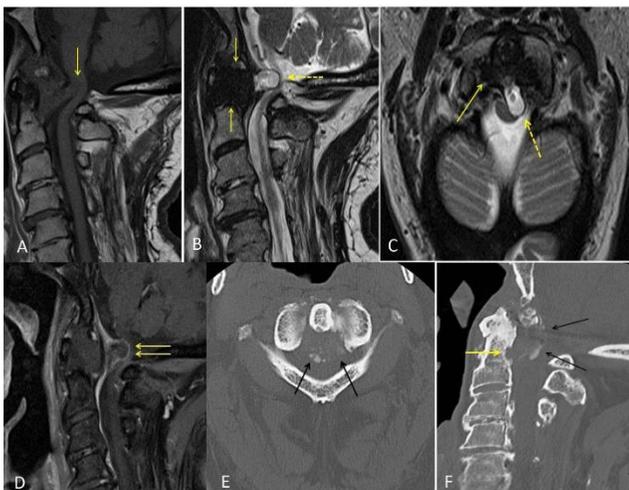
TABLE 1. Con't.			
Case	5	6	7
Age (yrs)/ Sex	92/M	86/M	85/F
Symptoms at presentation	Acute headaches, neck pain and stiffness	Acute cervical pain and decreased range of motion	Right temporal headaches
Elevated ESR/CRP	Yes/Yes	NC	Yes/No
First evoked diagnosis	Infectious meningitis	Acute neck pain	Giant cell arteritis
Previous / Concurrent peripheral arthritis	No	No	Yes
X-rays peripheral joints	Chondrocalcinosis	None	None
Cervical CT imaging for CDS	Calcifications	Calcifications	Cranial settling, calcifications, erosion of dens
Treatment	Prednisone	Prednisone, colchicine	Colchicine
Outcome	Resolution of symptoms	Improvement in symptoms	Complete resolution

NC, Not check; NSAID, non-steroidal anti-inflammatory drug

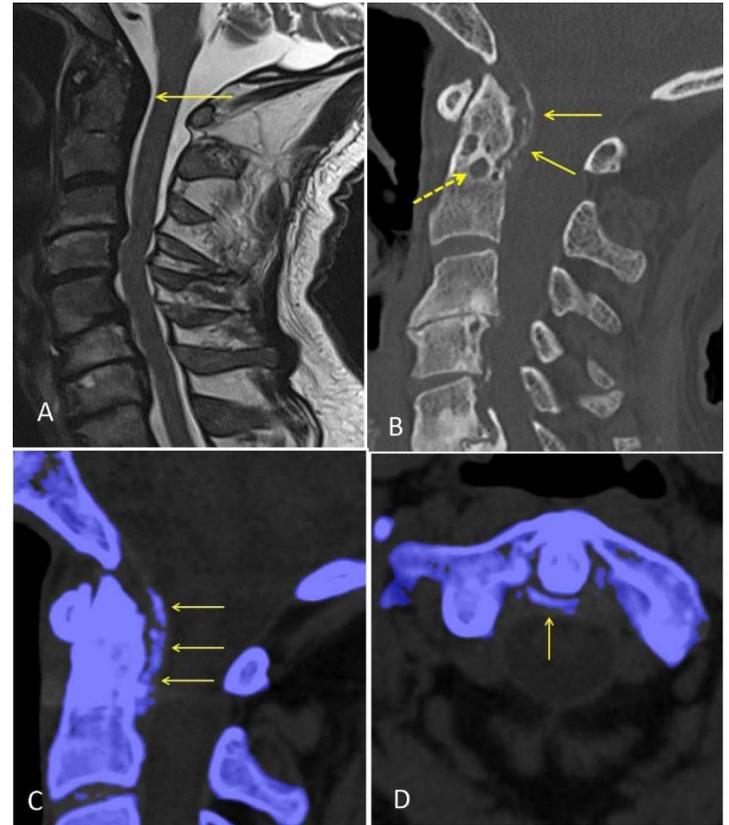
**Figure 1.** Cervical spine MRI sagittal T2-weighted (A) and axial T2-weighted images (B,C). Pannus with low T2-weighted mineralization (arrows), compatible with CPPD, surrounds the dens, extends within the spinal canal and causes severe central canal stenosis and compression of the spinal cord. (D) Head CT scan, axial image. Pannus with amorphous mineralization (arrows), compatible with CPPD, surrounds the dens, extends within the spinal canal, and causes severe central canal stenosis and compression of the spinal cord.



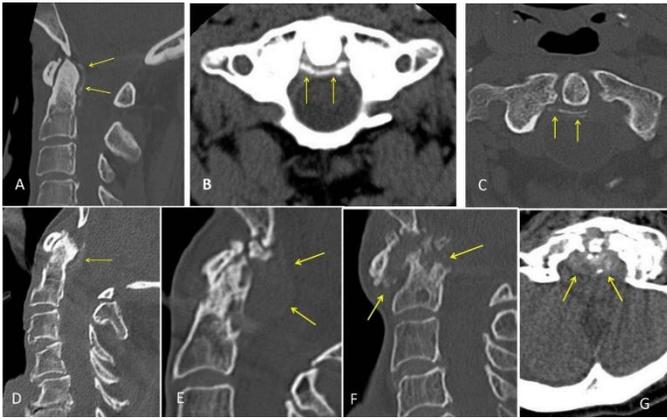
**Figure 2.** Cervical spine MRI sagittal T1-weighted (A), sagittal T2-weighted (B), axial T2-weighted (C) and T1 fat saturation post contrast images (D). Nonenhancing pannus (double arrow) with low T1 and T2-weighted mineralization (single arrows) and a cystic component (dashed arrow), compatible with CPPD, surrounds the dens, extends within the spinal canal and causes severe central canal stenosis and compression of the spinal cord. Cervical spine CT scan axial (E) and sagittal images (F). Pannus with amorphous mineralization (black arrows), compatible with CPPD, surrounds, and slightly erodes the dens (yellow arrow), extends within the spinal canal and causes severe central canal stenosis and compression of the spinal cord.



**Figure 3.** Cervical spine MRI sagittal T2-weighted image. (A) Pannus with low T2-weighted mineralization (arrow), compatible with CPPD, surrounds the dens and slightly extends within the spinal canal without compression of the spinal cord. Cervical spine CT sagittal (B), dual energy sagittal (C), and dual energy axial images (D). Pannus with amorphous mineralization (arrows), compatible with CPPD, surrounds and slightly erodes the dens (dashed arrow). On the dual energy images (B,C), the CPPD is blue due to calcium, which matches the same blue color as the adjacent bone.



**Figure 4.** Cervical spine CT sagittal (A) and axial with soft tissue algorithm images (B). Mineralization surrounding the dens (arrows), compatible with CPPD. Cervical CT axial (C) at the C1/C2 level showing linear calcifications of the transverse ligament of the atlas. Cervical spine CT sagittal (D). Pannus with amorphous mineralization (arrow), compatible with CPPD, surrounds the dens. CT cervical spine sagittal (E) shows extensive soft tissue thickening surrounding the dens with erosion secondary to CPPD. The pannus formation displaces the cervicomedullary junction posteriorly; (F) shows extensive erosion of the dens with adjacent osseous debris and mineralization, compatible with CPPD. Marked diastasis of the atlantodental interval and cranial settling are due to a chronic C1 Jefferson fracture, which are not completely visualized on this image. CT cervical spine axial image (G) shows mineralization and pannus surrounding an eroded dens, compatible with CPPD.



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