

CPPD Crowned Dens Syndrome with clivus destruction: A case report

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ABSTRACT

We report a case of CPPD crowned dens syndrome in an 87 year white old male with a known history of pseudogout, with clinical and radiological features characteristic of this syndrome. Interestingly, there was significant mass effect on the clivus, with clivus erosion and destruction, a finding that has not previously been described with this syndrome. The clinical and radiological characteristics of Crowned Dens syndrome, as well as CPPD are reviewed. We suggest that CPPD crowned dens syndrome may be included in the differential diagnosis when clivus destruction or erosion, in association with a soft tissue mass with calcification, is seen.

CASE REPORT

CASE REPORT

CPPD Crowned Dens Syndrome is uncommon and has been reported in the form of individual case reports [1-14]. Erosion of the clivus associated with crowned dens syndrome has not been reported previously.

We report a case of an 87 year old white male with a known history of pseudogout and chronic renal failure, who presented to the Emergency Room with a history of chronic neck pain, but now with clinical features of acute progressive quadriplegia. He had been able to walk half a block on his own approximately 1 week prior to presentation. Over that week the patient began using a cane. On the day of presentation the patient woke up unable to move his legs and get out of bed.

On presentation, he was awake and fully oriented. His cranial nerve function was intact and he had retained sensation throughout. However he demonstrated decreased rectal tone and complained of significant urinary retention. He had partial 3/5 strength in his deltoids and biceps, but was 0/5 in his triceps, hand grip, and below that. He was also 0/5 strength in his bilateral lower extremities. There was no evidence of spinal tenderness. He did not have fever or an elevated white

count. Aside from elevated creatinine, laboratory assessment was unremarkable.

There was no evidence of acute intracranial abnormality on CT scan of the head. The patient was not able to undergo MRI due to the presence of a pacemaker, and could not receive IV contrast due to his poor renal function.

His neurological examination was consistent with cord compression, and a cervical noncontrast CT scan showed the presence of extensive degenerative disk disease with diskal and soft tissue calcification. The dens was eroded and remodeled posteriorly, with a calcified soft tissue mass that demonstrated internal but predominantly peripheral calcification (Figures 1- 3). The soft tissue mass had an obtuse angle with the posterior vertebral body suggesting an extradural or dural based lesion, with anterior compression of the lower brainstem and upper cervical cord (Figure 2) . The inferior portion of the clivus was eroded by the soft tissue mass indicating the presence of significant mass effect. However the lesion was identified retrospectively on CT scans of the head from 6 and 8 years prior (Figure 4), which indicated that the lesion was slow growing. The radiological diagnosis was that of an inflammatory lesion, either calcium

pyrophosphate deposition disease or, less likely, rheumatoid arthritis. The differential diagnosis included neoplasms such as meningioma, chordoma, chondrosarcoma, metastases, and PVNS (pigmented villonodular synovitis).

The patient was taken to the Operating Room, and posterior surgical decompression with spinal fusion was performed. However the patient did not recover from his quadraparesis. A few days after surgery, the patient subsequently expired suddenly following PEA (pulseless electrical activity) cardiac arrest.

A surgical sample from the posterior cervical elements showed chalky white deposition in the gross specimen (Figure 5), and microscopy showed chondrocalcinosis with the presence of rhomboid shaped positively birefringent crystals, consistent with calcium pyrophosphate deposition disease (Figures 6,7).

DISCUSSION

The entity of Crowned Dens Syndrome was first described by Bouvet et al in 1985, who described it as a clinical and radiological entity characterized by acute neck pain and evidence of calcium deposits around the dens on radiographs[1]. Some authors have differentiated it from the purely radiological entity of isolated calcification around the dens [2]. The deposition of calcium around the dens of C2 is well known, and may occur in Calcium hydroxyapatite deposition disease, CPPD, and ankylosing spondylitis. Clinical features of Crowned Dens Syndrome include acute onset of severe neck pain, restricted range of motion (especially rotation) and positive inflammatory indicators. More chronic neck pain has also been described[3,4].

In the English language literature, only 35 patients from 14 different institutions have been reported [1-14]. However, Goto et al suggests that this entity may be more common than previously thought [15]. In their study, 40 patients out of 2023 studied demonstrated the clinical and radiological features to support the diagnosis of CDS (crowned dens syndrome). Whether this may be related to the ethnicity of the patient population (the study was performed in Japan) will require further investigation. More than two thirds of cases are found in patients older than 70 years of age and females are slightly more affected [15]. The exact incidence of CDS in patients with CPPD arthropathy is not known. A falsely low incidence is suspected as patients receive treatment for neck pain in the outpatient community clinic setting and the fact that most physicians are not aware of this uncommon syndrome.

The clivus is a central skull base bone that forms a shallow depression behind the dorsum sellae that slopes obliquely backward. It forms a gradual sloping process at the anterior most portion of the basilar occipital bone at its junction with the sphenoid bone. On axial planes, it sits just posterior to the sphenoid sinuses. Just lateral to the clivus bilaterally is the foramen lacerum which contains the internal carotid artery, proximal to its anastomosis with the Circle of

Willis. Posterior to the clivus is the basilar artery. The clivus supports the upper part of the pons. Tumors that can be seen originating in and around the clivus with a similar appearance to CPPD Crowned Dens Syndrome are chordoma, chondrosarcoma, and meningioma.

Calcium pyrophosphate deposition disease is a metabolic arthropathy caused by the deposition of calcium pyrophosphate dihydrate (CPPD) in and around joints, especially in articular and fibrocartilage. The etiology of CPPD is unknown, although increased breakdown of adenosine triphosphate resulting in increased pyrophosphate levels in joints, is believed to be one mechanism by which crystals develop[16]. There is also evidence that the gene ANKH (ankylosis progressive homolog) is involved in crystal related inflammatory reactions and inorganic phosphate transport[16]. Excess calcium also has a probable relationship with chondrocalcinosis with hemochromatosis and hyperparathyroidism being two major risk factors.

Although CPPD is often asymptomatic, with only radiographic changes such as chondrocalcinosis, various clinical manifestations may occur, including acute arthritis (pseudogout) and chronic arthritis. In pseudogout, the deposition of crystals in the synovial space can lead to an intense inflammatory reaction leading to clinical features that resemble infection or acute gouty arthritis. Bone destruction may occur as a result of the inflammation.

Although almost any joint may be involved by CPPD, the knees, wrists, and hips are most commonly affected[16]. This condition is the most common cause of secondary metabolic osteoarthritis.

There is no cure for CPPD arthropathy, treatment usually consists of symptomatic control. Nonsteroidal anti-inflammatory drugs and corticosteroids are the mainstay of treatment for exacerbations, with colchicine used to prevent recurrent attacks. Corticosteroids may be either given in pill form or injected directly into the affected joint. The prognosis of Crowned Dens Syndrome is excellent. Using high dose nonsteroidal anti-inflammatory drugs and/or corticosteroids, the majority of patients fully recover within one week of treatment [15]. Only 9 of the 40 patients with CDS in the Goto et al study relapsed within 9 months of original treatment [15]. Although severe neurological complications are rare, extensive CPPD deposits may result in myelopathy or cervical stenosis, for which surgical decompression may be necessary as in our case.

In CPPD Crowned Dens Syndrome, the calcification is typically deposited in the synovial membrane, transverse ligament and alar ligaments [15]. The inflammatory reaction that occurs with the deposition of these CPPD crystals typically leads to erosions and cystic change in the dens, with the presence of a slowly enlarging inflammatory soft tissue component occurring as the disease progresses. This soft tissue component may cause mass effect on the cervical spinal cord, but mass effect on the clivus with erosion and destruction of the inferior clivus as in our patient, has to our knowledge, never been described.

TEACHING POINT

CPPD Crowned Dens Syndrome should be included in the differential of clivus erosion by a calcified soft tissue mass, in addition to other lesions such as chordoma or chondrosarcoma of the clivus.

REFERENCES

1. Bouvet JP, le Parc JM, Michalski B, Benlahrache C, Anquier L. Acute neck pain due to calcifications surrounding the odontoid process: the crowned dens syndrome. *Arthritis Rheum.* 1985;28:1417-20. PMID: 4084331
2. Malca SA, Roche PH, Pellet W, Combalbert A. Crowned dens syndrome: a manifestation of hydroxy-apatite rheumatism. *Acta Neurochir (Wien).* 1995;135:126-30. PMID: 8748801
3. Aouba A, Vuillemin-Bodaghi V, Mutschler C, De Bandt M. Crowned dens syndrome misdiagnosed as polymyalgia rheumatica, giant cell arteritis, meningitis or spondylitis: an analysis of eight cases. *Rheumatology.* 2004;43:1508-12. PMID: 15956089
4. Wu DW, Reginato AJ, Torriani M, Robinson DR, Reginato AM. The crowned dens syndrome as a cause of neck pain: report of two new cases and review of the literature. *Arthritis Rheum.* 2005;53:133-7. PMID:15696551
5. Baysal T, Baysal O, Kutlu R, Karaman I, Mizrak B. The crowned dens syndrome: a rare form of calcium pyrophosphate deposition disease. *Eur Radiol.* 2000;10:1003-5. PMID:10879718
6. el-Khoury GY, Tozzi JE, Clark CR, Foucar E, Menezes AH, Smoker WR. Massive calcium pyrophosphate crystal deposition at the craniovertebral junction. *AJR Am J Roentgenol.* 1985;145:777-8. PMID: 2994449
7. Wells CR, Morgello S, DiCarlo E. Cervical myelopathy due to calcium pyrophosphate dihydrate deposition disease. *J Neurol Neurosurg Psychiatry.* 1991;54:658-9. PMID:1895139
8. Wendling D. Syndrome de l'odontoide couronnee. *Synoviale.* 1992;10:25-7.
9. Alcalay M, Debiais F, Marco M, Wendling D, Tavernier Ch, Le Goff P, Le Parc JM. The crowned dens syndrome and apatite deposition disease. A sexlinked disease? *Arthritis Rheum.* 1992;35:s212.
10. Constantin A, Bouteiller G. Acute neck pain and fever as the first manifestation of chondrocalcinosis with calcification of the transverse ligament of the atlas. Five case-reports with a literature review. *Rev Rhum Engl Ed.* 1998;65:583-5. PMID:9809363
11. Mula M, Bordin G, Naldi P, Gaviani P, Leone M, Monaco F. Crowned dens syndrome in an elderly man. *Neurology.* 2001;56:275. PMID:11160974
12. Kobayashi Y, Mochida J, Saito I, Matui S, Toh E. Calcification of the alar ligament of the cervical spine: imaging findings and clinical course. *Skeletal Radiol.* 2001;30:295-7. PMID:11407723
13. Sato Y, Yasuda T, Konno S, Kuwayama A, Komatsu K. Pseudogout showing meningoencephalitic symptoms: crowned dens syndrome. *Intern Med.* 2004;43:865-8. PMID:15497527
14. Ritter J, Kerr LD, Valeriano-Marcet J, Spiera H. ACTH revisited: effective treatment for acute crystal induced synovitis in patients with multiple medical problems. *J Rheumatol.* 1994;21:696-9. PMID:8035395
15. Goto S, Umehara J, Aizawa T, Kokubun S. Crowned Dens syndrome. *J Bone Joint Surg Am.* 2007 Dec;89(12):2732-6. PMID: 18056506
16. Saadeh C. Calcium Pyrophosphate Deposition Disease. <http://emedicine.medscape.com/article/330936-overview>. September 2009

FIGURES

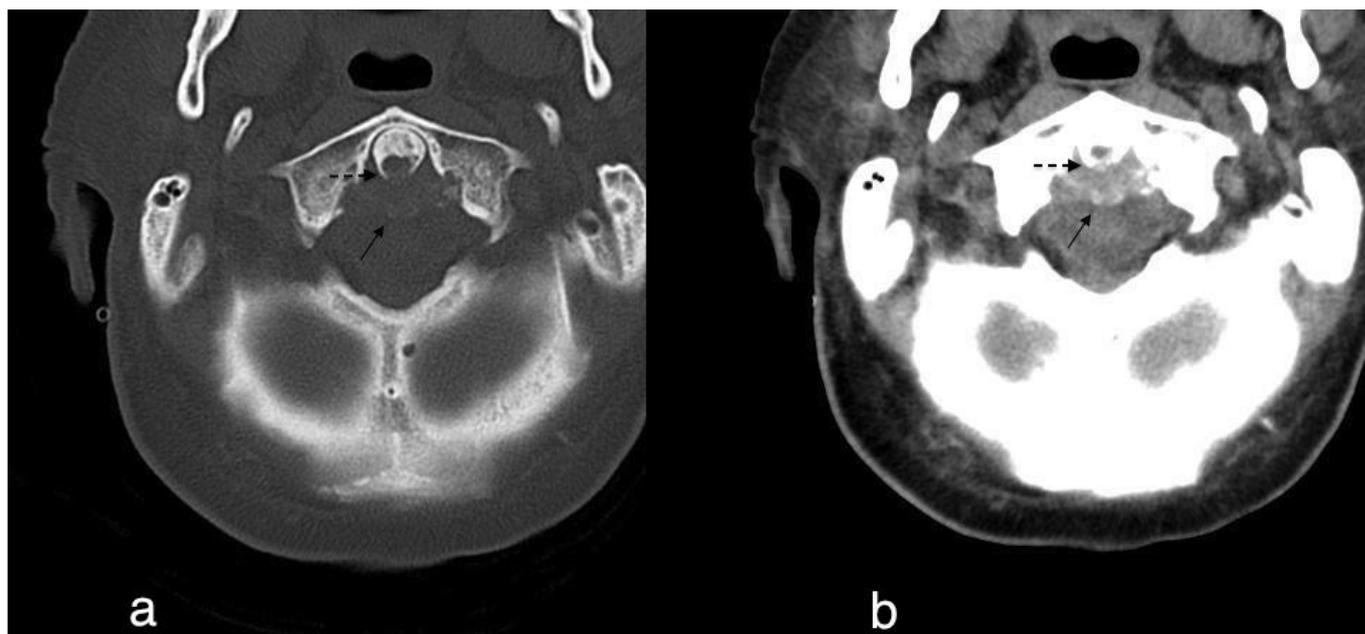


Figure 1: 87 year old male with Crowned Dens Syndrome. Axial images of the cervical spine at the level of the dens, in bone (a) and soft tissue (b) windows, demonstrate erosion and posterior remodeling of the dens (dashed arrows) with a peripherally calcified soft tissue mass in a "halo or crown-like" appearance (solid arrows). Protocol: Siemens CT, mAs 376, KV 120, 3 mm slice thickness, without contrast.

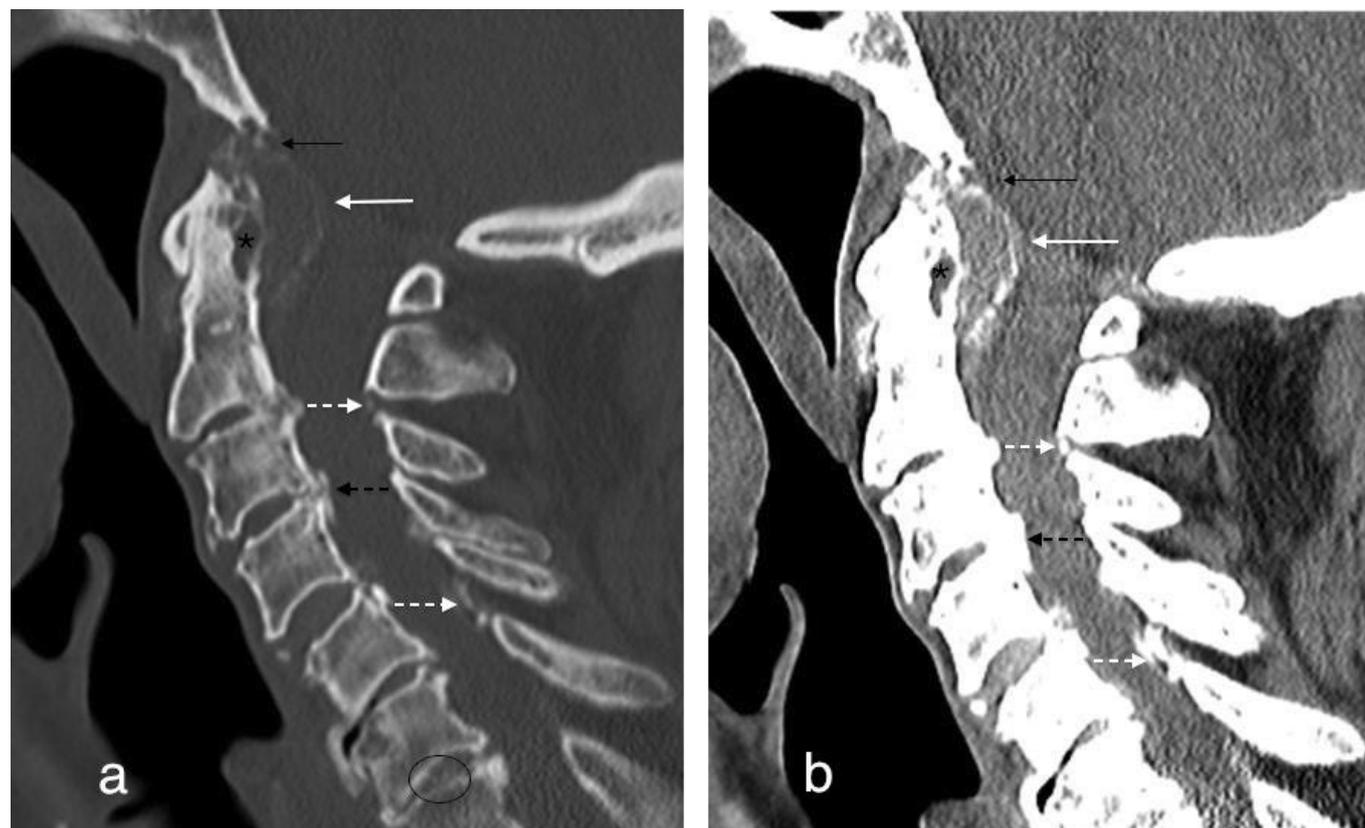


Figure 2: 87 year old male with Crowned Dens Syndrome. Sagittal reconstructed CT images of the cervical spine in bone (a) and soft tissue (b) windows demonstrate erosion and posterior remodeling of the dens (asterisk) with a peripherally calcified soft tissue mass in a "halo or crown-like" appearance (solid white arrows). The inferior portion of the clivus is destroyed (solid black arrows) indicating mass effect. Calcium pyrophosphate dihydrate deposition of the spine can be seen within the ligamentum flavum at C2-3 and C5-6 (dashed white arrows), posterior longitudinal ligament at C3-4 (dashed black arrows), and within the C6-7 intervertebral disc (black circle). Protocol: Siemens CT, mAs 376, KV 120, 3 mm slice thickness, without contrast.

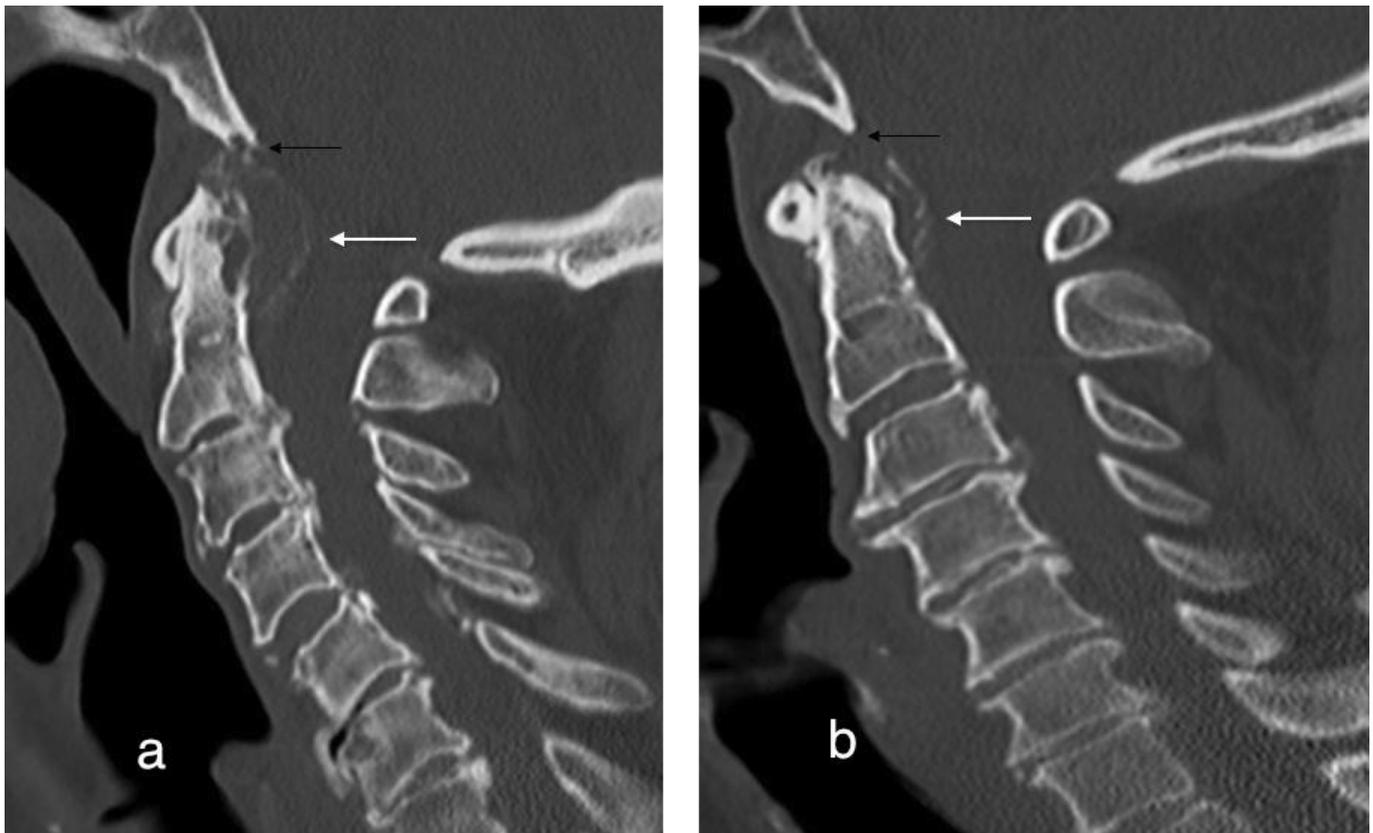


Figure 3: (a) 87 year old male with Crowned Dens Syndrome. Sagittal reconstructed CT images of the cervical spine showing clivus destruction (black arrow). (b) 65 year old asymptomatic female. Comparison case of crowned dens at our institution showing intact clivus (black arrow). Peripherally calcified soft tissue mass in a "halo or crown-like" appearance about the dens in both patients (white arrows). Protocol for both: Siemens CT, mAs 376, KV 120, 3 mm slice thickness, without contrast

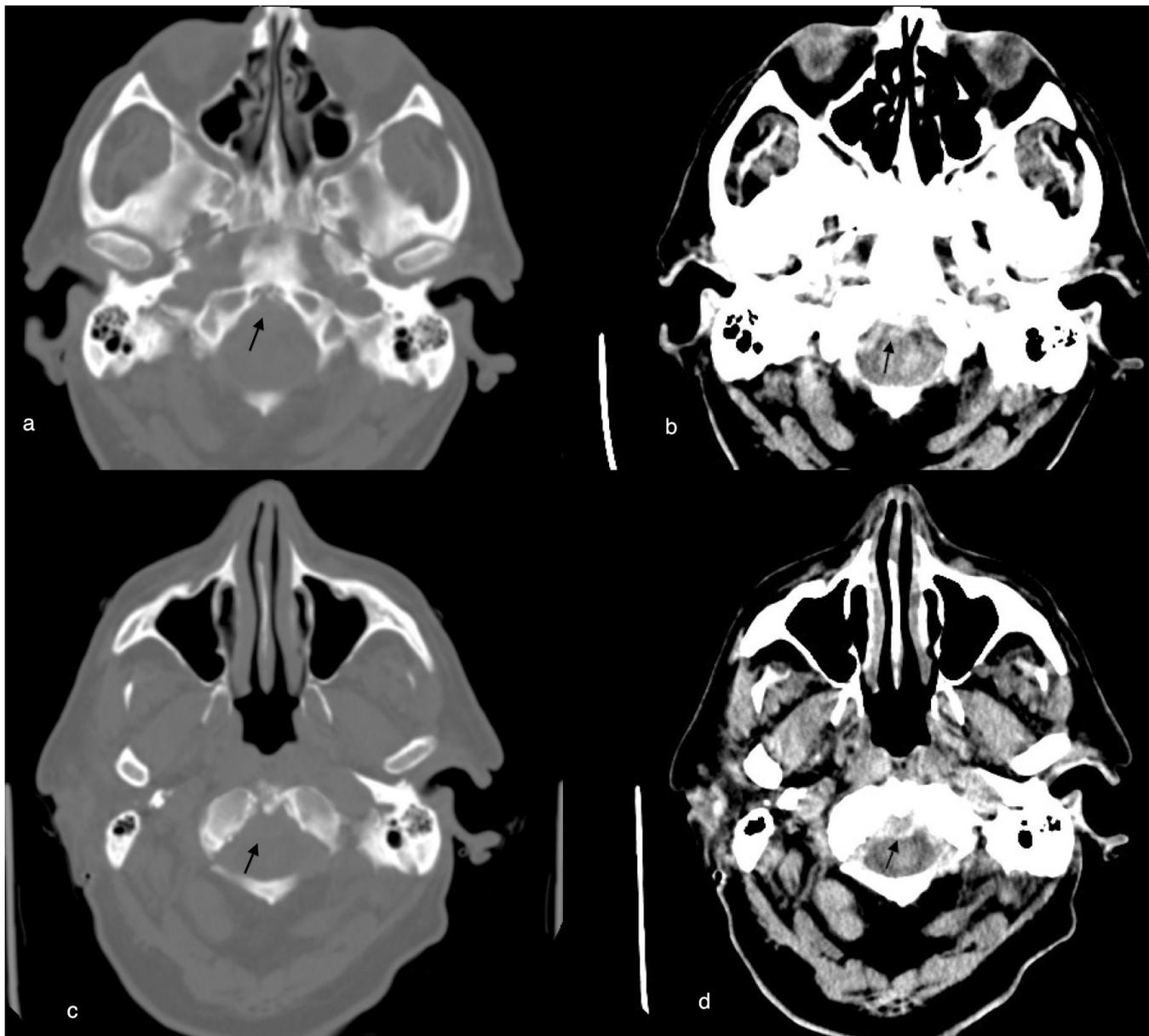


Figure 4: Axial images of the brain in bone (a) and soft tissue (b) windows of the same 87 year old male patient with Crowned Dens Syndrome taken 8 years prior to the current presentation. Axial images of the brain in bone (c) and soft tissue (d) windows taken 6 years prior to the current presentation. Images demonstrate a lesser degree of erosion of the dens with associated calcified soft tissue component (black arrows). A mild increase in size of the 6 year relative to the 8 year study can be seen. These findings indicate the chronic slow growing nature of the lesion. Protocol for both: Siemens CT, mAs 370, KV 120, 4.8 mm slice thickness, without contrast.

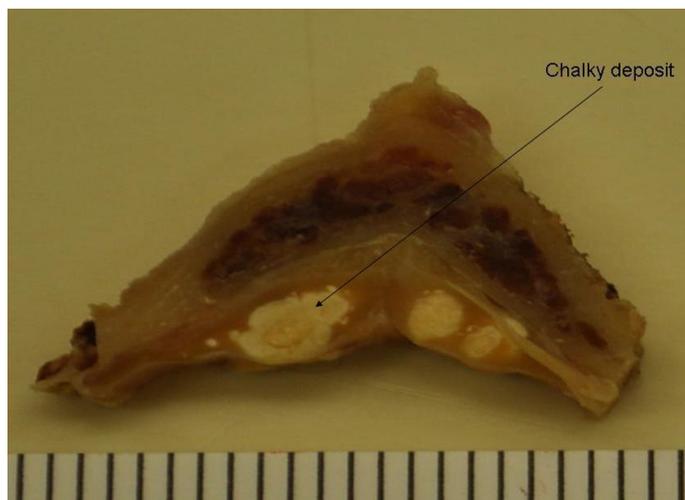


Figure 5 (left): 87 year old male with Crowned Dens Syndrome. Photograph of the cut section of the surgical specimen from the posterior cervical elements showing the chalky yellow-white deposits of calcific material (arrow).

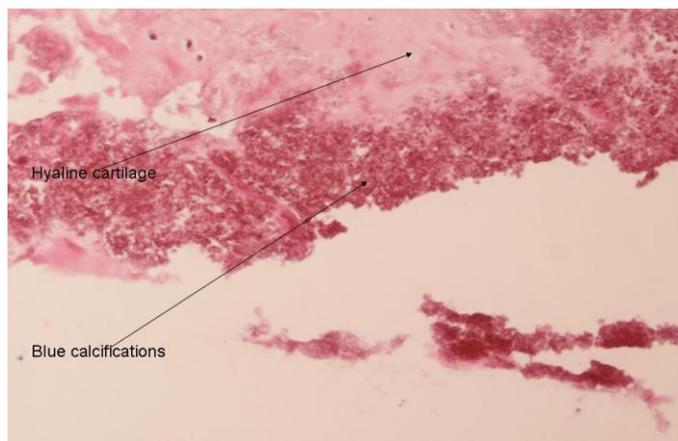


Figure 6: 87 year old male with Crowned Dens Syndrome. Microscopic evaluation of the surgical specimen demonstrates hyaline cartilage (labeled arrow) with dark blue calcifications (labeled arrow) representing chondrocalcinosis.

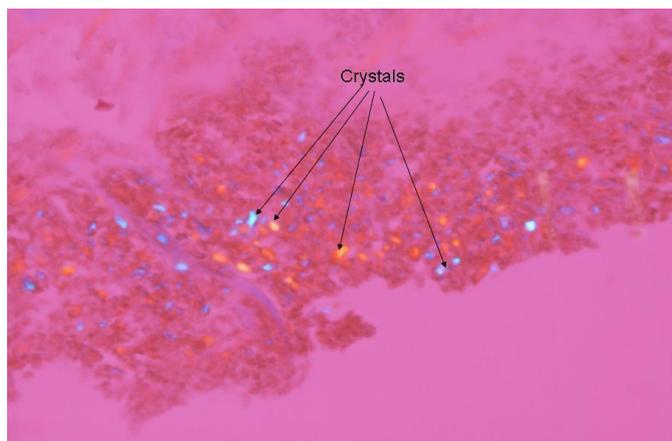


Figure 7: 87 year old male with Crowned Dens Syndrome. Microphotograph (200X) of the surgical specimen under polarized light showing rhomboid crystals (arrows) that were positively birefringent.

Etiology	Calcium pyrophosphate deposition in and around the dens of C2 [1]
Incidence	Very low worldwide, 40/2023 in Japan[15]
Age	Two thirds of patients are greater than 70 years old [15]
Male/Female Ratio	0.6 [15]
Risk Factors	Genetics, Hyperparathyroidism, Hemochromatosis
Treatment	NSAIDS, Colchicine, Steroids
Prognosis	No cure, but majority of patients recover within one week of high dose NSAID or steroid therapy [15]
Imaging Findings on CT	<ul style="list-style-type: none"> -Calcium deposition in and around the dens in a “halo or crown-like” pattern -Erosions and cystic change of the dens -Soft tissue inflammatory component posterior to the dens, which may cause mass effect on the spinal cord or osseous structures -Chondrocalcinosis of the disks and facet joints

Table 1: Summary table of Crowned Dens Syndrome

Entity	Xray	CT	Pathology
Crowned Dens Syndrome	<ul style="list-style-type: none"> • Degenerative disk disease • Chondrocalcinosis 	<ul style="list-style-type: none"> • Calcium deposition around the dens • Erosions and cystic change of the dens • Chondrocalcinosis of the disks and facets 	<ul style="list-style-type: none"> • Calcium pyrophosphate deposition • Rhomboid crystals • Weakly positive birefringent crystals
Rheumatoid Arthritis	<ul style="list-style-type: none"> • Purely erosive • Osteopenia • Predental space widening 	<ul style="list-style-type: none"> • Pannus around dens • C1-2 subluxation • Erosions of facets 	<ul style="list-style-type: none"> • Thick synovium containing plasma cells • Lymphocytes and Multinucleated Giant cells
Chordoma	<ul style="list-style-type: none"> • Well circumscribed mass in the central clivus 	<ul style="list-style-type: none"> • Central clival soft tissue mass • Lytic bone destruction 	<ul style="list-style-type: none"> • Semitranslucent • Gelatinous • Physaliphorous cells
Chondrosarcoma	<ul style="list-style-type: none"> • Mass at the petro-occipital fissure • Usually greater than 3cm 	<ul style="list-style-type: none"> • Chondroid Calcium in matrix • Sharp transition zone • Dense soft tissue 	<ul style="list-style-type: none"> • Smooth and gray • Pleomorphic chondrocytes • Multinucleated Giant cells
Meningioma	<ul style="list-style-type: none"> • Extra axial soft tissue mass arising from the clivus 	<ul style="list-style-type: none"> • Isodense on noncontrast • Homogeneous enhancement with contrast • Hyperostosis 	<ul style="list-style-type: none"> • Firm with dural attachment • Psammoma type is most common • Arises from arachnoid "cap" cells
PVNS	<ul style="list-style-type: none"> • Usually monoarticular • Effusion with subchondral cysts 	<ul style="list-style-type: none"> • Subchondral cysts, • Effusion • Soft tissue mass with no calcification 	<ul style="list-style-type: none"> • Frond-like proliferation of the synovial membrane • Joint fluid of dark blood • Hemosiderin laden macrophages • Multinucleated Giant cells

Table 2: Differential diagnosis of Crowned Dens Syndrome

ABBREVIATIONS

ANKH - ankylosis progressive homolog (mouse)
 C1 - Cervical vertebral body 1
 C2 - Cervical vertebral body 2
 CDS - Crowned Dens Syndrome
 CPPD - Calcium pyrophosphate dihydrate
 CT - Computed Tomography
 IV - intravenous
 MRI - Magnetic resonance imaging
 NSAIDS - Nonsteroidal ant-inflammatory drugs
 PEA - Pulseless electrical activity
 PVNS - Pigmented Villonodular Synovitis
 Xray - Plain radiographs

KEYWORDS

Calcium pyrophosphate dihydrate deposition disease; dens; clivus

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