

Crowned dens syndrome: an uncommon cause of cord compression

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A 65-year-old man presented to our hospital in June 2015 with a 2-week history of neck pain and progressive weakness in four limbs. There was no recent trauma history. He had a history of cervical myelopathy with decompression performed in 2011. On physical examination, an old scar on his neck was unremarkable with no signs of infection. Neurological examination revealed generalised weakness in all four limbs, more marked in bilateral upper limbs. All limbs were hypertonic with hyperreflexia. There was no sensory loss. His C-reactive protein level was elevated to 39.6 mg/L, white blood cell count was also elevated to $14.8 \times 10^9/L$. His cervical radiograph showed indistinct dens (Fig 1). No abnormal soft tissue thickening was seen. Screws of the previous posterior cervical decompression were in-situ. Computed tomography was performed and revealed erosion of the dens and some mildly hyperdense periodontoid soft tissue (Fig 2). Further study with magnetic resonance imaging showed T1 intermediate, T2 heterogeneously hypointense periodontoid soft tissue with patchy enhancement (Fig 3). The cervicomedullary junction was moderately compressed with internal T2 hyperintense cord signal, signifying cord oedema or myelomalacia. Radiograph of other joints found chondrocalcinosis in the triangular fibrocartilage of the right wrist, which is also a common manifestation of calcium pyrophosphate dihydrate (CPPD) crystal deposition disease (Fig 4). Overall features were compatible with crowned dens syndrome.

Crowned dens syndrome was first described by Bouvet et al in 1985.¹ It is a rare entity that presents clinically with severe upper neck pain and radiologically a crown-like density surrounding the odontoid process caused by deposition of CPPD crystals, which is now more commonly described, or hydroxyapatite (HA).¹ It is more common in elderly patients with no history of trauma. Increased inflammatory indicators, such as an elevated C-reactive protein, are usually seen.² Diagnosis is not easy as crowned dens syndrome can mimic a wide range of diseases such as meningitis, osteomyelitis, degenerative cervical spine disease, ankylosing spondylitis, gout, rheumatoid arthritis, temporal arteritis, metastatic bone disease, and spinal tumours.³ Computed tomography is the gold standard for identifying crowned dens syndrome, as it is able to depict the shape and site of calcification

and any bone erosions. Radiography of other joints (wrist, knee, pubic symphysis) may help to ascertain whether the disease is due to CPPD or HA crystals,

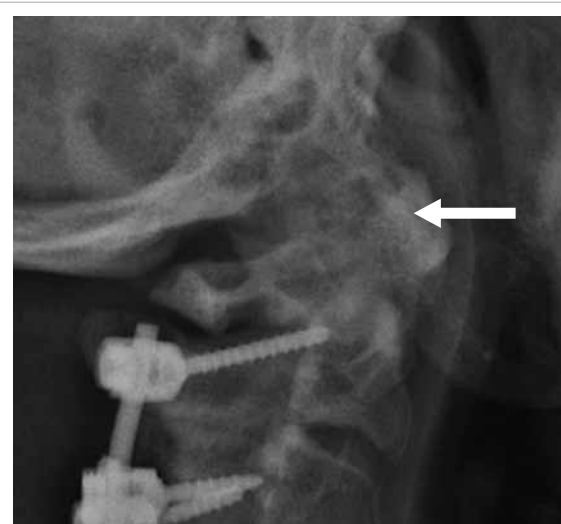


FIG 1. Lateral cervical spine radiograph demonstrating indistinct dens (arrow)

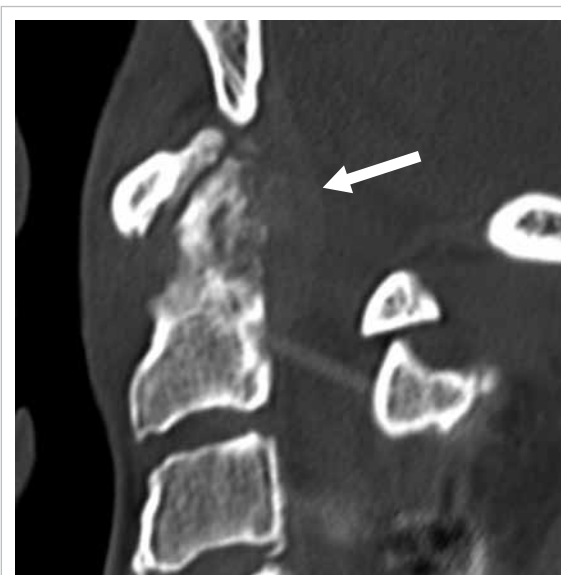


FIG 2. Sagittal reformatted of cervical spine computed tomographic scan revealing erosion of dens and hyperdense periodontoid soft tissue (arrow)

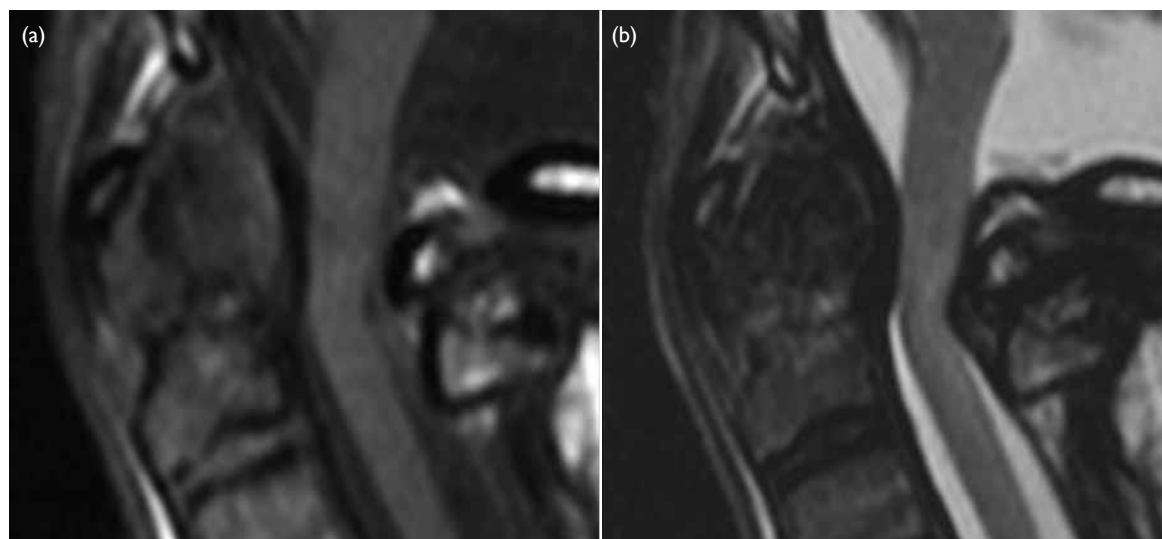


FIG 3. Sagittal (a) T1-weighted and (b) T2-weighted magnetic resonance images showing T1 intermediate, T2 heterogeneously hypointense periodontoid mass with cord compression



FIG 4. Radiograph of other joints found chondrocalcinosis of the triangular fibrocartilage in the right wrist, compatible with calcium pyrophosphate dihydrate crystal deposition disease

and is therefore recommended for routine patient management. Magnetic resonance imaging is indicated for the study of neurological complications

as in our patient.⁴ Prednisolone and non-steroidal anti-inflammatory drugs in combination are the recommended treatment for symptom relief.²

Crowned dens syndrome is an under-recognised disease. Familiarity with the clinical and radiological features will help doctors provide prompt and effective treatment.

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