29. CROWNED DENS SYNDROME – PSEUDO-RHEUMATOID OF THE ODONTOID PEG PRESENTING AS ACUTE INFECTIVE DISCITIS

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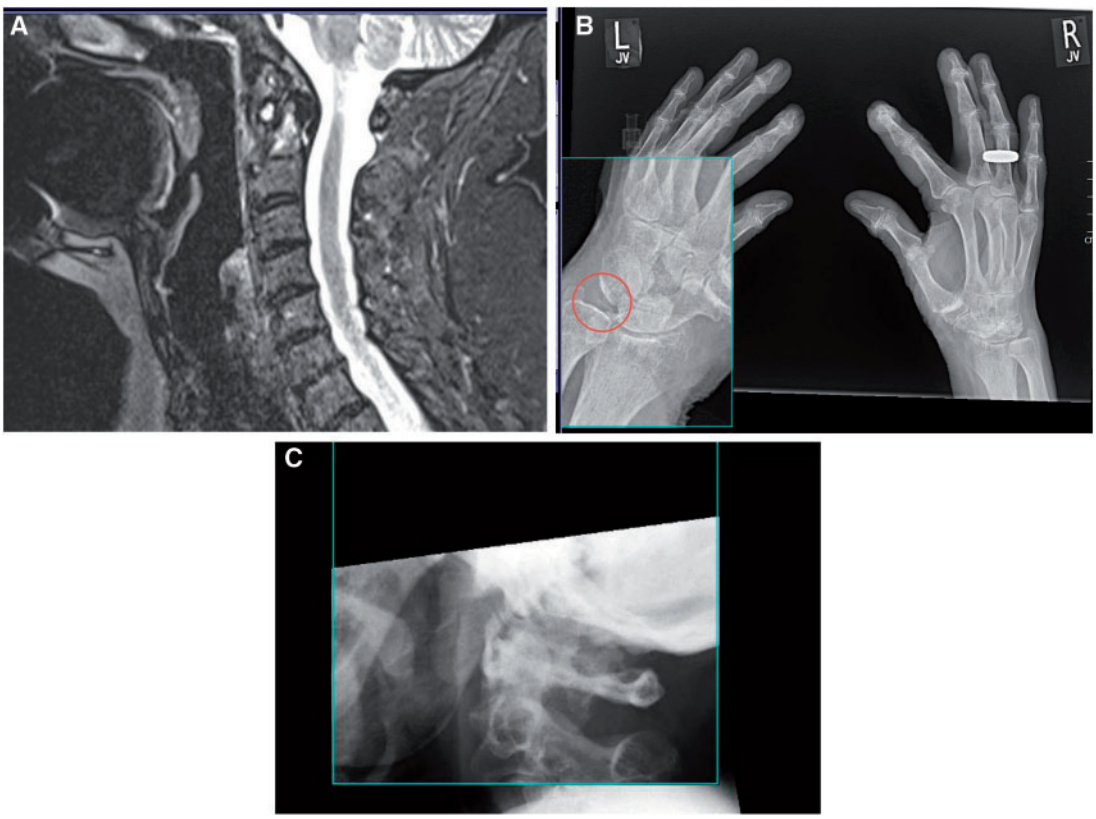
Introduction: We present a case of crowned dens syndrome, calcium pyrophosphate dihydrate (CPPD) deposition of the odontoid peg. The circumstances of this case led to the patient being misdiagnosed with acute discitis on clinical grounds, and rheumatoid arthritis on imaging.

Case description: A 76 year old lady presented to the Emergency Department with fever, rigors and acute onset of occipital headache radiating to her left ear, and associated with neck stiffness. She denied any trauma to the head or neck. Initial blood tests revealed C-reactive protein 51mg/L, WCC 19.5 x 109/L, and coagulase-negative Staphylococcus was isolated from 1 of 2 blood culture bottles. She was commenced on intravenous flucloxacillin for suspected cervical spondylodiscitis, and transferred to an Orthopaedic ward. On day 2 of her admission urinalysis showed leukocytes and blood, although culture was negative. A chest X-ray was normal, and there were no other obvious sources of infection. A magnetic resonance (MR) scan was performed on day 4 of her inpatient stay and showed high STIR signal in the bone of the odontoid peg, and the adjacent anterior arch of C1 (see Fig. 1A). There was no evidence of fluid collection but some mild soft tissue thickening and oedema, reported as in keeping with rheumatoid arthritis (RA). The transverse ligament was intact with no spinal instability. This prompted a request for rheumatology opinion. RA serology was requested and rheumatoid factor was found to be 28 U/mL, and cyclic citrullinated peptide antibodies were negative. On assessment by the rheumatology team, the lady described a history of longstanding degenerative arthritis of multiple joints, including the hands and wrists, as well as longstanding cervical spondylosis. Both of her knees had been replaced. She had experienced acute flares of neck pain with stiffness in the past but overall her history was not suggestive of RA. The patient demonstrated an excellent clinical response to antibiotics. Considering the positive blood culture, it was felt that septic arthritis of the atlantoaxial articulation was still a likely diagnosis, although a lack of oedema and effusion on MR were atypical even allowing for partial treatment at the time of the scan, hence the patient remained on antibiotics. After reviewing a subsequent cervical MR scan on day 8 of her admission, it was concluded that a coincident (probably urinary tract) infection was the cause of her systemic symptoms and this had potentially triggered a flare of local inflammatory arthropathy. With radiographic chondrocalcinosis and severe osteoarthritis elsewhere (see Fig. 1B), and in the absence of clinical or laboratory evidence of RA, the diagnosis of atlantoaxial calcium pyrophosphate arthropathy was made. This lady's neck symptoms resolved within a matter of days and colchicine was recommended for recurrences. Inflammatory markers improved, antibiotics were stopped and she was discharged from hospital. She remains systemically well but continues to experience occasional flares of neck pain and utilises topical non-steroidal gel as required.

Discussion: Spinal involvement is well described in RA and the spondyloarthritides, but is probably more common than previously reported amongst patients with calcium pyrophosphate dihydrate (CPPD) arthropathy. Crowned dens syndrome can often be identified as periodontoid calcification on plain radiography of the cervical spine, although, as in this case, plain radiography identified only a degenerative atlantoaxial articulation (see Fig. 1C). Erosions are sometimes seen, as well as incidental calcification identified on computed tomography (CT) scans performed to exclude fracture in elderly patients presenting with acute neck pain. In this case, as CT scan was not included in the initial diagnostic workup, diagnosis of crowned dens syndrome was probably delayed. Early identification of the diagnosis can prevent unnecessary prolonged inpatient intravenous antibiotic therapy. A clue to the diagnosis in this case was the presence of advanced osteoarthritis and chondrocalcinosis on X-ray of peripheral joints (see Fig. 1B). Finally, while crowned dens

FIG. 1 (**A**



syndrome can lead to elevated inflammatory markers in its own right, flares of CPPD arthropathy may be triggered by systemic infection. We consider this to have been the case in this patient with a probable urinary tract infection.

Key learning points: 1) CPPD deposition of the odontoid peg (crowned dens syndrome) is characterised by atraumatic episodes of cervical pain and stiffness, particularly on rotation, and can be misdiagnosed as fracture, local or meningeal infection, and neoplasm. 2) Plain film appearances can often suggest the diagnosis of crowned dens syndrome, although further imaging is usually required exclude important differential diagnoses. The identification of calcification, for example of the dens or the transverse and alar ligaments, on computed tomography is a typical finding. 3) Early recognition of the clinical features of crowned dens syndrome can prevent over-investigation, for example with lumbar puncture or biopsy, as well as over-treatment for suspected bacterial or viral central nervous system, or musculoskeletal infections. 4) Treatment of crowned dens syndrome is usually with non-steroidal anti-inflammatories unless contraindicated, and our experience suggests that topical therapy can be effective. Successful treatment has also been reported with colchicine and corticosteroids.