Crowned dens syndrome: a key differential for acute neck pain in the elderly
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Crowned dens syndrome (CDS) is an uncommon condition caused when calcium pyrophosphate dihydrate (CPPD) deposition occurs in cervical vertebral ligaments. Symptoms include acute neck pain with stiffness mimicking conditions like meningitis, epidural abscess, discitis and polymyalgia rheumatica or giant cell arteritis. It should be considered as a differential diagnosis for acute neck pain in elderly populations who are not septic and have normal neurology. CDS has a good prognosis, and an early diagnosis is essential to avoid extensive invasive investigations and inadvertent treatment.

Case vignette
An 82-year-old man presented with a four-day history of acute atraumatic cervico-occipital pain and stiffness without infective symptoms or neurological deficits. Past medical history included minimally invasive parathyroidectomy for primary hyperparathyroidism, ischaemic heart disease, hypertension, epilepsy and solitary lung nodule. Medications included alendronic acid, sodium valproate, atorvastatin, aspirin, cilazapril and tamsulosin.

Examination was pertinent for severe limitation in neck movements in all directions, including rotation, flexion and extension. He was afebrile with normal neurological examination. Blood results showed persistently normal white cell count with significantly elevated C-reactive protein (CRP) at 252mg/L. Renal function and electrolytes were normal, apart from low serum phosphate of 0.51mmol/L.

Initial treatment consisted of empirical ceftriaxone and patient-controlled analgesia (PCA) pump for pain management. Peripheral blood cultures, urine analysis, cerebrospinal fluid analysis and chest radiograph were negative for sepsis. Cervical spine x-ray showed facet joint osteoarthritis. Concurrently performed computed tomography (CT) and magnetic resonance imaging (MRI) of cervical spine ruled out infection, abscess or malignancy. A close review of CT cervical spine revealed calcified transverse ligament around dens (Figures 1 and 2). Review of old plain radiographs of knee and hip joints confirmed chondrocalcinosis. A diagnosis of crowned dens syndrome was made in the context of clinical, laboratory and radiological findings.

Within a week of commencing colchicine and tapering dose of prednisone, symptoms resolved completely and the CRP normalised. Serum phosphate returned to normal without supplementation.

Discussion
Crowned dens syndrome was described first in 1985 by Bouvet et al. It may be detected in up to 2% of patients over 70 years of age who present with acute neck pain. Typical presentation is with acute severe neck pain associated with neck stiffness and a significant restriction of cervical movements and occasional fever, and the inflammatory markers are often elevated. Neurological complications are rare; however, large cervical CPPD deposits may result in spinal stenosis or cervical myelopathy. Ageing is a major risk factor, and others include haemochromatosis, primary hyperparathyroidism, hypophosphatemia, hypomagnesaemia, hereditary predisposition to CPPD and use of oral bisphosphonates. The gold standard investigation for the diagnosis of CDS is CT scan because plain radiographs fail to detect the periodontoid calcifications. CPPD depo-
sition may be located posterior only (50%), posterolateral (27.5%), circular (12.5%), lateral (5%) or anterior (5%) to the odontoid process, and findings may persist for about three months after symptom relief.\textsuperscript{9}

Integrated single photon emission tomography (SPECT) scan has been performed to evaluate target sites for facet joint injection which showed avidly increased uptake at the anterior border of the dens and calcification of the transverse ligament of the atlas.\textsuperscript{10}

The majority of patients with CDS fully recover within a week of treatment with non-steroidal anti-inflammatory drugs (NSAIDs), corticosteroid, colchicine or combination therapy.\textsuperscript{3,11} There is some evidence of a role for colchicine in the prevention of both acute attacks of pseudogout and recurrent episodes of the other CPPD-related arthritides.\textsuperscript{12} In rare cases, surgical decompression and stabilisation have been considered for central cord syndrome.\textsuperscript{13}
**Figure 1:** CT cervical spine in axial plane–transverse ligament calcification and crystal deposition.

**Figure 2:** CT cervical spine in coronal plane–transverse ligament calcification and crystal deposition.
Competing interests:
Nil.

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