Title

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CLINICAL VIGNETTE

A Pain in the Neck: An Unusual Presentation of Calcium Pyrophosphate Dihydrate Deposition Disease (CPPD)

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Introduction

CPPD is characterized by precipitation of calcium pyrophosphate dihydrate crystals in joints and surrounding connective tissues. It can often mimic and at times co-exist with other arthropathies including gout, osteoarthritis, and rheumatoid arthritis. Calcium pyrophosphate dihydrate arthritis often initially presents in the knee joint, but also commonly involves wrists, shoulders, ankles, feet and elbows. Crowned Dens Syndrome (CDS) is a rare subtype of CCPD, characterized by crystal deposition in the joints of the spine. CDS typically manifests as axial neck pain associated with stiffness of the neck and shoulder girdle, occasionally accompanied by fever and headache.

Case Presentation

A 71-year-old male with a history of paroxysmal atrial fibrillation on anticoagulation, heart failure with preserved ejection fraction and obstructive sleep apnea/obesity hypoventilation syndrome presented to the emergency department with melanic stools, epigastric and joint pain. The patient's gastrointestinal bleeding resolved, and he was treated for esophagitis and gastritis seen on upper endoscopy. Further history revealed that the patient had been experiencing persistent diffuse joint pain and stiffness that began approximately 1 week prior to admission. Months prior to presentation, he reported similar but less severe episodes of joint pain which remitted with the use of analgesics.

Prior to his admission, the patient was functionally independent in all activities of daily living (ADLs) and instrumental ADLs. He had worked as a roofer in the past without limitation. In the week prior to admission, his pain and stiffness had become so severe that the patient could no longer perform his ADL's independently. On exam, the patient had synovial swelling and pain in several of his bilateral metacarpophalangeal joints (MCP's) and proximal interphalangeal joints (PIP's) as well as in his wrists, bilateral elbows and right olecranon bursa. His knees were warm and demonstrated the presence of an effusion. Bilateral shoulders and hips were tender to palpation. Examination of the cervical spine demonstrated severe tenderness to palpation with marked limitation in both active and passive flexion, extension, and rotation.

Labs were significant for a c-reactive protein of 15 and an erythrocyte sedimentation rate greater than 120. Rheumatoid factor (RF), anti-citrullinated peptide (anti-CCP), double stranded DNA (dsDNA), antibodies to Ro (SSA), antibodies to La (SSB), anti-topoisomerase I (Scl-70) and HLA-B5801 were all within normal limits. Aspiration of the knee joint demonstrated inflammatory synovial fluid with 25,100 nucleated cells with 96% segmented neutrophils and the presence monosodium urate crystals. A CT of the cervical spine demonstrated mineralization around the dens process involving the intervertebral discs, along with multilevel degenerative changes, suggesting a diagnosis of crowned dens syndrome (Figures 1 and 2).

Systemic corticosteroids, allopurinol and colchicine were initiated with marked improvement in the patient's pain and range of motion. However, given his persistent deficits in mobility and self-care, he required transfer to an acute inpatient rehabilitation hospital. He is now functionally independent with his ADL's and living back at home.

Discussion

CDS should be suspected when a patient presents with symptoms of inflammatory arthritis affecting both the peripheral and axial skeleton. Both acute CPPD and acute gout typically present in peripheral joints such as the knee, wrist, and shoulder, are often monoarticular, and are highly inflammatory. Axial joint involvement in gout is rare and predominantly affects the lumbar spine or sacroiliac joints while CPP crystal precipitation occurs in the cervical spine, as in this case of CDS.[1] CDS is commonly underdiagnosed and may mimic other causes of neck pain, fever and headache including polymyalgia rheumatica/temporal arteritis, meningitis, and cervical discitis.[2] Fortunately, CDS can be readily distinguished from other etiologies of neck pain by a CT demonstrating calcification of the periodontal ligaments around the dens process, appearing as a radiopaque "crown" or halo surrounding the dens.[3,4]

While CPP crystals were not observed in this patient's knee synovial fluid, CT cervical spine in conjunction with the clinical picture were consistent with a mixed crystalline

arthropathy. Identification of CPP crystals in synovial fluid analysis is limited by the fact that they are weakly positively birefringent via polarized light microscopy, polymorphic, and smaller than monosodium urate crystals.[1,5]

As this case illustrates, CPPD should be considered when an older patient demonstrates new deficits in mobility and ADL independence concurrent with joint pain and stiffness. CDS is an important cause of severe disability and can lead to spinal instability, thus, early recognition of this pathology is essential to avoid long-term sequela of untreated disease.

NSAIDs are the gold standard treatment of acute CPPD, including CDS. Steroids and colchicine are alternatives. Colchicine has also been used to prophylactically reduce the incidence of flares and accumulation of joint damage.[6]

Figures

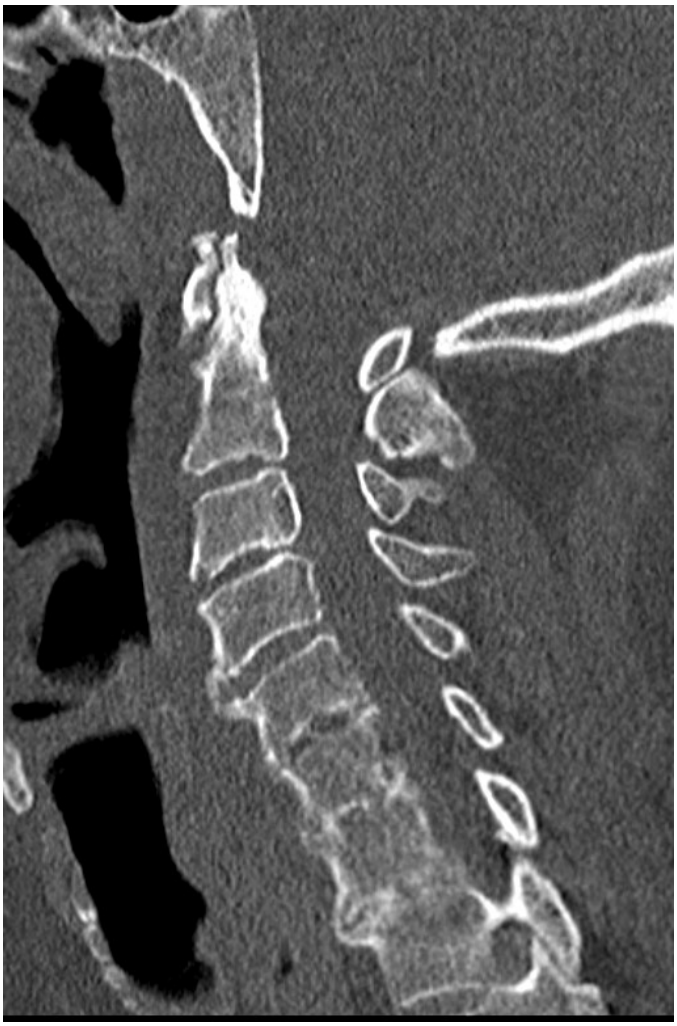

Figure 1: CT C-spine without contrast: chronic erosive process of the tip of the dens process and mineralization of several intervertebral disks

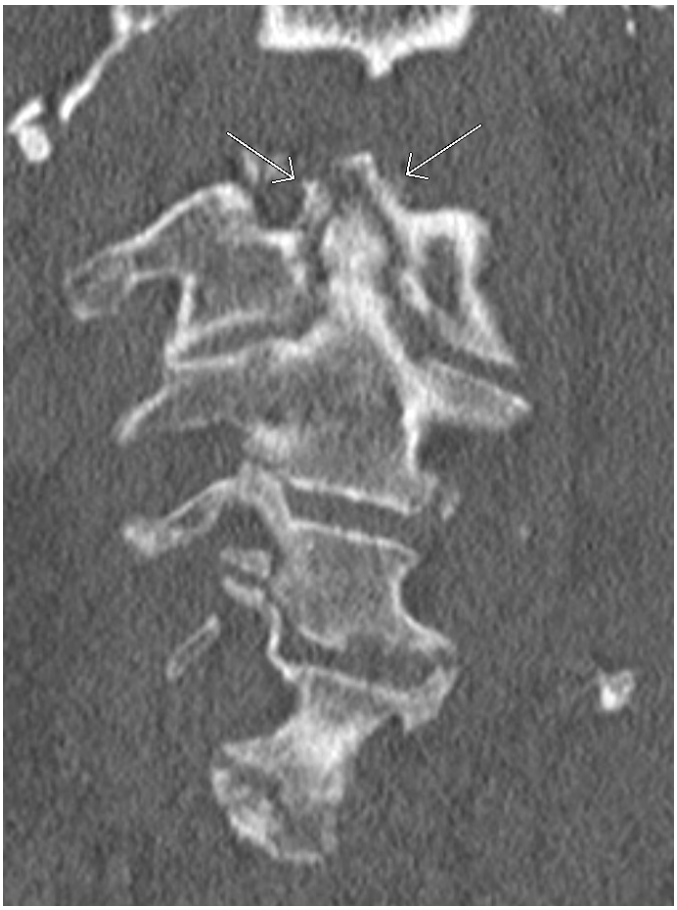

Figure 2: CT C-spine without contrast: mineralization of the ligaments surrounding the dens process consistent with calcium pyrophosphate dihydrate deposition

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