Crowned dens syndrome
An unusual cause of fever and neck stiffness

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Discussion

Calcium pyrophosphate dihydrate (CPPD) crystal deposition disease, also known as pseudogout, has varied clinical presentations, dependent on the anatomical area of crystal deposition (Figures 2 and 3). Rarely, pseudogout involves the atlanto-axial joint of the cervical spine, causing acute severe neck pain and fever with raised inflammatory markers, and, sometimes, altered mental status. This has been called crowned dens syndrome, and is often misdiagnosed as meningitis or cervical discitis, resulting in inappropriate broad-spectrum antimicrobial therapy.1 The diagnosis is best confirmed with unenhanced CT imaging of the upper cervical spine. Optimal treatment is with short-term non-steroidal anti-inflammatory drugs, corticosteroid, or colchicine.2 Early suspicion and diagnosis leads to avoidance of unnecessary further testing, invasive procedures like lumbar puncture, and inappropriate antimicrobial treatment in a population at high risk of Clostridium difficile-associated disease.

References

Case

An 81-year-old gentleman presented with a 4-day history of acute neck pain and fever. Examination revealed fever at 38.2 °C with marked neck stiffness and limitation of movement due to pain. Serum C-reactive protein concentration was raised at 50 mg/L. Unenhanced computed tomographic (CT) imaging of the cervical spine was performed, which confirmed the clinical suspicion of crowned dens syndrome (Figure 1). The patient improved dramatically with a course of oral colchicine and was subsequently discharged.

Figure 1. Unenhanced coronal (left) and axial (right) computed tomographic images of cervical spine, showing crown-shaped calcium deposits surrounding the odontoid process (blue arrow), and curvilinear calcification in the transverse ligament of the atlas (blue arrowhead).

Figure 2. Pathogenesis of calcium pyrophosphate (CPP) crystal deposition. Articular cartilage vesicles concentrates substrates, enzymes, and transporters necessary for CPP crystal formation. AMP Adenosine monophosphate; ATP Adenosine triphosphate; ANK Progressive ankylosis gene product; CPPD Calcium pyrophosphate deposition disease; PPI Pyrophosphate

Figure 3. Typical joint involvement with CPPD.

Consider crowned dens syndrome in older patients presenting with fever and neck pain and who have raised inflammatory markers.

Unenhanced cervical spine CT is the radiologic investigation of choice.

Early diagnosis prevents unnecessary invasive testing and broad-spectrum IV antimicrobial therapy in this vulnerable population.

Figure 2

Very common
Common

Articular cartilage vesicle

Adenosine + Pi

AMP

ATP

PPi

ANK

Progressive ankylosis gene product

CPPD crystals

CPP crystal says

Print A0 size
Laminated
Thanks!