

Acute Pseudogout of the Neck: "Crowned Dens" Revisited

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Calcium pyrophosphate dihydrate crystal deposition disease is associated with an acute mono- or pauci-arthritis, termed "pseudogout" in elderly patients, involving a large joint (including the knees, ankles) or a chronic arthropathy manifesting as mild joint pain and stiffness. Pseudogout is a crystal-deposition disease of peripheral joints, usually encountered in elderly patients. However, acute presentation of pseudogout around the odontoid process comprises a "crowned-dens" appearance, and requires contemplation of differential diagnoses. We recently experienced a case of pseudogout in the cervical spine presenting with fever and acute neck pain that was successfully treated with a colchicine and low-dose oral steroid. We reported this case with a review of the relevant literature. (*J Rheum Dis* 2016;23:122-124)

Key Words. Pseudogout, Cervical

INTRODUCTION

Pseudogout is a common cause of inflammatory arthritis in old age that is characterized by the calcification of articular tissues due to calcium pyrophosphate dihydrate (CPPD) crystal deposition [1,2]. It can present as acute, monoarticular arthritis resembling pyogenic arthritis, or can show chronic polyarticular involvement. CPPD crystal usually deposits on the knee, ankle, elbow and rarely temporomandibular joint or ligamentum flavum of spine.

Recently, we experienced a perplexing case of pseudogout in cervical spine mimicking an infectious spondylitis or osteomyelitis, presenting with fever and acute neck pain that was successfully treated with a colchicine and low-dose oral steroid. We reported this case with a review of the relevant literature.

CASE REPORT

A 72 year-old female with previously known knee osteoarthritis was admitted to the department of infectious diseases with complaints of spiking fever for 2 days and

severe pain in the posterior cervical area. Blood pressure was normal, pulse rate was 90 beats/min and body temperature was 38.8°C.

On physical examination, she had tenderness on the posterior neck, and a deformed, mildly swollen left knee on palpation. Lung sound was clear and there was no significant finding on physical examination of the abdomen. After initiating broad-spectrum antibiotics for indolent infectious condition, we conducted an intensive search for the causative disease of the fever and several differential diagnoses were considered, including infectious spondylitis and systemic rheumatic diseases.

The patient had leukocytosis (16,300/mm³), mild thrombocytosis (476,000/mm³), a markedly elevated C-reactive protein level (132.5 mg/L; normal <5 mg/L) and an elevated erythrocyte sedimentation rate (64 mm/h). Serum rheumatoid factor was negative and serum uric acid level was within normal range (4.3 mg/dL; normal, 4 to 7 mg/dL). Blood cultures showed no growth of organism. Abdomen, pelvis and chest computed tomography scans showed no abnormal finding.

Computed tomography (CT) of cervical spine area re-

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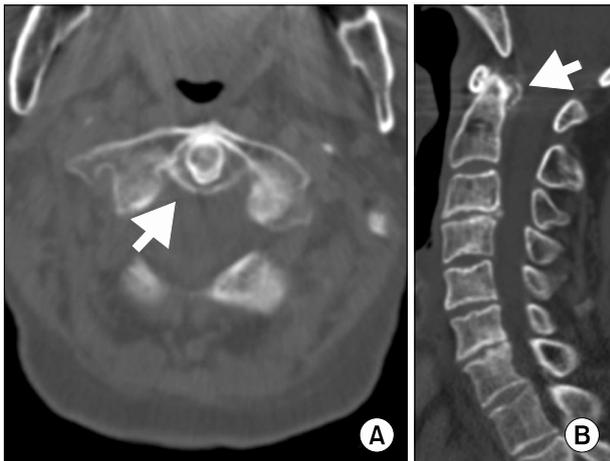


Figure 1. Axial (A) and sagittal (B) image of the cervical computed tomography scan at the C1/C2 level shows curvilinear calcifications of the transverse ligament (arrows).

vealed curvilinear calcifications of the transverse ligament at the C1~2 level, i.e., “crowned dens” appearance (Figure 1A and 1B). To identify bone marrow edema, soft-tissue change, and to exclude the infectious spinal pathology, magnetic resonance imaging (MRI) was done. Sagittal fat-suppressed T2-weighted, and T1-weighted gadolinium enhanced MRI showed a soft tissue edema and diffuse enhancement around the dens, suggesting inflammatory change (Figure 2A and 2B).

On the second day of admission, the patient complained of aggravating pain on the left knee joint. An arthrocentesis of the knee joint showed a yellowish aspirate, with 9,000 white cells per cubic millimeter (85% polymorphonuclear leukocytes, 20% lymphocytes). No organism was seen in the Gram’s stain and cultures were sterile. Rhomboid shape crystal deposition was identified under polarizing light microscopic examination of the joint aspirate (Figure 3). Typical chondrocalcinosis features were absent on plain radiograph of the knee joint.

Finally, the patient was diagnosed with pseudogout of the cervical spine and knee joint. Antibiotics were discontinued and colchicine (100 mg, twice a day, per oral) plus non-steroidal anti-inflammatory agent (NSAID; nabumetone 500 mg, twice a day, per oral) and 5 mg prednisolone was administered. After 3 days of anti-inflammatory treatment, fever and neck pain subsided. Acute phase reactant returned to normal in 1 week, and the patient has been visiting rheumatology clinic with intermittent administration of NSAID.



Figure 2. Sagittal fat-suppressed T2-weighted (A), and T1-weighted gadolinium enhanced (B) magnetic resonance image shows a soft tissue edema and diffuse enhancement around the dens, suggesting inflammatory change.

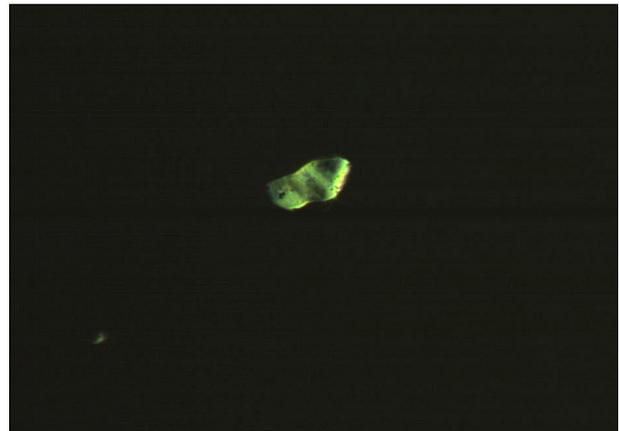


Figure 3. By the aspiration of synovial effusion of left knee joint, square, rhomboid-shaped crystal was founded under polarized light microscope ($\times 200$).

DISCUSSION

Pseudogout of the cervical spine also known as “crowned dens syndrome (CDS)”, was first described in 1985 by Bouvet et al. [3]. It is a clinical-radiologic condition with crystal deposits within the fibers of the transverse ligament of the atlas. Similar to peripheral joint CPPD diseases, the crystal-induced inflammatory process causes local and systemic symptoms [4-6].

CDS usually affects elderly women more than 60 years of age. Prevalence is around 2% among cases of acute

neck pain. While up to 70% of peripheral CPPD deposition disease may have radiographic evidence of cervical spine CPPD deposition, the majority are asymptomatic [7].

Diagnosis of CDS is usually based on the identification of periodontoid calcification on CT scan. CT is more sensitive and accurate than conventional plain radiography in detecting calcific deposits and bony change. Up to 90% of CT imaging comprises posterior, posterolateral, or circular periodontoid calcification, configuring the “crowned dens” appearance [8]. Plain radiography is often not helpful.

In differential diagnosis of CDS, MRI is useful to exclude crucial differentials like infectious spondylitis, discitis, myelopathy and rare case of malignancy [9]. Histopathologic examination of periodontoid tissue or peripheral joint fluid aspiration, are helpful but not essential.

In case of this patient, acute neck pain with 2 days of unexplained fever was the main complaint. Though the crowned-dens appearance was initially identified by conventional CT scan, the cause of fever could be investigated by MRI imaging, and consequently prompt administration of colchicine with concomitant use of NSAID resulted in prompt resolution of symptoms.

Administration of NSAID and/or low dose glucocorticoid is the mainstay of medical treatment [10]. Colchicine is effectively used in resistant cases [11]. The prognosis is usually excellent with rapid improvement of clinical symptom and laboratory sign of inflammation. However, surgical decompression is required in cases of atlantoaxial instability, spinal cord compression and neurologic symptom [12].

SUMMARY

In conclusion, CDS is a pseudogout of the cervical spine characterized by a crystal deposition around periodontoid articular tissue resulting in acute neck pain and systemic inflammatory sign. Clinicians should be aware of this underdiagnosed cause of neck pain in elderly patients, and avoid delay in diagnosis with therapeutic intervention.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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