Crowned dens syndrome presenting as pyrexia of unknown origin (PUO)

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Background. Deposition of calcium pyrophosphate crystals in the cervical spine around the odontoid process may lead to neck pain and fever. This condition is called crowned dens syndrome (CDS).

Case report. An 89-year-old female presented with complaints of fever for one-month duration and recent onset neck pain. During her admission, she developed right knee pain with evidence of chondrocalcinosis on X-ray. Considering her clinical presentation in setting of pseudogout, she had a CT scan of her neck that revealed erosion of the dens and hyperdense soft tissue surrounding the odontoid process. Based on her clinical and radiologic presentation, she was diagnosed with crowned dens syndrome and started on NSAIDs. Unfortunately, she did not respond to NSAIDs and was switched to Colchicine, which resulted in immediate improvement in her symptoms.

Conclusions. We present this case to stress the importance of keeping crowned dens syndrome as one of the differentials in an elderly patient presenting with fever and neck pain.

Key words: crowned dens syndrome; fever; elderly.

INTRODUCTION

Calcium pyrophosphate crystal deposition (CPPD) disease affects approximately 4 to 7% of adult population of the United States and is especially common amongst persons of advanced age [1, 2]. Deposition of CPPD crystals in the cervical spine around the odontoid process may lead to neck pain and fever. This condition is called crowned dens syndrome (CDS) and was first described by Bouvet *et al.* in 1985 [3]. We herein describe the case of an 89 year-old female who was admitted to our hospital with recent onset posterior neck pain and persistent fevers for one-month duration that had failed to resolve with broad-spectrum antibiotics. On the 6th day of admission, she was diagnosed as having crowned dens syndrome based on clinical (febrile neck pain), radiologic (CT scan findings of calcification surrounding the odontoid process) and therapeutic findings (drastic improvement in symptoms with colchicine).

CASE REPORT

An 89-year-old female with past medical history significant for multiple deep vein thrombosis (DVTs in both legs and right arm), and inclusion body myositis (on chronic prednisone 5 mg) was admitted to our hospital with complaints of

gradually worsening posterior neck pain since a week and persistent fever for about 1 month duration. She was discharged from the outside hospital on Vancomycin with a peripherally inserted central catheter (PICC) line in-situ in view of suspected infection. At admission, her body temperature was 102 F and neurologic examination significant for stiff cervical spine in all ranges without any evidence of neurological deficit. Her lab work revealed leukocytosis (13,000/mm³) and an elevated CRP of 14.2 mg/L. In addition, X-ray of cervical spine did not show any particular abnormalities.

In addition to the vancomycin she had been receiving, we started her on piperacillin-tazobactam for her persistent fever and NSAIDs for her neck pain, but unfortunately there was no symptomatic improvement. She had a lumbar puncture in view of suspected meningitis, but her CSF examination was unremarkable. In addition, there was no pathogenic growth on blood cultures. Furthermore, the levels of serum pro-calcitonin (0.43 ng/mL) were normal indicating more towards a non-infectious etiology. The lab work for connective tissue disorders or vasculitis, such as rheumatoid factor, anti-cyclic citrullinated peptide antibodies, anti-nuclear, and anti-neutrophil cytoplasmic antibody (ANCA) antibodies were all negative. In an attempt to rule out inclusion body myositis flare (on prednisone for IBM), we measured CPK and Aldolase levels which were within normal limits.

On the 4th day of admission, patient started having pain over the right knee. On physical examination, the right knee was erythematous and tender. She had an X-ray of the right knee, which showed evidence of chondrocalcinosis (Figure 1) (no past history of previously diagnosed CPPD). The levels of serum uric acid were normal. To confirm the diagnosis of CPPD, a tap was attempted but there was not enough synovial fluid.

Figure 1. Chondrocalcinosis in the right knee (as directed by arrows).

Figure 2. CT cervical spine showing erosion of dens and hyper dense soft tissue around the odontoid process (as directed by arrow).

Our patient had a CT scan of the neck on 6th day of admission, which revealed erosion of the dens and hyper-dense soft tissue surrounding the odontoid process (Figure 2). In addition, the cervical

MRI showed hyperintense soft tissue surrounding the odontoid process on T2 weighted imaging, with no evidence of cervical cord compression. A diagnosis of CDS was made according to the clinical (febrile neck pain in association with calcium pyrophosphate crystal deposition in her right knee) and radiologic findings (erosion of dens and hyperdense peri-odontoid soft tissue). She was started on oral colchicine. Patient's fever resolved within 1 day of therapy and neck pain improved within 2 days of therapy. Also, the inflammatory markers (ESR and CRP) returned to within normal range after a week of therapy.

DISCUSSION

Amongst 100 consecutive admissions to an acute geriatric unit, the prevalence of radiographic calcium pyrophosphate deposition was 15% in age group 65-74 years, 36% in age group 75-84 years and almost 50% in individuals > 84 years. Majority of individuals with CPPD deposition are asymptomatic whereas others can present either with acute episodic mono- or oligo-arthritis (pseudogout) involving large joints (knee, ankle or wrists) or with chronic arthropathy [4].

Our case report describes an elderly female presenting with crowned dens syndrome (CDS) as the first presentation of her calcium pyrophosphate deposition disease (CPPD). Our case report is unique for the following reasons: firstly, to the best of our knowledge, this is the first case of crowned dens syndrome presenting as pyrexia of unknown origin (temperature > 101 F lasting more than 3 weeks in duration and failure to reach a diagnosis despite one week of inpatient investigation). Secondly, in most of the cases reported, the symptoms of crowned dens syndrome improved either with NSAIDs or moderate dose steroids. However, our patient did not respond to NSAIDs but responded well to Colchicine with symptomatic relief in 2 days.

The crowned dens syndrome (CDS) is a relatively rare clinical condition which is characterized by fever and severe neck pain; elevated inflammatory markers (CRP, ESR); and crystalline deposits surrounding the odontoid process [5]. Calcium pyrophosphate dehydrate (CPPD) and hydroxyapatite (HA) constitutes the major components of crystalline deposits. CDS should be suspected in individuals with medical history of crystalline deposition disease in other joints [6]. The presence of right knee pain and evidence of chondrocalcinosis prompted us to obtain a CT

cervical spine in order to look for crystal deposition around the odontoid process. Cervical CT scanning focusing on C1 and C2 is the gold standard for diagnosis of CDS whereas X-ray findings are mostly unremarkable [7]. Cervical MRI can support the findings in the evaluation of CDS with evidence of soft tissue hyper-intensity around the odontoid process on T2 weighted imaging.

Because of the clinical symptoms of fever and posterior neck pain, CDS may be misdiagnosed as meningitis, osteomyelitis or cervical metastatic spondylitis in a patient with history of malignancy [8, 9]. CDS can also mimic polymyalgia rheumatica (PMR) or giant cell arteritis if morning stiffness in shoulder girdle or jaw claudication is present in addition to febrile neck pain [8]. In our patient, we performed a lumbar puncture but the CSF findings were unremarkable for meningitis.

To the best of our knowledge, most of the cases on crowned dens syndrome present as neck pain and sudden onset fever lasting for 2-3 days. However, we report a case of CDS presenting as pyrexia of unknown origin. Unfortunately, our patient unnecessarily received antibiotics and had a PICC line in place for more than one-month

duration despite the non-infectious cause behind her clinical presentation. This resulted in high health-care cost, increased predisposition to antibiotic resistance and discomfort from PICC line. Therefore, CDS should be considered as one of the differentials especially in elderly patients with fever and posterior neck pain to prevent unnecessary invasive procedures and antibiotic treatment. Presence of chondrocalcinosis on X-rays, even if the patient is asymptomatic and has no past history of CPPD, may be helpful in making the diagnosis of CDS.

Also, most of the cases have described improvement with NSAIDs. In patients unresponsive to NSAIDs, moderate-dose steroids form an important part of treatment [10]. However, not many cases have illustrated the use of colchicine in patients with crowned dens syndrome. There are further studies needed to demonstrate the efficacy of colchicine in patients with CDS.

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Introducere. Depunerile de cristale de pirofosfat de calciu din jurul proceselor odontoide ale coloanei cervicale pot duce la dureri și febră. Această patologie este cunoscută sub numele de sindromul coroanei dense (crowned dens syndrome-CDS).

Prezentare de caz. O pacientă în vârstă de 89 de ani cu febră și durere cervicală veche de o lună s-a prezentat la spitalul nostru. În timpul internării pacienta a acuzat și dureri la nivelul genunchiului drept cu evidențierea condrocalcinosei pe radiografie. Analiza CT cervical a demonstrat eroziune și hiperdensitatea țesutului moale din jurul procesului odontoid. A fost diagnosticată cu CDS și s-a inițiat tratamentul cu AINS (anti-inflamatoare non-steroidiene). Din păcate pacienta nu a răspuns la tratamentul cu AINS și a fost tratată cu colchicină ceea ce a dus la o îmbunătățire rapidă a simptomatologiei.

Concluzii. Acest caz subliniază importanța excluderii CDS ca diagnostic la pacienții vârstnici cu febră și durere cervicală.

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