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The Spine Journal 15 (2015) e65–e68

THE SPINE JOURNAL

Case Report

The early and late stages of crowned dens syndrome: two case reports

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Received 27 June 2014; revised 21 March 2015; accepted 19 May 2015

Abstract

BACKGROUND CONTEXT: Crowned dens syndrome (CDS) is a rare form of calcium phosphate crystal depositions and often presents with recurrent neck pain, stiffness of neck, increased erythrocyte sedimentation rate, and episodes of fever.
PURPOSE: The goal of this report is to identify the early and late stages of CDS and its consequences as the result of repeated attacks of CDS at cervical spine in its late stage.
STUDY DESIGN: This is a case report.
METHODS: We reported one case of early-stage CDS and one late-stage CDS.
RESULTS: The two patients shared some common clinical features of acute attack of CDS, such as increased erythrocyte sedimentation rate, C-reactive protein, episode of fever, and increased white blood cells along with high blood glucose levels. The first case showed early phase of CDS with computed tomography (CT) scan that only showed mild calcification around the dens. The second case had appearance of late stage of CDS with more severe chronic degenerative changes of cervical spine.
CONCLUSIONS: Early stage of CDS can be difficult to identify because of mild clinical symptoms, but CT scan is a preferable method to demonstrate densities surrounding the top and sides of the odontoid process. In the late stage of CDS, radiographic features often include diffuse periodontoid calcifications, diffuse destructive discopathies, and apophyseal joint destruction, and patient might have severe neurological symptoms.

Keywords: Crowned dens syndrome; Pseudogout; Calcium phosphate crystal depositions (CPPD); Neck pain; Cervical spine; Arthropathy

Introduction

Bouvet et al. [1] first described crowned dens syndrome (CDS) in 1985 in four women who had neck pain, and the computed tomography (CT) scan showed revealing radiopaque densities surrounding the top and sides of the odontoid process in a crown- or halo-like distribution. The diagnosis of CDS is based on the clinical symptoms and identifying calcifications of all odontoid articular structures (ie, synovial membrane, articular capsule, and ligaments) [2]. It is known that patients with CDS present with recurrent neck pain, stiffness of neck, increased erythrocyte sedimentation rate (ESR), and episodes of fever [3]. Computed tomography scan is the preferred method to detect the calcifications around the odontoid process. Currently, there is no guideline with regarding to the treatment of CDS, and the mainstay of treatments is short oral steroid therapy or nonsteroidal anti-inflammatory drugs. The goal of this report is to describe early and late stage findings in CDS.

Case report

Case 1

A 63-year-old man with a medical history significant for hypertension and diabetes was admitted to the hospital because of uncontrolled diabetes. He also reported a 1-week history of headache, fever, neck pain, and difficulty in moving his neck. He had experienced similar neck symptoms in the past, which were treated with nonsteroidal anti-inflammatory drugs and rest. Spine surgery consultant was

FDA device/drug status: Not applicable.

Author disclosures: ***HZ:*** Nothing to disclose. ***DJ:*** Nothing to disclose. ***ES:*** Nothing to disclose.

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http://dx.doi.org/10.1016/j.spinee.2015.05.019
1529-9430/

called because of neck pain and stiffness. Initial evaluation showed an elevated ESR and C-reactive protein level to 60 mm/h and 12 units, respectively. Physical examination showed tenderness over the neck and occipital regions. The range of motion of neck, including flexion, extension, and lateral rotation, was limited. The neurological examination was normal. A CT of cervical spine showed calcifications in soft tissues adjacent to the odontoid characterizing CDS (Fig. 1) and only mild degenerative changes in the lower cervical region. The magnetic resonance imaging showed no obvious compression on cervical spinal cord. The diagnosis of CDS was made based on his CT image and clinical symptoms. He was treated with only nonsteroidal anti-inflammatory drug and insulin to control his blood glucose. His neck symptoms improved over the next few days, and his blood glucose and ESR returned to normal before discharge.

Case 2

An 81-year-old man, with a medical history significant for hypertension, hypercholesterolemia, Type II diabetes mellitus, and cardiovascular disease, presented with 1-month history of progressive neck pain and decreased muscle power in his upper limbs. One month before, symptoms started suddenly with right shoulder pain and spread to the neck and scalp with difficulty of moving his neck. He also noted progressive weakness in his upper extremities. He was admitted to hospital because of progressively worsening neck pain and decreased muscle power of upper extremities. On musculoskeletal examination, passive rotation of the neck was limited. His right knee was swollen and tender, with moderate effusion. Neurological examination showed his muscle power on bilateral upper extremities was III/VI. Laboratory evaluation showed an elevated ESR of 75 mm/h and C-reactive protein 9.8 units. Synovial fluid showed calcium phosphate crystal deposition (CPPD) crystals with no organisms. The CT images on sagittal view revealed foci of increased densities surrounding the odontoid (Fig. 2); on coronal view, there were multiple calcified foci around the tip of dens because of the atlantoaxial synovial calcifications in a crown- or halo-like distribution (Fig. 2). Computed tomography images also showed diffuse destructive arthropathy in cervical spine

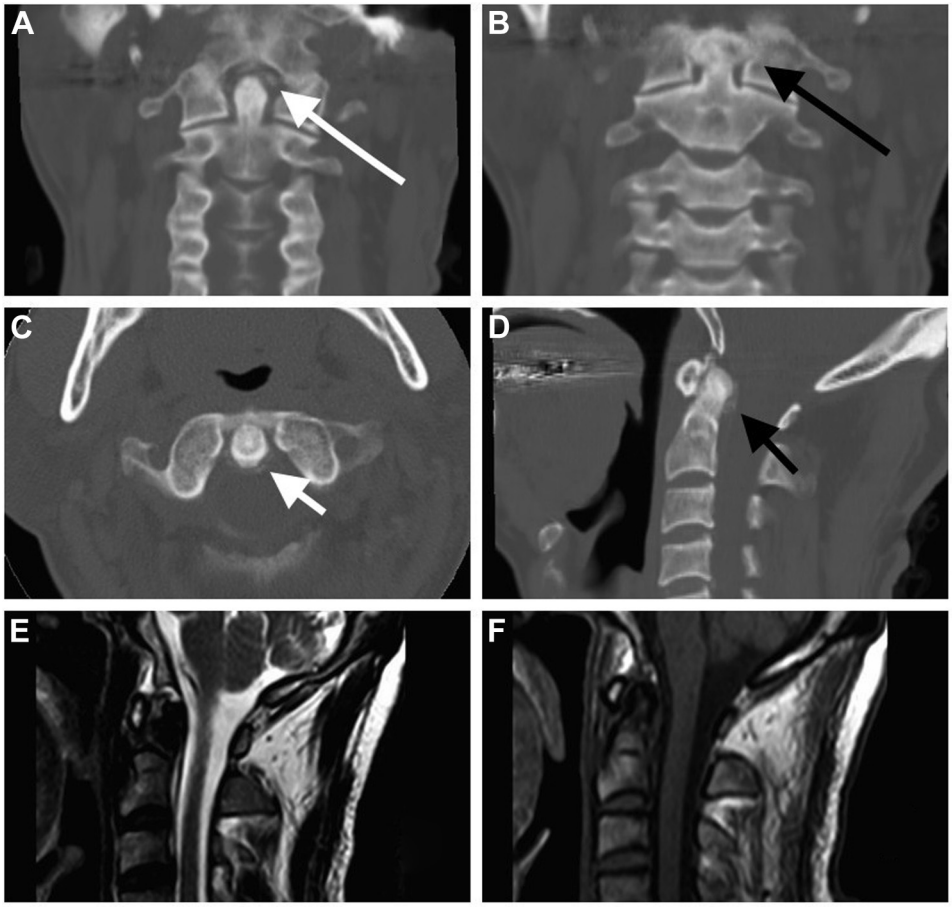

Fig. 1. (A) Computed tomography of cervical spine, on coronal view, showed calcifications on the top of dens (white long arrow); (B) another coronal view showed calcifications around the dens and fused to the lateral mass of C1 (black long arrow); (C) an axial view showed calcifications on the cruciform ligament (white short arrow); (D) a sagittal view showed calcifications behind the dens (black short arrow); (E) T2-weighted magnetic resonance imaging (MRI) showed no obvious spinal cord compression of calcified tissue; and (F) T1-weighted MRI showed similar findings.