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The early and late stage of Crowned Dens Syndrome: Two Cases Report

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Key words: Crowned Dense syndrome; Pseudogout; Calcium phosphate crystal depositions (CPPD)

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Abstract

Background. Crowned dens syndrome is a rare form of CPPD 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 stage and late stage of CDS and it consequences as the result of repeated attacks of CDS at cervical spine in its late stage.

Study design. 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 ESR, CRP, episode of fever, and increased WBC along with high blood glucose levels. The first case showed early phase of CDS with CT scan only showed mild calcification around the dens. The second case had appearance of later stage of CDS with more severe chronic degenerative changes of cervical spine.

Conclusions. Early stage of CDS can be difficult to identify due to 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, radiolographic features often include diffuse periodontoid calcifications, diffuse destructive discopathies and apophyseal joint destruction, and patient might have severe neurological symptoms.

Introduction

Bouvet first described crowned dens syndrome (CDS) in 1985 in four women who had neck pain and the CT scan showed revealing radiopaque densities surrounding the

top and sides of the odontoid process in a crown- or halo-like distribution [1]. The diagnosis of CDS is based on clinical symptoms and identifying calcifications of all odontoid articular structures (i.e. 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, and episodes of fever [3]. CT 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 non- steroidal 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 due to 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 NSAID's and rest. Spine surgery consult was called due to due to neck pain and stiffness. Initial evaluation showed an elevated erythrocyte sedimentation rate (ESR) and C-reactive protein level to 60 mm/hour 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 characterising CDS (Figure 1) and only mild degenerative changes in the lower cervical region. The MRI showed no obvious compression on cervical spinal cord. The diagnose of CDS was made based on his CT image and clinical symptoms.

He was treated with only NSAID and insulin to tighten 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 DM, cardiovascular disease, presented with one 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 due to 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 erythrocyte sedimentation rate (ESR) 75 mm/hour and C-reactive protein 9.8 units. Synovial fluid showed CPPD crystals with no organisms. The computed tomography (CT) images on sagittal view revealed foci of increased densities surrounding the odontoid (figure 2); on coronal view, there were multiple calcified foci around the tip of dens due to the atlantoaxial synovial calcifications in a crown- or halo-like distribution (figure 2). CT images also showed diffuse destructive arthropathy in cervical spine shown in sagittal and coronal view (Figure 2). His Pelvis CT also showed multiple calcifications foci around pubic symphysis area. Magnetic resonance image (MRI) of cervical spine showed enhancement of the odontoid process and adjacent soft tissues, compressing spinal cord at C2 level (Figure-2). He was consulted by spine surgery team due to cervical myelopathy and decreased

muscle power of upper extremities. He was treated conservatively with cervical spine collar and was started on 40 mg of prednisone for 3 days with gradually tapering down dose. His muscle power and neck movement returned to normal within 5 days. Before discharge, his ESR returned to 30 mm/hour with significant improvement of his general condition.

Discussion

The crowned dens deposits are caused by Calcium phosphate crystal depositions (CPPD) crystals. CPPD diseases are caused by the formation of CPPD crystals in joints and periarticular tissues [4]. The diagnosis of CPPD disease is made clinically by chondrocalcinosis on Xray and CPPD crystals in the synovial fluid [5]. The CPPD deposits can also be observed in the spine, especially in the intervertebral discs, posterior longitudinal ligament, facet joints, and ligamentum flavum [6]. Few studies in the past have investigated the natural history of CDS. The effectiveness of medical or surgical treatment of CDS with long term follow up has not been established. In this paper, two CDS cases in early and late stage were reported alone with their clinical and radiographic features.

In the early stage CDS, the spine may be the only site of CPPD disease and is generally asymptomatic [7]. The first case in this report was a middle-age man, who had only mild neck symptoms and never seeks medical help for his neck symptoms. The diagnosis of CDS was made based on his typical clinical symptoms and CT scan images, which showed only few sections calcifications around the dens in the coronal views of CT scan, while other levels of subaxial cervical spine remains less affected. Sekijima Y [8] reported a series of 14 CPPD patients with periodontoid calcification. 6 of them had calcifications in other area of cervical spine, and average age of this group was 83; while in patients with only calcifications around the dens, the average age was 73. This data and our first case shared the similar clinical findings that in the early stage of CDS calcifications could only involve around the dens and the diagnosis could be easily missed. Our imaging finding also confirmed that CT scan

allowed for an easier demonstration of minute calcifications in the area of the odontoid process, with sensitivity above that of MRI in the diagnose of CDS [9], especially in the early stage of CDS.

In the later stage, patients usually have severe neck symptoms with neurological deficits due to the long standing repeat attacks of CDS in cervical spine [10]. CPPD crystal may deposit in the intervertebral disc and apophyseal joint, causing chronic inflammation and destructive lesions. Our second patient had severe CPPD diseases and erosive cervical spondyloarthritis, probably due to the repeated attack of CDS. His CT images showed periodontoid calcifications and advanced spondyloarthritis, which involved the atlantoaxial joint, the intervertebral discs and appophyseal joints, causing diffuse destructive discopathies as well as destruction of facet joints in cervical spine. He also had diffuse calcifications of pubic symphysis from his CPPD. In this case of late stage CDS, medical treatments instead of surgical treatments were provided to him due to his poor general condition and cervical spine bone quality.

In summary, due to the rarity of the diseases and non-specific presentation the early stage of CDS is often missed by clinicians and radiologists. Undiagnosed CDS can lead to severe chronic inflammatory destruction of bone structure of spine, causing neurological deficit as shown in our second case. The treatment of late stage of CDS can be difficult due to its extensive bone destruction, poor bone quality, and neurological symptoms. It is very important to identify the CDS in the early stage in order to prevent severe complications in the late stage.

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Legend of figures

Figure-1. A: CT 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 MRI showed no obvious spinal cord compression of calcified tissue; F: similar findings in T1 weight MRI image.

Figure 2. A: Coronal view of computed tomography (CT) images revealed multiple foci of increased densities surrounding the odontoid, and atlantoaxial synovial calcifications in a crown- or halo-like distribution around the odontoid process (white long arrow); B: Sagittal view of CT image showed foci of increased densities behind the odontoid (black long arrow); C: an axial image showed calcified tissues around craniocervical ligaments (white short arrow); D: a view of pelvic CT showed calcification around pubic symphysis joint (black short arrow); E: another coronal view showed severe destructive arthropathy in the apophyseal joint; F: Magnetic resonance image (MRI) of cervical spine shows that revealed enhancement of the odontoid process and adjacent soft tissues, compressing spinal cord at C2 level.