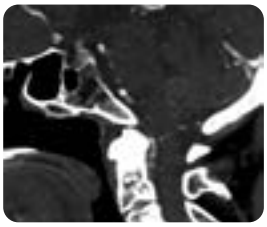

CROWNED DENS SYNDROME, AN UNUSUAL MANIFESTATION OF CERVICAL PAIN: A CASE REPORT

Síndrome de la apófisis odontoides coronada, una manifestación inusual de dolor cervical: presentación de un caso

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Chondrocalcinosis
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Cefalea

Summary
The calcium pyrophosphate dehydrate (CPPD) or hydroxyapatite (HA) crystal deposition disease can manifest in any joint and when it comprises the cervical spine can lead to pain. Crowned dens syndrome is a rare clinical condition that is featured by crown-like calcification of ligaments around the odontoid process. We describe the case of a 70-year-old man with cervical pain, headache and fever for over a week in whom a neurological condition was suspected. A CT scan revealed lentiform calcifications of the transverse ligament of the atlas. Treatment with steroids and non-steroidal anti-inflammatory improve the symptoms. A proper clinical history and imaging studies avoid unnecessary procedures and can let this entity to be included as a differential diagnosis in acute cervical pain.

Resumen
Las enfermedades por depósito de cristales de pirofosfato dihidratado de calcio o hidroxiapatita se pueden manifestar en cualquier articulación y cuando se acumulan en la columna cervical pueden causar dolor. El síndrome de la apófisis odontoides coronada es una rara condición clínica que consiste en la calcificación en forma de corona de los ligamentos que rodean a la apófisis odontoides. Se describe el caso de un hombre de 70 años de edad con una semana de cervicalgia, cefalea y fiebre, que ingresa por sospecha de patología neurológica. Se realiza TC de cráneo que documenta calcificaciones lentiformes del ligamento transverso del atlas. Se instaura manejo con corticosteroides y antiinflamatorios no esteroideos, con mejoría. Una adecuada anamnesis, junto con estudio por imagen, evita procedimientos innecesarios y permite incluir esta entidad en el diagnóstico diferencial del dolor cervical agudo.

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Introduction

In 1982, Ziza and co-workers (1) described for the first time a case of acute suboccipital cervical pain in a patient with hydroxyapatite crystal deposition disease, whose radiological finding indicated crown-shaped calcifications around the odontoid process. Three years later, in 1985, Bouvet and collaborators reported the

same finding in four patients, and called it the crowned dens syndrome (CDS) (2).

Both calcium pyrophosphate crystal deposits and hydroxyapatite deposits affect the cervical spine around the odontoid process, the synovial membrane, the joint capsule, the transverse ligament, the cruciate ligaments and the wings. Its incidence is unknown; however, calcifications in the odontoid process are observed in 40-60% of symptomatic patients and in 15% of asymptomatic people (2).

CDS is associated with episodes of mono-articular arthritis, whose clinical findings range from a quiescent form of the entity to acute cervical pain - with marked restriction of the arches of movement - with irradiation to the upper limbs; sometimes it simulates meningeal signs due to the inflammatory response that develops in the course of the disease, due to feverish pictures and headache of temporary predominance (triad: headache, fever and morning nuchal rigidity) (3).

A predilection for women has been seen, with an age of onset of 70 years and a prevalence of cervical pain of 2% of the patients studied with CDS (4).

Case presentation

70-year-old male patient who was admitted to the "Hermanos Ameijeiras" Clinical Surgical Hospital due to a one-week history of temporary headache of moderate intensity, associated with fever and cervical pain exacerbated by arches of movement. Relevant history: transitory ischemic stroke 10 months prior to admission, arterial hypertension in management with Enalapril 20 mg every 12 hours.

On physical examination he was in regular condition, painful, alert, with temperature of 38.5 °C, heart rate of 90/min, respiratory rate of 20/min, SO2: 97%, FIO2: 98%. GLASGOW 15/15. Nuchal rigidity and dubious Kernig's sign were observed. There was no involvement of cranial pairs or signs of focalization.

Complementary studies showed leukocytosis of 15,000/mm3 with neutrophilia of 9,500/mm3, C-reactive protein (CRP) at 29 mg/L, VDRL: non-reactive. Upon suspicion of meningitis, cranial CT with contrast medium was performed, which did not document intra- or extra--axial lesions, meningeal enhancement or hydrocephalus. Towards the cranio-cervical junction, lentiform calcification of the atlas transverse ligament surrounding the odontoid process was observed, suggestive of crowned dens syndrome (CDS) (Figures 1 and 2). Under this suspicion, uric acid was examined in the blood, with a result of 12 mg/dL, which confirmed the diagnosis of CDS. Management with diclofenac 75 mg every 12 hours, prednisone 20 mg day and colchicine 1 mg day was instituted, which resulted in improvement of symptoms on the fourth day and a decrease in the acute phase reactants. He was discharged on the sixth day with indomethacin 25 mg every 12 hours for 8 days. To date, he has attended a neurological check-up, without recurrence of the clinical picture and with adequate adherence to his chronic medication.

Discussion

One form of the presentation of arthritis by crystal deposition (formerly called pseudogout) in the cervical spine is the crowned dens syndrome (CDS), named after the crown-shaped configuration around the odontoid process. The clinical spectrum is variable, from headache to meningeal picture, due to the inflammatory response and the catabolic effect of the crystals on the chondrocytes and without oviocytes (5).

Diagnosis by conventional radiology is limited by the interposition of anatomical structures, movements of the neck, the failure to open the mouth in the frontal radiographic projection of odontoids or the taking of the examination by a non-expert technician, which often make it difficult to detect calcifications surrounding the atlanto-odontoid joint and its adjacent structures (6-7).

The study of choice is the head and cranio-cervical junction CT scan, which makes it possible to characterize the curvilinear-looking calcifications of the atlas transverse ligament (halo or crown sign) around the odontoid process (4, 6).

Magnetic resonance imaging (MRI) provides an additional study of the soft tissue, although it is limited by the absence of a signal from the calcifications (7).

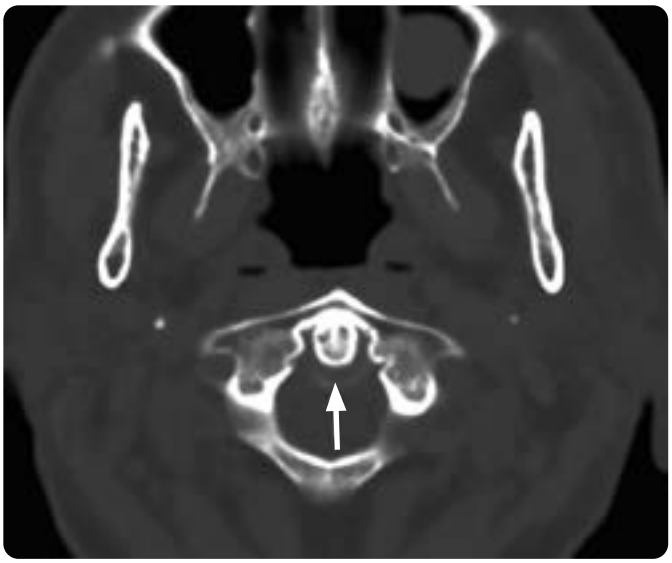

Figure 1. Cranio-cervical junction CT: lentiform calcification surrounding the odontoid process (arrow).

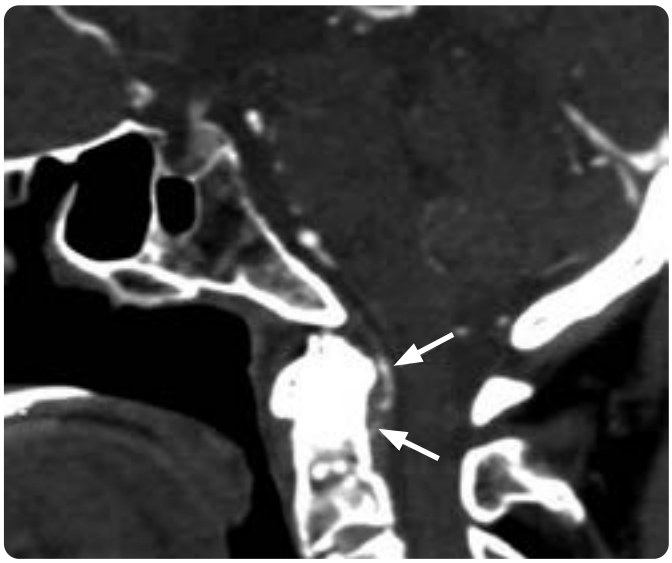

Figure 2. Sagittal reconstruction, calcification of the atlas transverse ligament (arrows) associated with OCAS.

Single photon emission tomography (SPECT) studies can help if active degenerative involvement is suspected, which is seen as a marked uptake around the calcification (6).

The clinical picture resolves in a few weeks with the administration of high doses of non-steroidal anti-inflammatory drugs (NSAIDs), corticosteroids, colchicine or combination therapy, with an excellent prognosis (8).

Differential diagnoses include: rheumatoid arthritis, rheumatoid polymyalgia, meningitis, giant cell arteritis, dyscitis, ankylosing spondylitis and cervical spondylosis (6).

Conclusion

The CDS is an entity with a broad clinical presentation, but with a specific radiological diagnosis. It should be considered in patients with crystal deposition diseases who present clinical picture of headache, nuchal rigidity and febrile episodes.

Conflict of interest

The authors declare that they have no conflict of interest in the preparation of this publication.

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