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CROWNED DENS SYNDROME: AN ILL-KNOW CAUSE OF ACUTE NECK PAIN

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BACKGROUND

Crowned dens syndrome (CDS) is a rare and ill-know etiology of acute occipital and neck pain, with less than 200 case reports since it was first described in 1985 by Bouvet et al. It is caused by crystal deposition and, in 80% of the cases, calcium pyrophosphate dihydrate crystal (CPPD). More frequently described in women over 60 years of age, CDS is defined by the acute neck or occipital pain associated with elevated inflammatory markers and radiological calcifications of the cruciform ligament around the odontoid process. However, patients may experience fever and cervical stiffness. Therefore, among differential diagnoses, infectious causes such as meningitis and tuberculosis should be ruled out, whereas polymyalgia rheumatica and spondyloarthritis should be evaluated among rheumatologic diseases. The first line of treatment is nonsteroidal anti-inflammatory drugs with rapid relief of symptoms within 4 days followed by normalization of inflammatory markers and, in some cases, disappearance of periodontoid calcifications.

CASE REPORT

A 59-year-old woman developed acute neck pain with limited flexion, extension, internal and external rotation of the head associated with elevated inflammatory markers (erythrocyte sedimentation rate: 190 mm/s and C-reactive protein: 470 mg/dL). A magnetic resonance imaging (MRI) was requested and showed atlantoaxial synovitis and calcification of the transverse and alar ligaments (Fig. 1). At the time, it was decided to start prednisone 60 mg/day and methotrexate 15 mg/week. However, 4 weeks after treatment, the patient showed no improvement, remaining with the same pain intensity and limited head movement when she was admitted to our service. After ruling out infectious causes, including tuberculosis spondylitis (Pott's disease), she underwent a new cervical MRI with evidence of progression of synovitis to C5 with signs of joint instability. She was evaluated by the neurosurgery team that performed a posterior arthrodesis with C1-C5 duraplasty. After the procedure, methotrexate was suspended and there was a progressive reduction in corticosteroids. Nonsteroidal anti-inflammatory drugs were introduced with an impressive improvement of pain in only 2 weeks. Up until now, she remains asymptomatic in outpatient follow-up.

Figure 1. Magnetic resonance imaging demonstrating (red arrow) crystal deposition in sagittal (A) and coronal (B) sections.

CONCLUSION

Crowned dens syndrome is often overlooked because of being ill-know by the doctors. Therefore, it is often misdiagnosed and patients undergo inappropriate treatments as seen in this case report. Another point in this case is the challenging and atypical presentation since arthritis is rarely reported in CDS. Therefore, with more knowledge and reports of CDS, it is possible to avoid delay in diagnosis and appropriate treatment can be instituted.