Unilateral spontaneous ankylosis of the C1/C2 joint with “crowned dens syndrome”: Computed tomography scan is imperative in diagnosis

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A 55-year-old female presented to our outpatient services with history of neck stiffness and restricted neck movements of 3 years duration. She had occasional moderate neck pain which was intermittent and had no radicular component or paresthesias. On examination, she had restricted neck movements in all directions with mild pain and no neurological signs. She had undergone magnetic resonance imaging (MRI) of the craniovertebral (CV) junction thrice [Figure 1] over 3 years. Pannus at C2 was diagnosed and she had been advised surgical fusion by at least four different surgeons and had received over 6 months of antitubercular therapy. A computed tomography (CT) scan of the CV junction was recommended at our outpatient department [Figure 2], which revealed a crowned dens syndrome (CDS) with fusion of the left C1/C2 joint. She was managed conservatively with indomethacin and was symptom-free on follow-up. Serum rheumatoid factor and QuantiFERON gold test for tuberculosis were negative. She had prior history of arthritis of the knees but had no evidence of swelling in any large joint at the time of presentation, and hence no arthrocentesis was performed.

CDS is a type of crystal-induced pain syndrome, first described on X-ray imaging in 1982.^[1] It is diagnosed

on both clinical and radiological criteria.^[1,2] There have subsequent reports which have delineated the radiographic features, management, and outcome in such patients.^[1,2] CDS occurs with deposition of hydroxyapatite (HA) or calcium pyrophosphate dihydrate (CPPD) crystals.^[1] HA deposits may occur in the longus colli tendon and intervertebral discs.^[1] CPPD deposition is also seen in the ligaments, discs, and apophyseal joints and ligaments around the CV junction.^[1] An attempt has been made to classify CDS into an early and a late variety.^[2] The former is seen in a younger patient with little calcification around the dens with little or no neurological symptoms, and the diagnosis is easily missed.^[2] Based on these criteria, the present patient would be in an early stage of CDS. CT scan has been proven to be more sensitive than MRI in diagnosis.^[2]

We reiterate that a high degree of suspicion is required to diagnose CDS, and a CT scan of the spine proves more sensitive and is imperative to the diagnosis.

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Conflicts of interest

There are no conflicts of interest.

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Figure 1: Magnetic resonance imaging of the craniovertebral junction revealing T1 sag images (a and b) and T2 axial images (c and d). Figure 1a reveals heterogeneous signal involvement anterior to the dens (black arrow). Figure 1d depicts hypertrophy of the left lateral mass of C1 and C1/C2 facet joint. There was no demonstrable cord compression or atlantoaxial dislocation

Figure 2: Computed tomography scan of the craniovertebral junction. Figure 2a and b is the sagittal reconstructions, Figure 2c is the coronal reconstruction and Figure 2d and e is the axial images. Several areas of calcification at the craniovertebral junction are demonstrated. Figure 2 (a, b, and d) depicts calcifications of the transverse and apical ligaments (white arrows). Complete fusion of the right C1/C2 joint is noted (black arrow - Figure 2c) and hypertrophy of the region is visualized on axial sections [Figure 2e]

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