CLINICAL VIGNETTE

Crowned Dens Syndrome

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Clinical Case

A 72-year-old man with past medical history of anxiety, neuropathy, gout and acid reflux presented for evaluation of subacute low grade fevers, neck pain, and neck stiffness. Over the course of days, his symptoms progressed to include stiffness and pain in the shoulders, wrists, hands, and hips. Evaluation by primary care physician revealed elevated ESR (49) and CRP (2.3). Complete blood count and comprehensive metabolic panel were normal. The patient was started on methylprednisolone dose pack and referred to rheumatology due to suspicion for Polymyalgia Rheumatica.

At the time of rheumatology evaluation, the patient reported improved symptoms with methylprednisolone therapy. He denied temporal headache, jaw claudication, visual symptoms, or scalp tenderness. He reported a similar episode approximately five years prior to the current presentation. At that time he recalled he was also successfully treated with a short course of methylprednisolone, and had been diagnosed with arthritis in his cervical spine. He disclosed a history of gout and had self-discontinued Allopurinol approximately four weeks prior to the present episode. He described infrequent symptoms of bilateral podagra which he self-managed with ibuprofen in recent years. His last recorded uric acid from two years prior was 10.

Radiographs of the cervical spine from two years prior showed diffuse degenerative disease. CT Scan of the Cervical-spine showed granular calcifications around the dens and in the transverse ligament, consistent with Crowned-Dens Syndrome (Figure 1). The patient was diagnosed with likely calcium-pyrophosphate deposition disease and was treated with a four-week steroid taper, which led to complete resolution of symptoms and laboratory abnormalities.

Discussion

The crowned dens syndrome is a lesser-known presentation of crystalline arthritis which was first described in 1980.[1] The majority of cases are due to calcium pyrophosphate deposition disease affecting the cruciform ligament around the odontoid process.[2] Few cases have been reported as secondary to deposition of calcium-hydroxyapatite crystals and also in the setting of hyperuricemia.[2,3]

Patients typically present with complaints of fever, headaches, and neck stiffness. They have elevated ESR and CRP. This presentation commonly raises concern for meningitis, and patients frequently undergo lumbar puncture.[4] Because the cervical stiffness seen in crowned dens syndrome may be associated with shoulder girdle symptoms and jaw claudication, Polymyalgia Rheumatica and Giant Cell Arteritis are other diagnosis which are commonly considered. Therefore, many patients will undergo unnecessary temporal artery biopsy. Unlike meningitis, polymyalgia rheumatica, and giant cell arteritis, the patient may report similar episodes in the past. They appear less likely to have severe hip girdle symptoms than those with polymyalgia rheumatica.[2] Much less common but more severe presentations consisting of neurologic deficits secondary to cervical spinal cord compression have been reported with extensive cervical spine involvement with calcifications producing mass effect.[5]

The imaging modality of choice and gold standard for diagnosis of crowned dens syndrome is CT scan of the cervical spine focusing on C1/C2. The characteristic finding is calcification of the transverse ligament of the atlas, which creates the appearance of a crown or halo. [6,7] CT is superior to MRI for identifying calcification of this structure. Plain radiography may detect calcification, but will not allow for determination of involved anatomic structure.

Management options for crowned dense syndrome include colchicine, oral steroids, or non-steroidal anti-inflammatory drugs. These are the standards of therapy for flares of calcium pyrophosphate deposition disease.

It is important to consider this entity in the setting of head and neck symptoms, as many patients with Crowned Dens Syndrome may report no prior history of calcium pyrophyosphate deposition disease or other crystalline arthropathy.[8] Evaluation and correct diagnosis will avoid unnecessary, invasive procedures and allow for correct treatment. Misdiagnosis of crowned dens as meningitis leads to incorrect treatment with antibiotic therapy, which will prove ineffective. Failure to differentiate Crowned Dens Syndrome from Polymyalgia Rheumatica or Giant Cell Arteritis will lead to a more prolonged course of steroids than is necessary to treat the symptoms of Crowned Dens Syndrome.

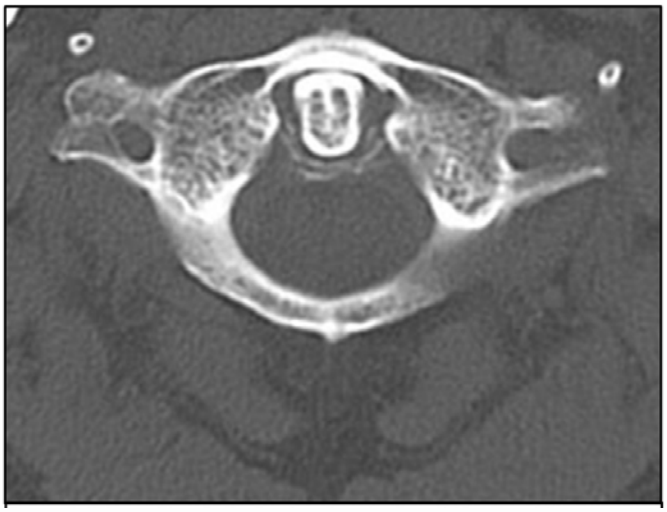

Figure 1: CT Scan of cervical spine showing granular calcification of the transverse ligament.

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