Neuroimaging Highlight

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Calcium Pyrophosphate Dihydrate Deposition Disease Resulting in Cervical Myelopathy

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An 86-year-old woman presented with a six-month history of bilateral hand and feet numbness and diffuse weakness. On examination cranial nerves were intact. The motor examination revealed decreased bulk throughout with no fasciculations. There was diffuse weakness (grade 3 to 4-) in all muscle groups with diffuse 3+ hyperflexia. Sensory examination showed 'glove and stocking' pattern loss of proprioception and vibration with preservation of 'pin prick'pain.

Computed tomography (CT) of the cervical spine (Figure 1A-C) revealed a large, slightly hyperdense mass, containing multiple small calcifications, located at the C1/2 level. This extended from the tip of the clivus superiorly to the lower third of C2 vertebra inferiorly, along the posterior aspect of C2 vertebra. There was erosion of the dorsal portion of the dens with marked compression of the cord at C2 level. On MRI the lesion was isointense to muscle on T1-weighted (Figure 2) and hypointense on T2-weighted images (Figure 3). In addition to compression of the cord there were also changes of myelomalacia. The posterior aspect of the dens appeared to be remodeled with preservation of the cortical line. A benign extraosseous etiology was favored as the lesion showed remodeling of bone rather than destruction, was hypointense on T2-weighted images rather than hyperintense, and had smooth obtuse angles of contact with the dens. The differential diagnosis included synovial osteochondromatosis, inflammatory masses and benign chondroid lesions.

Posterior decompression from the lower part of the occiput to the upper part of C3 vertebra was performed and a biopsy of the epidural mass was obtained. The specimen showed robust calcium pyrophosphate crystal deposition with no evidence of neoplasm or inflammation (Figures 4-6). A final diagnosis of calcium pyrophosphate dihydrate (CPPD) crystal deposition disease resulting in compressive cervical myelopathy was established. At four months post surgery follow-up there was complete clinical recovery with no residual neurological deficit.

Calcium pyrophosphate dihydrate deposition disease is a general term for a disorder characterized by the presence of CPPD crystals in or around joints.[1] These weakly, positively birefringent crystals can cause an inflammatory arthropathy and lead to characteristic chondrocalcinosis.[1] One of the clinical patterns associated with this crystal deposition is called pseudogout, due to its similarity with gouty arthritis.[1]

Calcium pyrophosphate dihydrate crystal deposition may take place in the peripheral joints, particularly the knees, ankles, wrists, and second and third metacarpophalangeal joints.[1] Though involvement of the lumbar spine is not uncommon,[2] CPPD deposition in the cervical spine is, however, rare.[3]

Calcium pyrophosphate dihydrate deposition disease is found in both males and females and is generally observed in the middle aged and elderly.[1] However, the mechanism of CPPD crystal precipitation in cartilage is yet unknown.[1] Various theories include increased production of pyrophosphate dihydrate crystals in articular structures, a leakage of crystals from the articular cartilage and a decrease in rate of clearance of normal CPPD crystals from cartilage or synovium.[4]

In the spine there is frequent involvement of the intervertebral disc resulting in characteristic, though not specific, calcifications. Calcium pyrophosphate dihydrate deposition in the spine can also be found in the ligamentum flavum, posterior longitudinal ligament, interspinous and supraspinous ligaments and the interspinous bursae.[5] To the best of our knowledge, cervical myelopathy resulting from CPPD deposition has been reported in only 26 patients to date in the English literature,[3] of which 80% have been older women. This gender predilection also applies to our patient.

Apart from presenting with chronic cervical symptoms, crystal deposition diseases like CPPD and hydroxyapatite deposition disease can sometimes manifest as acute neck pain.[6,7] When this clinical presentation is associated with a tomographic appearance of calcification surrounding the odontoid process, the condition is termed 'Crowned dens syndrome'.[6,7] This disease (which affects only adult females) presents with inflammatory signs and can be usually treated with nonsteroidal anti-inflammatory drugs with good recovery.[6,7]

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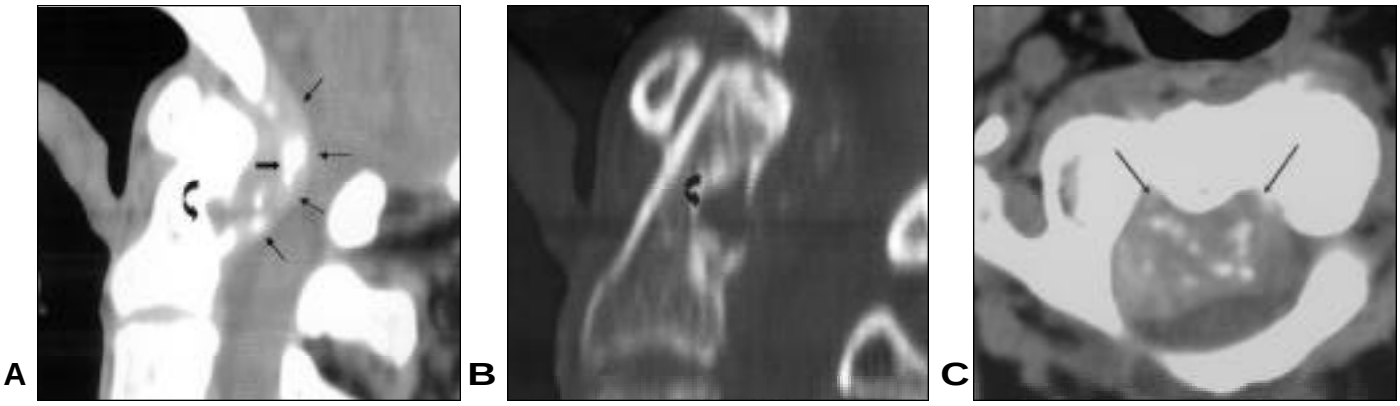

Figure 1: *Sagittal reconstruction CT image (Figure 1a) reveals a mildly hyperdense mass (thin small arrows) in the retro-odontoid area with foci of calcification (small broad arrow). Erosion with remodeling is noted at the posterior aspect of the dens (curved arrow-Figure 1a and b). Axial CTimage (Figure 1c) shows the mass (arrows) compressing the cervical cord posteriorly.*

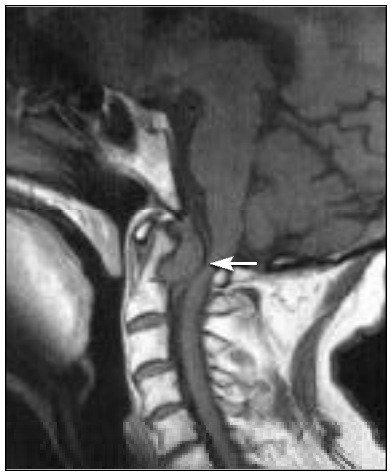

Figure 2: *The mass is isointense on SE T1-weighted sagittal MRI and shows compression of the cervicomedullary junction (black arrow).*

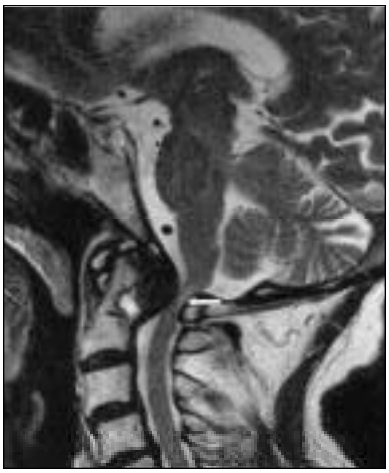

Figure 3: *The mass has low signal intensity on TSE T2-weighted sagittal MRI and the compressed cord shows hyperintensity (white arrow) suggestive of edema/myelomalacia.*

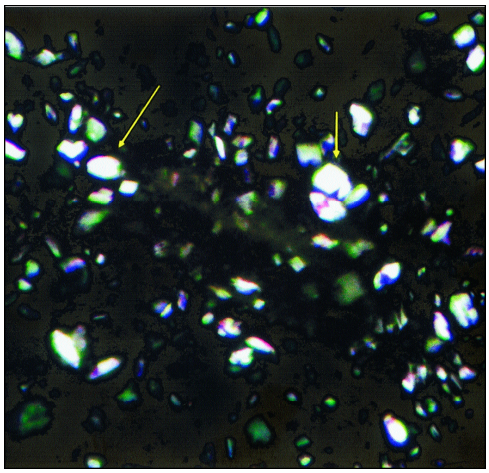

Figure 4: *The calcium pyrophosphate crystals display birefringence under polarized light. (Haematoxylin and eosin, X40).*

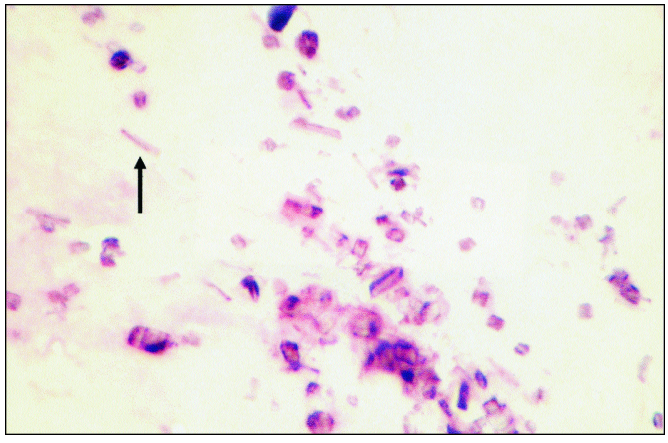

Figure 5: *High power photomicrograph showing rhomboid and needle-like morphology of calcium pyrophosphate crystals. (Haematoxylin and eosin, X40).*

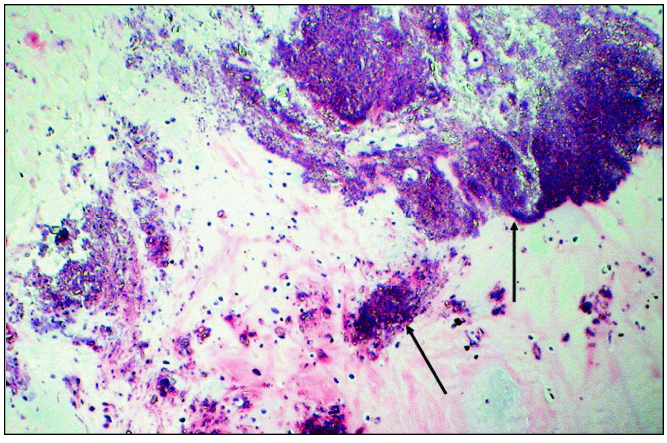

Figure 6: *Fibrocartilage containing dense focal deposits of basophilic calcium pyrophosphate crystals. (Haematoxylin and eosin, X10).*

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Zunkeler et al[8] concluded that CPPD deposition disease should be included in the differential diagnosis of craniocervical junction masses since it has characteristic imaging features and is amenable to early surgical intervention. In their study, CPPD deposition disease in the periodontoid area showed areas of calcification on CTand was isointense to muscle on T1-weighted and iso to hyperintense on T2-weighted MRI. However, in our patient the lesion was isointense on T1-weighted and hypointense on T2-weighted images. This suggests that CPPD deposition disease may be of variable signal intensity on MRI and hence a definitive diagnosis may not be possible in all cases. 'Pseudotumor' of the dens is another differential diagnosis of masses in this region and shows low signal intensity on T1 and T2 weighted images.[9] This entity is an inflammatory mass composed of fibrous granulation tissue and usually results from acute or subacute infection via direct or hematogenous spread.[9]

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