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Calcium Pyrophosphate Dihydrate Deposition in the
Transverse Ligament of the Atlas: An unusual cause of
cervical myelopathy

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Abstract

A 75-year-old male presented with progressive myelopathy due to retro-odontoid massive deposits of calcium pyrophosphate dehydrate (CPPD) crystals. Magnetic resonance imaging revealed a non-enhanced isointense extradural mass on T1-weighted image and heterogeneous intense mass on T2-weighted image. Computed tomography (CT) showed linear calcification within the mass. The mass was resected via a posterolateral approach resulting in marked improvement of the symptoms. Histological examination revealed birefringent rhomboid crystals consistent with CPPD. The preoperative differential diagnosis of periodontoid CPPD deposition disease in the elderly population should be considered, particularly if CT studies demonstrate small areas of calcification within the retroodontoid mass.

Key Words: pseudogout, calcium phosphate dihydrate, odontoid process, craniocervical junction, myelopathy, CT
Introduction

Extradural mass lesions situated posterior to the odontoid process may cause symptomatic anterior compression of the cervicomedullary junction. The differential diagnosis for these lesions includes os odontoideum, inflammatory lesions, and neoplasms such as foramen magnum meningioma, osteoblastoma, or extradural metastatic disease [1,2,3]. Symptomatic anterior cervicomedullary compression is rarely caused by calcium pyrophosphate dihydrate (CPPD) deposition.

CPPD deposition is not a rare condition among the elderly and it probably increases with age and has been estimated to reach a rate of 45% within the population group aged 85 years and over [4]. CPPD commonly involves the joints of the extremities, especially the knee joints, but rarely the spine. Symptomatic involvement of the ligamentum flavum in the cervical and lumbar spine with resulting focal neurological deficits due to nerve root or spinal cord compression has been described in a number of case reports [5,6,7]. Several studies have demonstrated a high frequency of CPPD in the transverse ligament of the atlas in patients known to have pseudogout [8,9]. However, CPPD deposition may rarely cause neurologically symptomatic mass lesions in the cervicomedullary junction. To our knowledge, 16 cases have been reported in the English literature [10,11,12,13,14,15,16,17,18,19].
We report a rare case of massive CPPD deposition at craniocervical junction causing cervical myelopathy

Case Report

A 75-year-old male first presented with numbness of hands and feet persisting for a few months. He was admitted to our university hospital with a 2-month history of progressive weakness and paresthesias in the upper and lower limbs. He had no history of knee joint pain or inflammation of other joints.

Physical examination showed multiple areas of tenderness overlying the occiput and the midline of the neck and restriction of neck motion. Neurological examination revealed diffuse weakness of upper and lower extremities, and glove and stocking type paresthesia. The deep tendon reflexes were hyper active with several beats of ankle clonus.

Plain cervical radiograph showed a partially calcified large mass in the retro-odontoid area (Fig. 1). Sagittal T2-weighted magnetic resonance (MR) imaging showed a heterogeneously intense mass with significant extradural compression (Fig. 2-A). T1-weighted MR imaging revealed an isointense mass with peripheral enhancement after administration of contrast medium (Fig. 2-B, C). Axial MR imaging showed that the spinal cord compression was greater on the right side than on the left
side (Fig. 3-A, B). Computed tomographic (CT) myelography showed a linear calcified retro-odontoid mass causing marked compression of the spinal cord (Fig. 4).

The mass was removed via a posterolateral approach. C-1 laminectomy and upper partial hemilaminectomy of C2 was performed. After the dura was retracted gently on the right side, a firm yellowish mass with a heterogeneous consistency was removed piece by piece. Histological examination revealed rhomboid or rectangular crystals consistent with calcium pyrophosphate dehydrate deposition in fibrocartilage tissue (Fig. 5). Polarized light microscopy demonstrated rhomboid positively birefringent crystals consistent with CPPD.

After surgery the patient was immobilized in a Philadelphia collar. He reported gradual improvement of neurologic function and at the last visit to our clinic one half year after surgery, he could use chopsticks and walk without aid. The only persisting symptom was numbness of the hands. Postoperative MR imaging showed the decompression of the upper cervical cord.

Discussion

Tumours at the craniocervical junction are uncommon and difficult to diagnose clinically. The tumours most often encountered are usually neoplasms; meningiomas when bone erosion is absent, and neurofibromas, chordomas and metastases when bone
erosion or destruction is present [20,21]. Many non-neoplastic mass lesions also have been reported. These have included rheumatoid arthritis, synovial cyst, ossifying posterior longitudinal ligament, and CPPD deposition disease. CPPD deposition disease involving the spine has been increasingly reported in the literature. A study by Markiewitz et al [22] demonstrated axial involvement in 33% of patients with CPPD deposition disease, with 87% of these being in the lumbar spine. Despite this high incidence of lumbar spine involvement, symptomatic lumbar disease is very rare with only a few reported cases [23]. With respect to the cervical spine, symptomatic crystal accumulation in the ligamentum flavum causing myelopathy has been reported [5,6,7]. Involvement of the transverse ligament of the atlas by pseudogout was demonstrated first by Dirheimer et al [8] in a report of 27 patients with pseudogout and no neurologic symptoms. Calcifications in the syndesmoid-odontoid region were found in 12 of these cases (44%). In a similar study, Constantin et al [9] found that 14 of 21 neurologically asymptomatic patients had demonstrable calcification of the transverse ligament (66%). These two reports suggested a high incidence of involvement of the transverse ligament of the atlas. However, involvement of the craniocervical junction is rarely symptomatic. To our knowledge, only 16 neurologically symptomatic cases have been reported in the English literature [10,11,12,13,14,15,16,17,18,19].
Signs and symptoms of CPPD deposition disease involving the cervical spine include neck pain, stiffness, and myelopathy. CT typically shows speckled calcification within the transverse ligament of the atlas or retro-odontoid mass. These lesions are typically isointense to neural tissue on T1-weighted MR images and of mixed intensity on T2-weighted images, and demonstrate peripheral enhancement with gadolinium [14], as shown in this case. Considering the relatively frequent occurrence of CPPD deposition disease, particularly in the elderly population, a preoperative differential diagnosis of periodontoid CPPD deposition should be considered, if CT studies demonstrate small areas of calcification within the retroodontoid mass.

The pathologic diagnosis was based on polarized light examination of tissue obtained by surgery. It seems obvious that the most direct and least expensive means of diagnosis is demonstration of positively birefringent crystals by simple examination, under polarized lenses with a red compensator, of the surgical specimen to distinguish other types of mineral deposition disease [16].

The pathophysiology of CPPD crystal deposition in the articular joint and ligaments is still unknown. It has been postulated that mechanical trauma to the joint cartilage or the ligaments is the initial event that leads to formation of these crystals [14]. Several factors, such as age, heredity, and various metabolic imbalances may predispose to
CPPD crystal formation. In the present case, a patient with degenerative disease of the cervical spine developed a partial tear of the transverse ligament of the atlas, and a cycle of attempted repair and mass formation might have ensued, because the patient had neither metabolic imbalance nor hereditary CPPD deposition disease.

Calcium pyrophosphate dihydrate deposition disease is a rare cause of large, symptomatic mass lesions in the periodontoid location of elderly people. A preoperative differential diagnosis of periodontoid CPPD deposition disease should be considered, particularly if CT studies demonstrate small areas of calcification within the retroodontoid mass. Histologically, the disease is characterized by the deposition of birefringent rhomboid crystals, which can be seen using light microscopy when examining untreated frozen tissue sections or gently processed hematoxylin and eosin-stained sections. Decalcification procedures, commonly employed when sectioning bone or cartilaginous specimens, may wash out the crystals and thus obviate a correct histological diagnosis [14]. Therefore, the tissue should be fixed in 70% ethanol so that water-soluble crystals do not dissolve in formalin. It is recommended that the pathologist should be informed of the preoperative diagnosis so that the surgical specimen is handled appropriately to reveal the diagnostic birefringent rhomboid CPPD crystals.
References


Figure legends

**Figure 1.** Plain cervical radiograph showing a partially calcified large mass in the retro-odontoid area (arrows).

**Figure 2.** Sagittal magnetic resonance (MR) images. T2-weighted MR imaging showing the mass with heterogeneous intensity with significant extradural compression (A). T1-weighted MR imaging showing an isointense mass without enhancement (B) and peripheral enhancement after administration of contrast medium (C).

**Figure 3.** Axial magnetic resonance (MR) images showing the spinal cord compression was greater on the right side than on the left side. T2-weighted MR imaging (A) and T1-weighted MR imaging (B).

**Figure 4.** Computed tomographic (CT) myelography showing a linear calcified retro-odontoid mass causing marked compression of the spinal cord.

**Figure 5.** Photomicrograph of the surgical specimen revealing rhomboid or rectangular crystals consistent with calcium pyrophosphate dehydrate deposition in fibrocartilage tissue