Retro-odontoid Massive Calcium Pyrophosphate Crystal Deposition
—Case Report—

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Abstract

An 86-year-old male presented with progressive myelopathy due to retro-odontoid massive deposits of calcium pyrophosphate dihydrate (CPPD) crystals. Magnetic resonance imaging revealed a non-enhanced isointense extradural mass on the T1-weighted image and heterogeneously intense mass on the T2-weighted image. Computed tomography showed typical punctate and linear calcifications within the mass. The mass was resected via a lateral approach resulting in marked improvement of the symptoms. Histological examination revealed birefringent rhomboid crystals consistent with CPPD. CPPD deposition should be considered in the differential diagnosis of retro-odontoid extradural mass because surgical therapy is beneficial even for elderly patients.

Key words: calcium pyrophosphate dihydrate deposition, pseudogout, odontoid process, myelopathy

Introduction

Calcium pyrophosphate dihydrate (CPPD) deposition is not a rare condition in the elderly with a frequency of 1 per 1000 including asymptomatic cases. The incidence increases with age and approaches 45% in people aged 85 years old or older. CPPD is also referred to as articular chondrocalcinosis or pseudogout. CPPD commonly involves the joints of the extremities, especially the knee joints, but rarely the spine. Myelopathy may occur secondary to nodular deposition of CPPD crystals in the ligamentum flavum of the cervical spine. We report a rare case of massive CPPD deposition at the craniocervical junction causing myelopathy.

Case Report

An 84-year-old male first presented with numbness of hands and feet persisting for a few months in January 1997. He had no rectourinary disorder or neck pain, but developed clumsiness of hands and mild gait disturbance shortly before the first admission to our hospital in February 1997.

Neurological examination revealed clumsy hands, mild weakness of the left upper extremity, and glove and stocking type paresthesia. The deep tendon reflexes were diminished. Magnetic resonance (MR) imaging showed a large retro-odontoid mass. Initially, this mass was thought to be some kind of neoplasm and surgical therapy was recommended but the patient refused because of his age. His symptoms remained stable for another subsequent year but then gradually progressed.

He was readmitted in March 1999. He was unable to use chopsticks and could walk only for a short distance in a walker. He had moderate weakness of all extremities with the sensory level at C-5. His preoperative neurosurgical cervical spine scale (NCSS) was 3:2:2:B. Routine laboratory studies including serum calcium, phosphorus, uric acid, and alkaline phosphatase were all within normal limits. Cervical radiography showed extensive ossification of the anterior longitudinal ligament consistent with diffuse idiopathic skeletal hyperostosis (Fig. 1). The posterior longitudinal ligament was also ossified and there was a partially calcified large mass with peripheral curvilinear calcification in the retro-odontoid region. The shortest distance between this mass and the C-1 posterior arch was 4 mm. Computed tomographic (CT) myelography showed a

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387
punctate and linear calcified retro-odontoid mass causing marked compression of the spinal cord (Fig. 2). There was no calcification in the yellow ligament. T₁-weighted MR imaging revealed an isointense mass with dural enhancement (Fig. 3 left, center). The size of the mass was essentially the same as shown by previous studies. T₂-weighted MR imaging showed the mass as heterogeneous intensity (Fig. 3 right). Bilateral vertebral angiography revealed no tumor stain or displacement of the major vessels. The mass was removed via a lateral approach as
An epidural approach to the mass was attempted but failed due to extensive adhesion of the ventral dura. The dura was opened on the lateral side, and a very hard mass ventral to the spinal cord and covered by the dura was removed piece by piece with disc forceps, preserving the accessory nerve and C-2 nerve root. Histological examination revealed mineral deposits in fibrocartilage tissue (Fig. 4). Polarized light microscopy demonstrated rhomboid positively birefringent crystals consistent with CPPD. Postoperatively, he was immobilized in a Philadelphia collar. His symptoms gradually improved and at the last visit to our clinic one year after the surgery, he was able to use chopsticks and walk normally. The only persisting symptom was numbness of the hands. His final NCS was 5:5:3:D/E. Postoperative MR imaging clearly showed the decompression of the upper cervical cord (Fig. 5).

**Discussion**

CPPD in the craniocervical junction is rare with only 12 reported cases. An epidural approach to the mass was attempted but failed due to extensive adhesion of the ventral dura. The dura was opened on the lateral side, and a very hard mass ventral to the spinal cord and covered by the dura was removed piece by piece with disc forceps, preserving the accessory nerve and C-2 nerve root. Histological examination revealed mineral deposits in fibrocartilage tissue (Fig. 4). Polarized light microscopy demonstrated rhomboid positively birefringent crystals consistent with CPPD. Postoperatively, he was immobilized in a Philadelphia collar. His symptoms gradually improved and at the last visit to our clinic one year after the surgery, he was able to use chopsticks and walk normally. The only persisting symptom was numbness of the hands. His final NCS was 5:5:3:D/E. Postoperative MR imaging clearly showed the decompression of the upper cervical cord (Fig. 5).

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This extradural mass lesion is not enhanced and not associated with obvious bone destruction, so important differential diagnoses include synovial cyst and retroodontoid disc herniation. These rare diseases both manifest as non-enhanced retro-odontoid mass but lack calcification in the lesion. Synovial cysts appear as homogeneous on T2-weighted MR imaging. Neoplasms such as chordoma, chondrosarcoma, and osteoblastoma, and rheumatoid arthritis may reveal similar radiological features in this region, but usually also show contrast enhancement and evident bone destruction.

Hydroxyapatite deposition can also cause myelopathy with similar radiological features to CPPD, but no case has occurred in the retro-odontoid region. CPPD is transformed to hydroxyapatite and

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the coexistence of both substances has been reported.

Extensive ossification of the anterior longitudinal ligament, compatible with diffuse idiopathic skeletal hyperostosis (DISH), in our patient may be associated with CPPD deposition because myelopathy due to calcified yellow ligament (nature of the calcification not described) associated with DISH has been reported. Retro-odontoid CPPD is not so uncommon in the elderly, so this disease must be included in the differential diagnosis of the retro-odontoid non-enhanced extradural mass lesion.

Most cases of cranio cervical junction CPPD were treated by transoral resection with posterior fixation. The posterior approach has never been used for this disease, probably because the mass is so hard that direct exposure of the lesion is necessary. Our lateral approach does not require any fixation procedure, carries less risk of infection, and is less invasive, so seems to be a better procedure for elderly patients.

References


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