Case Report

Cervical Myelopathy from Retro-Odontoid Calcium Pyrophosphate Dihydrate Mass: A Case Report

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Calcium pyrophosphate dihydrate (CPPD) crystal deposition disease is one of the most common forms of crystal-associated arthropathy in the elderly. However, cervical spine is rarely affected, especially in upper cervical area. There have been previous reported cases of symptomatic retro-odontoid CPPD deposition disease in English literature but this case is the first reported in Thai patients. This is a case report of a 67-year-old man who presented with neck pain with progressive myelopathy. Neurologic examination demonstrated a cervical myelopathy with muscle weakness and sensory disturbance of both extremities. Imaging studies showed extradural retro-odontoid mass compressing the spinal cord. The patient underwent occiput to C3 fusion with plating, posterior arch of atlas resection, transoral odontoidectomy, and mass removal. Histological examination of the mass revealed fibrocartilage tissue and rhomboid shaped crystals that showed positive biferingent in polarized light microscopy consistent with CPPD crystals. After surgery, no complication was found, and his neurological function had improved.

Keywords: *Pseudogout, Myelopathy, Cervical spine, Calcium pyrophosphate dihydrate, Odontoid process*

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Calcium pyrophosphate dehydrate (CPPD) deposition disease that was first reported by McCarty in 1962[1] is a crystal induced inflammatory joint disease characterized by intraarticular or periarticular positive birefringent rhomboid crystal deposition. The CPPD deposition disease frequently affected peripheral joint such as knee, wrist, shoulder, elbow, and hip joints. The cervical spine is rarely affected, especially in the upper cervical area. The authors reported an elderly man who had progressive cervical myelopathy and atlantoaxial instability from retro-odontoid mass. To our knowledge, there have only been 49 reported cases of symptomatic cervical CPPD deposition in English literature[2-16] but this is the first case reported in a Thai patient.

Case Report

A 67-year-old man presented with neck pain, progressive weakness, and paresthesia of both extremities, spastic gait, and urinary incontinence for two weeks. Past medical history revealed dyslipidemia and fully recovered right side hemiplegia from ischemic stroke. He had no history of joint pain or prior spinal surgery.

At the time of admission, physical examination revealed limited range of motion in all direction of the neck. According to the Medical Research Council scale, motor strength was graded 3/5 in deltoids, biceps, wrist extensors, triceps, and lower extremities of both sides. Finger flexors and intrinsic function were graded 2/5 of both hands. Pin prick and light touch sensation were diminished below C4 dermatome level on both sides. Proprioceptive sensation was normal. His muscle tone was increased. Deep tendon reflex showed increased biceps, triceps, and ankle reflex bilaterally. Hoffman and scapulohumeral reflex were positive but his jaw jerk was negative. His nurick score had five points. Laboratory tests showed serum C-reactive protein (CRP) concentration was 8.11 mg/L. White blood cell count was 6,690/cmm. Erythrocyte sedimentation rate (ESR) was 62 mm/hour. The lateral view of cervical spine radiographs showed spondylotic change with osteophyte formation at C4 to C7 level. The flexion and extension dynamic lateral radiographs showed C1-C2 instability, increasing in the atlanto-dental interval (ADI) from 3 mm in neutral position to 6 mm in flexion (Fig. 1).

Computed tomographic (CT) scan showed sclerosis at the tip of dens, multiple small calcification

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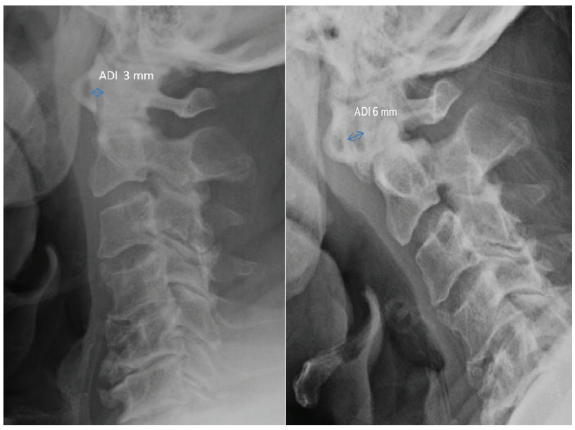

Fig. 1 Plain cervical flexion and extension dynamic radiograph showed instability of cervical spine. The ADI in neutral position was 3 mm (A) then increased to 6 mm in flexion position (B).

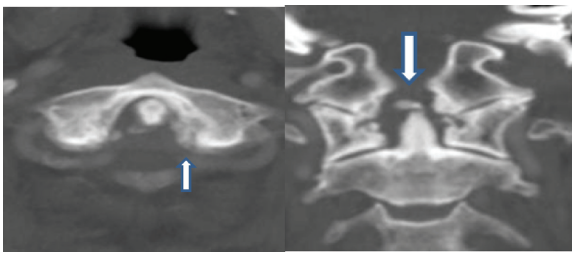

Fig. 2 CT cervical spine showing sclerotic at tip of dens, multiple small calcification around dens (arrow) in axial view (A) and coronal view (B).

around dens, and erosion of both lateral mass of atlas on both sides (Fig. 2).

Sagittal cervical magnetic resonance images (MRI) showed extradural retro-odontoid mass with hypointensity in T1-weighted MRI, slightly isointensity in T2-weighted MRI, and increased signal intensity in T1-weighted fat suppressed MRI after contrast media administration. The bulging mass compressed the spinal cord with increased signal in the spinal cord in T2-weighted MRI (Fig. 3).

Transoral odotoidectomy was performed. In addition, Posterior arch of atlas was resected, then occiput to C3 plating with posterior fusion with iliac crest autograft was done. Intraoperative finding revealed yellowish fibrous mass with minimal adherence to dural sac. Histological examination indicated fibrocartilage tissue and rhomboid shaped crystals that showed positive biferingent in polarized light microscopy consistent with calcium pyrophosphate dehydrate (CPPD) crystals (Fig. 4).

After surgery, the patient was immobilized in a Philadelphia collar. One week after surgery, the patient had right knee swelling and tenderness. Radiographic of right knee showed narrowing of joint space with chrondrocalcinosis. The joint fluid examination showed CPPD crystal. Symptomatic improvement was found in a few days after colchicines and non-steroidal anti-inflammatory drugs were prescribed. The patient's neurological status was gradually improved after surgery. His muscle grading improved to 4/5 MRC scale in all extremities. He could walk independently with a walker at 6 months after surgery. Pin prick and light touch sensation had slightly improved.

According to the Siriraj Institutional Review Board (SIRB) regulation, a case report with only one patient does not need the approval from SIRB.

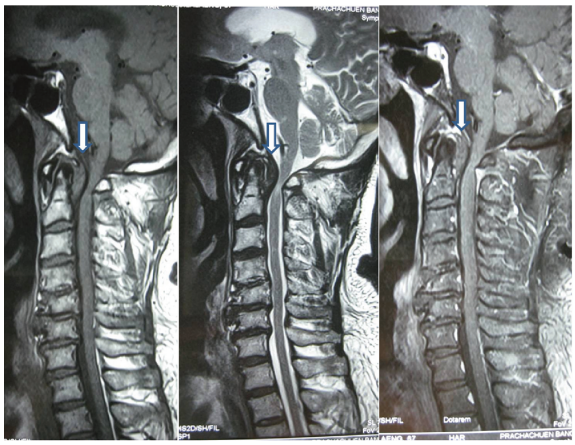

Fig. 3 Sagittal MRI of the cervical spine. The T1-weighted MR images showing an hypointense mass without enhancement (arrow) (A). The T2-weighted MR image showing the mass with heterogeneous intensity with significant extradural compression (arrow) (B) and heterogenous enhancement after administration of contrast medium (arrow) (C).

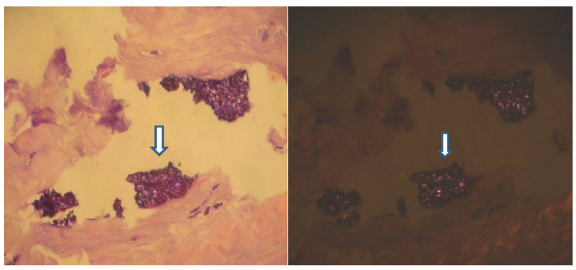

Fig. 4 pathological examination reveal necrotic fibro-cartilage with focal calcification with rhomboid crystal (CPPD) (arrow) (A) on polarized light (B).

Discussion

Calcium pyrophosphate dihydrate (CPPD) crystal deposition disease is one of the most common forms of crystal-associated arthropathy in the elderly that can be associated with some metabolic disease including diabetes mellitus, hyperparathyroidism, hypothyroidism, hypophosphatemia, hypomagnesaemia and gouty arthritis[8,17]. CPPD deposition disease involving the spine that can be found in the ligamentum flavum, posterior longitudinal ligament, interspinous and supraspinous ligament has been increasingly reported in the literature[18]. Markiewitz et al showed axial involvement in 33% of patients with CPPD deposition disease, with 87% of these being in lumbar spine[19]. Despite this high incidence of lumbar spine involvement, symptomatic lumbar disease is very rare with only a few reported cases[20]. In contrast, symptomatic cervical spine involvement appears to be more common. With regard to the cervical spine, symptomatic crystal accumulation in the ligamentum flavum causing myelopathy has been reported[21,22]. Involvement of the transverse ligament of the atlas by pseudogout was first demonstrated by Dirheimer et al[23].

The presence of calcifications of the transverse ligament of the atlas (TLA) in patients with chondrocalcinosis occurs in 71%[24]. Taking into account that calcifications of TLA may manifest as Crowded Dens Syndrome (CDS) in 45% of these patients[24]. Periodontoid CPPD crystal deposition presenting as mass lesions are a rare cause of cervical pain accompanied by compressive myelopathy placing patients at risk for odontoid fracture[25]. The differential diagnosis includes tumor, bone and connective tissue disease including, Paget's disease, ossification of posterior longitudinal ligament, acromegaly, rheumatoid arthritis, gouthy tophi[2]. The tumors most often encountered are meningiomas when bone erosion is absent, and neurofibromas, chordomas, osteoblastoma, osteoclastoma or metastases when bone erosion is present[13]. To our knowledge, forty-nine neurologically symptomatic cases of retro-odontoid CPPD deposition have been reported in the English-language literature[2-16].

Signs and symptoms of CPPD deposition disease involving the cervical spine include neck pain, fever, and restricted neck rotation[26]. 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, of mixed intensity on T2-weighted images, and demonstrate peripheral enhancement with gadolinium[6]. Sushil et al demonstrated four important diagnostic features of CPPD deposition disease in seven patients including (1) a mostly isointense mass on the T1-weighted MR image, (2) areas of calcification within the mass on the CT scan, (3) a mixed density mass with the signal ranging from hypo- to hyper-intense in T2-weighted MR image, and (4) a peripheral enhancement of the mass on the postcontrast MR image. Based on these findings, the retro-odontoid CPPD deposition lesions could be diagnosed with a high degree of certainty preoperatively[27].

Symptomatic patients with retro-odontoid CPPD deposition require surgical decompression. Most reported cases were usually performed either posterior decompression or transoral anterior decompression and combined with posterior stabilization. Most patients experienced rapid and substantial improvement after surgical decompression.

In this patient, CPPD crystal deposits in the transverse ligament of the atlas compressed the spinal cord anteriorly. In addition, there was evidence of C1-C2 instability preoperatively. The combined approach between anterior transoral approach and posterior C1 posterior arch resection was chosen because adequate decompression and tissue diagnosis was archived. Then C1-C2 instability was stabilized posteriorly with an occiput plating, C2 pedicular screws, and C3 lateral mass screws combined with posterior fusion by using iliac autograft because C1 lateral mass erosion may have affected C1 and C2 fixation. The intraopertive finding was typically a firm, yellowish, and avascular mass, which is the same as in the previous report.

The pathologic diagnosis was based on polarized light examination of tissue obtained by surgery. If CPPD deposition disease is suspected preoperatively, the pathologist should be alerted so as to be able to use a polarizing light for birefringence during tissue diagnosis because CPPD crystals may be lost during routine decalcification procedures when processing bone or cartilaginous tissue[6].

The previous studies revealed that symptomatic retrodontoid CPPD deposition is a rare condition and usually requires surgical decompression. Either posterior decompression or transoral anterior decompression provide good prognosis. The present study showed that the combined decompressive surgery (odontoidectomy and posterior C1 arch resection) is safe and beneficial in symptomatic retro-odontoid

CPPD deposition patients. The occiput to C3 plating may be the alternative procedure especially in the patients who have C1 lateral mass osteolytic lesion that may preclude C1 instrumentation.

Conclusion

CPPD deposition in periodontoid area is a rare cause of cervical myelopathy that may be diagnosed preoperatively by using high degrees of suspicion combined with proper radiographic study. The definite pathological diagnosis can be confirmed by using polarized-light microscope. Decompressive surgery via anterior, posterior, or combined approach provides good prognosis.

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Potential conflicts of interest

None.

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ก้อนเกาต์เทียมด้านหลังกระดูกสันหลังส่วนคอที่สองกดทับไขสันหลัง: รายงานผู้ป่วย

ปัญญา ลักษณะพฤกษา, อารีศักดิ์ โชติวิจิตร, ศิริชัย วิลาศรัศมี

โรคเกาต์เทียมเป็นโรคที่เกิดจากผลึกที่พบได้บ่อยในผู้ป่วยสูงอายุโดยพบรอยโรคที่กระดูกสันหลังส่วนคอได้น้อย โดยเฉพาะอย่างยิ่งกระดูกสันหลังส่วนบน แม้ว่ามีรายงานผู้ป่วยกลุ่มนี้รายงานในวารสารต่างประเทศ แต่รายงานผู้ป่วยฉบับนี้เป็นครั้งแรกในผู้ป่วยชาวไทยโดยผู้ป่วยเป็นผู้ป่วยชายไทยอายุ 67 ปี มาพบแพทย์เนื่องจากปวดต้นคอร่วมกับมีอาการทางระบบประสาท การตรวจร่างกายพบมีกล้ามเนื้ออ่อนแรงและชาบริเวณแขนขาทั้งสองข้าง ภาพถ่ายรังสีพบว่ามีก้อนอยู่ด้านหลังต่อกระดูกสันหลังส่วนคอที่สองกดเบียดไขสันหลัง ผู้ป่วยได้รับการผ่าตัดใส่โลหะยึดตรึงกะโหลกลงมาจนถึงกระดูกสันหลังส่วนคอที่ 3 ตัดกระดูกสันหลังส่วนคอที่ 1 ทางด้านหลัง ร่วมกับการตัดปุ่มกระดูกโอดอนตอยด์ทางปาก ผลการตรวจทางพยาธิวิทยาพบมีเนื้อเยื่อพังผืด ร่วมกับผลึกเกาต์เทียม ซึ่งให้ผลบวกจากการตรวจการกระจายแสง ภายหลังการผ่าตัดไม่พบภาวะแทรกซ้อน และอาการทางระบบประสาทดีขึ้นโดยลำดับ