Calcium Pyrophosphate Dihydrate Crystal Deposition Disease in Cervical Radiculomyelopathy

One patient had cervical spinal canal stenosis with radiculomyelopathy due to deposition of calcium pyrophosphate dihydrate within the ligamentum flavum. The MRI of cervical spine showed a calcified nodule over C5-6 level ligamentum flavum with obvious cord compression. After posterior decompressive laminectomy with removal of the calcified nodule, the symptoms and signs relieved remarkably, and the pathology showed calcium pyrophosphate dihydrate deposition within the ligamentum flavum. We presented this case and reviewed the literature to acknowledge so-called “pseudogout syndrome.”

CASE REPORT

This 73-year-old female was admitted to Taipei Veterans General Hospital in September, 2001 with the chief complaint of severe nuchal pain with weakness and tingling sensation in both hands for two more months. Unsteady gait was also noted. In the recent weeks, the clinical symptoms and signs were exacerbated with progressive weakness and tingling of the left hand. Except the history of hypertension and bilateral total knee arthroplasty for...
osteoarthritic knee, she denied any other history such as gout, diabetes mellitus or spinal surgery. The physical and neurological examination showed numbness in left C5, C6 dermatome, decreased muscle power (3+/5+) in left arm, and increased deep tendon reflex in C5, C6. Tests for Hoffman sign, grip & release sign and finger escape sign all revealed positive results.

The laboratory studies including uric acid, complete blood count, erythrocyte sedimentation rate and C-reactive protein were within normal limits. X-rays of C-spine showed spondylosis without obvious calcification or spondylolisthesis. MRI of C-spine revealed a low signal nodule over left C5-6 ligament flavum with cord compression in sagittal view of T1WI and axial view of T2WI (Fig. 1). Under the impression of C5-6 spinal stenosis with radiculomyelopathy, posterior decompressive laminectomy was performed and a 17 mm calcified nodule within ligamenta flavum with adhesion to dura mater and cord compression was found intra-operatively. The nodule was composed of fine granules and chalky white in color. In histologic examination CPPD deposition was present (Fig. 2). Post-operative condition was uneventful and the patient was discharged ten days later with remarkable neurologic improvement.

DISCUSSION

The incidence of CPPD is around 1/1000 with half of the victims older than 80 years of age. Occurrence in patients younger than 50 years of age is rare. In sex distribution, female predominance is found. The location of deposition can be divided into “articular calcification” and “extra-articular calcification.” The former is more common and mainly deposited in fibrocartilage, hyaline cartilage, synovium or capsule, especially in knee joint. The extra-articular type is relatively rare and deposited in tendon, dura matter, ligament soft tissue and usually asymptomatic.

CPPD deposition seldom results in spinal stenosis or myelopathy. The first case of cervical spine ligament...
CPPD deposition was reported by Ellman and Kawano. The patterns of deposition can be linear, nodular or transitional. The composition of CPPD is different from ordinary hydroxyapatite deposition. However, a recent study has proposed that the hydroxyapatite deposition can be transformed from the central part of CPPD of nodular type at the late growth stage. The mid-cervical spine is often affected, especially at C5-6 and C6-7 (81%) level, the thoracic spine is more common at T8 to T12 level (42%), and L2-3 level is usually involved in lumbar gion. The pathophysiology and mechanism of CPPD deposition disease is still unknown. Two hypotheses were described including (1) Primary: overproduction or decreased removal of CPPD crystals from cartilage or abnormality of underlying cartilage collagen. (2) Secondary: trauma or surgery to cause the intracellular pyrophosphate to release from the cells to deposit in the extracellular space. The phagocytosis of the polymorphonuclear leukocytes and the release of chemostatic factors result in acute inflammation, in the more mobile segments of C5-6 & C6-7, the ligamentum flavum undergoes repetitive stretch and minor trauma with aging in duces strong local re sponse. Asymptomatic patients can be observed in regular follow-up. For the acute and symptomatic patients, aspiration of joint effusions, intra-articular injection or oral NSAID/steroid are recommended in the CPPD deposition disease when the cervical spine is compressed posteriorly. Decompressive laminectomy is usually rec om mended in the CPPD deposition disease when the cervical spinal cord is compressed posteriorly. Low dose Colchicine in acute phase, and Ethane-l-hydroxy-1-diphosphate (EHDP; 600 mg qd for month) at chronic stage to reduce cal cium deposition are suggested. Decompressive laminectomy is usually recommended in the CPPD deposition disease as the final option. In conclusion, the calcified nodule to compress the spinal cord posteriorly warrants for decompressive laminectomy.

REFERENCES