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Clinical Studies

Calcium pyrophosphate deposition disease of the cervical and thoracolumbar spine: A report of two cases

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ABSTRACT

Background: Spinal calcium pyrophosphate deposition disease (CPPD) is uncommon, and often resembles more common spine pathologies causing pain and neural compression. Here, we present two unusual cases of CPPD of the cervical and thoracolumbar spines.

Case description: Case 1: A 71-year old female smoker presented with a large epidural mass causing rapidly progressive cervical myelopathy with weakness in the upper and lower extremities.

Case 2: A 66-year-old morbidly obese male presented with chronic back pain for several years associated with progressively worsening radicular pain in his left lower extremity.

Outcome: The first case is an example of tumoral CPPD involving the facet joint and expanding into the epidural space. The second case was an example of CPPD involving a thoracolumbar facet cyst, resulting in unilateral radiculopathy. Both patients were treated surgically and had significant improvement in symptoms postoperatively.

Conclusions: CPPD in the spine is an uncommon diagnosis but should be considered in the differential diagnosis of patients presenting with back pain and associated neurological symptoms. Accurate diagnosis of spinal CPPD is important in that it will guide postoperative management with anti-inflammatory medications and reduce risk of recurrence.

Background

Calcium pyrophosphate deposition disease (CPPD), commonly referred to as pseudogout, is an inflammatory arthropathy characterized by the presence of calcium pyrophosphate crystals in articular or periarticular tissues [1]. Spinal CPPD is uncommon, and often resembles more common spine pathologies causing pain and neural compression [2–16]. Previous reports of spinal CPPD have typically involved the cervical and lumbar spines [17–26]. Here, we present two unusual cases of CPPD of the cervical and thoracolumbar spines. Both patients were treated surgically and had significant improvement in symptoms postoperatively.

Case 1

A 71-year old female smoker presented to clinic with rapidly progressive cervical myelopathy with weakness in the upper and lower extremities. She was referred from an outside hospital for evaluation of an epidural mass at C7 with significant compression of the spinal cord at that level. The patient endorsed loss of manual dexterity, gait impairment, and difficulty with bladder control. Physical exam was notable for mild motor weakness in left hip flexion and knee flexion/extension. Sensation to light touch was intact bilaterally.

Radiographs were notable for gross spondylosis throughout the cervical spine with a 4.6 mm subluxation of C4 on C5 on upright views (Fig. 1A, 1B). Computed tomography (CT) without contrast showed a large, well-demarcated epidural mass with significant calcification extending from the midline of the C7 lamina, as well as subluxation of C4 on C5 (Fig. 2A, 2B). Magnetic resonance imaging (MRI) without contrast again demonstrated the mass at C7, hypointense on all sequences with significant compression of the cervical spinal cord (Fig. 3).

The patient was started on dexamethasone, and then subsequently underwent C2-T2 PSIF, C4-T1 laminectomy, and excision of the epidural mass at C6, C7, and T1 without intra- or post-operative complications. Post-operative medications included non-steroidal anti-inflammatory drugs (NSAIDs). Histopathology of the right C4-C5 facet and C7-T1 lamina and epidural mass revealed fragments of benign fibroconnective tissue and prominent multifocal nodules of dystrophic calcification, con-

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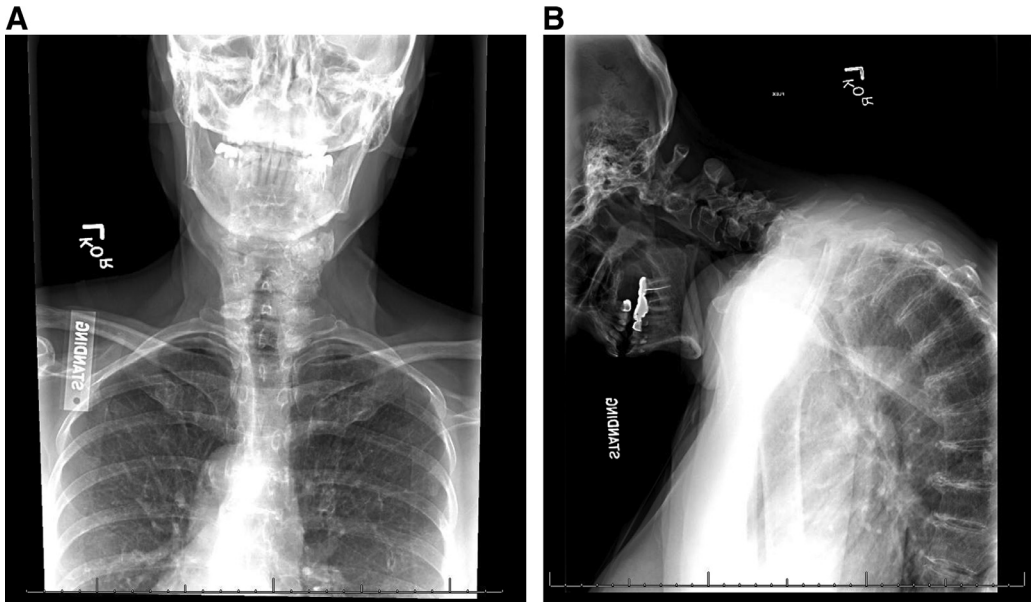

Fig. 1. A, 1B Pre-operative AP (1A) and Lateral (1B) view radiographs were notable for gross spondylosis throughout the cervical spine with a 4.6 mm subluxation of C4 on C5 on upright views.

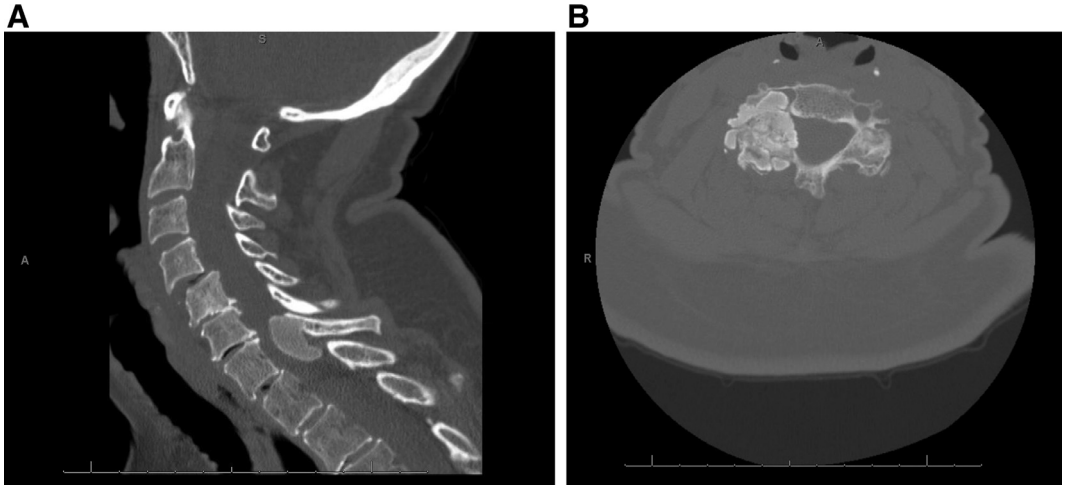

Fig. 2. A, 2B Pre-operative Sagittal (2A) and Axial (2B) view CT images without contrast demonstrating a large, well-demarcated epidural mass with significant calcification extending from the midline of the C7 lamina, as well as subluxation of C4 on C5.

sistent with CPPD. She had significant improvement in functional status and remained neurologically intact at her latest follow-up visit at 1 year post-operatively (Fig. 4A, 4B).

Case 2

A 66-year-old morbidly obese male was evaluated in clinic for chronic back pain for several years associated with progressively worsening radicular pain in his left lower extremity. He had a history of L1–L3 posterior spinal instrumented fusion (PSIF) with L1–L2 interbody fusion, left sacro-iliac fusion, and C5–C7 anterior cervical discectomy and fusion eight years prior to presentation. Physical exam demonstrated no apparent motor or sensory deficits, and was otherwise unremarkable.

Radiographs revealed mild kyphosis with disc space collapse at T12–L1 (Fig. 5A, 5B). His previous instrumentation was intact with no gross instability on flexion or extension views. MRI of the lumbar spine without contrast demonstrated a large facet cyst at T12–L1 with lateral recess stenosis and compression of the spinal cord (Fig. 6A, 6B). Laboratory findings were notable for elevated white blood cell count to 13.59 (ref. range, 4.00–11.00)), mildly elevated erythrocyte sedimentation rate (ESR) to 11 (ref. range, 0–10), and elevated C-reactive protein to 24.39 (ref. range, 0.00–10.90).

The patient underwent left-sided T12–L1 hemilaminectomy, partial facetectomy, and removal of facet cyst without intra- or post-operative complications. Histopathology of the T12–L1 facet cyst revealed CPPD. The patient received NSAIDs post-operatively, and had fully resolution of his radicular symptoms. He otherwise remained neurologically intact at his latest follow-up at 3 months (Fig. 7A, 7B). Particularly given his previous spine surgeries, he will need a longer follow-up to assess his symptoms post-operatively.

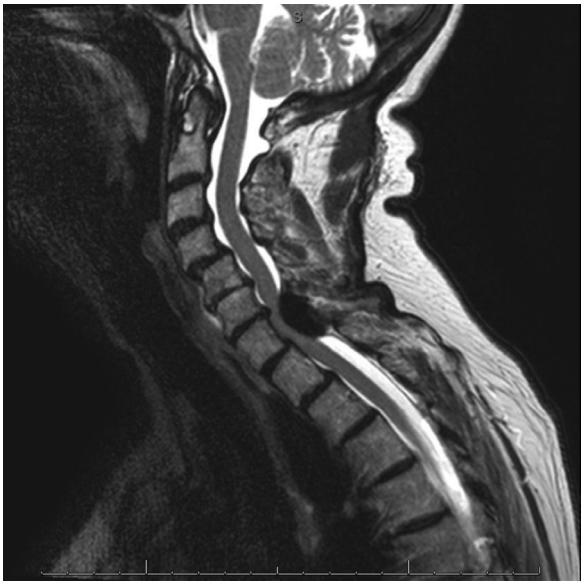

Fig. 3. Pre-operative Sagittal view MRI without contrast demonstrating the mass at C7, hypointense on all sequences with significant compression of the cervical spinal cord.

Discussion

CPPD is a common cause of inflammatory arthropathy in older patients, most commonly affecting peripheral joints such as the knees and wrists [1]. The sporadic type of CPPD is most common, but there are also familial forms and associations with diseases such as osteoarthritis, chronic kidney disease, hemochromatosis, hyperparathyroidism, Wilson's disease, hypothyroidism, hypophosphatasia, and hypomagnesemia [1,27–29]. Acute presentations typically manifest as a mono- or oligoarthritis with warmth, erythema and swelling at the involved joint(s), while chronic cases often resemble polyarthropathies such as degenerative osteoarthritis or rheumatoid arthritis [1,27,28].

While CPPD in the spine is uncommon, previously reported cases have involved the intervertebral disc, ligamentum flavum, facet joint, and neural foramen [2–26]. Spinal CPPD generally presents with symptoms of back stiffness, pain, and radiculopathy. Interestingly, the majority of spinal CPPD cases in the literature have been sporadic cases without the presence of any peripheral disease or associations with metabolic diseases. There appears to be a female preponderance in spinal CPPD, despite no gender predominance of the disease in general. The pathophysiology of the disease process remains unclear [30]. Treatment typically entails surgical decompression with adjuvant NSAIDs and steroids. The literature does not support one particular NSAID over another for the treatment of CPPD, and both traditional NSAIDs or selective cox-2 inhibitors can be used [31]. The patient in case 1 received Naproxen, while the patient in case 2 was prescribed Meloxicam.

The cases presented here highlight two unique cases of histopathologically confirmed CPPD of the spine. The first case presented as a large epidural mass causing rapidly progressive myelopathy. This type of focal deposition of calcium pyrophosphate crystals is called tophaceous or tumoral CPPD, and is a rare presentation of the disease [32–35]. In this case, the epidural mass began in the facet joint and expanded into the epidural space and surrounding soft tissues. Although facet joints are commonly involved in spinal CPPD, this is typically not the case in the cervical region. The majority of previously reported cases in the cervical spine have involved either the periodontoid structures, which is also known as crowned dens syndrome, or the ligamentum flavum [36–38].

In the second case, the patient developed CPPD involving a thoracolumbar facet cyst, resulting in a unilateral radiculopathy. Although he remained afebrile, his lab findings were consistent with an inflammatory process. Of note, this patient had a history of multiple spine surgeries. However, it is unclear what role these previous surgeries had in the development of CPPD. Although local trauma is a known risk factor for CPPD [39–42], our patient presented eight years after his previous spine surgeries. Ogawa et al. reported the first case of acute lumbar spinal pseudogout occurring four weeks after spinal instrumentation [39]. Similar to our case, the patient had back pain and abnormal blood levels of inflammatory markers. However, in their case there was a clear temporal association between the initial surgery and the development of CPPD.

CPPD in the spine is an uncommon diagnosis, but should be considered in the differential diagnosis of patients presenting with back pain and associated neurological symptoms. Index of suspicion should be increased in patients with progressive neurological compromise, signs of an inflammatory process, and those with known risk factors for CPPD. Accurate diagnosis of spinal CPPD is important in that it will guide post-operative management with anti-inflammatory medications and reduce risk of recurrence.

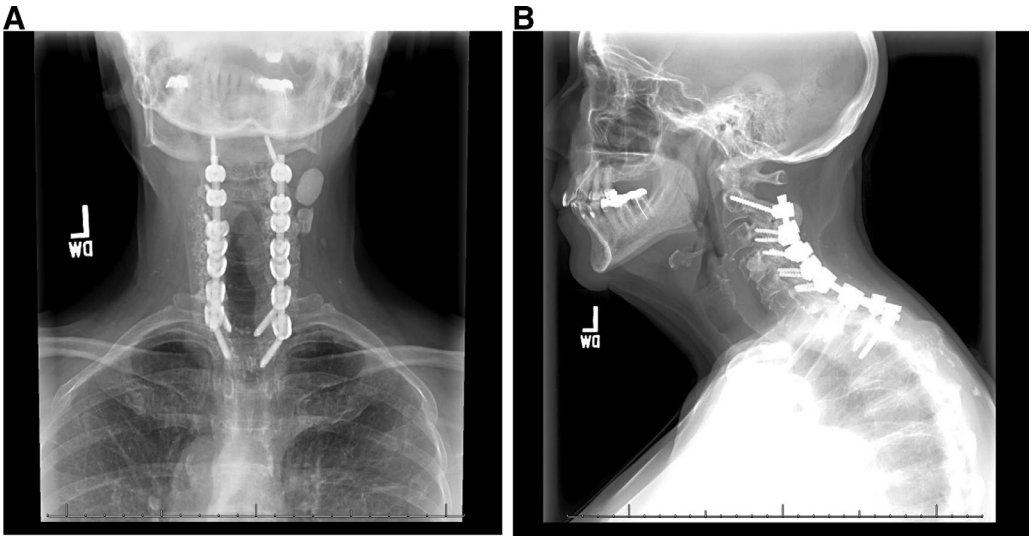

Fig. 4. A, 4B Post-operative AP (4A) and Lateral (4B) view radiographs demonstrate intact hardware without evidence of failure.

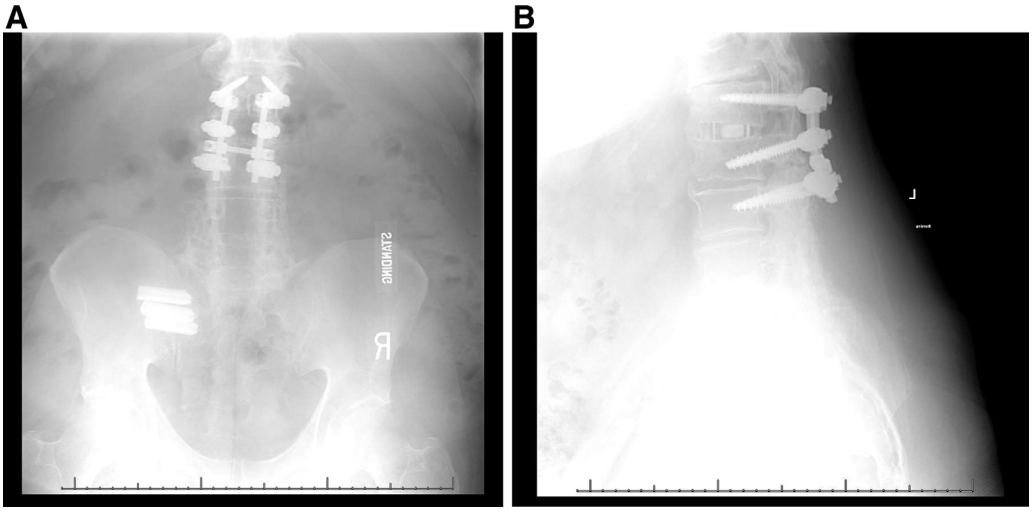

Fig. 5. A, 5B Pre-operative AP (5A) and Lateral (5B) view radiographs demonstrating intact hardware with disc space collapse at T12–L1.

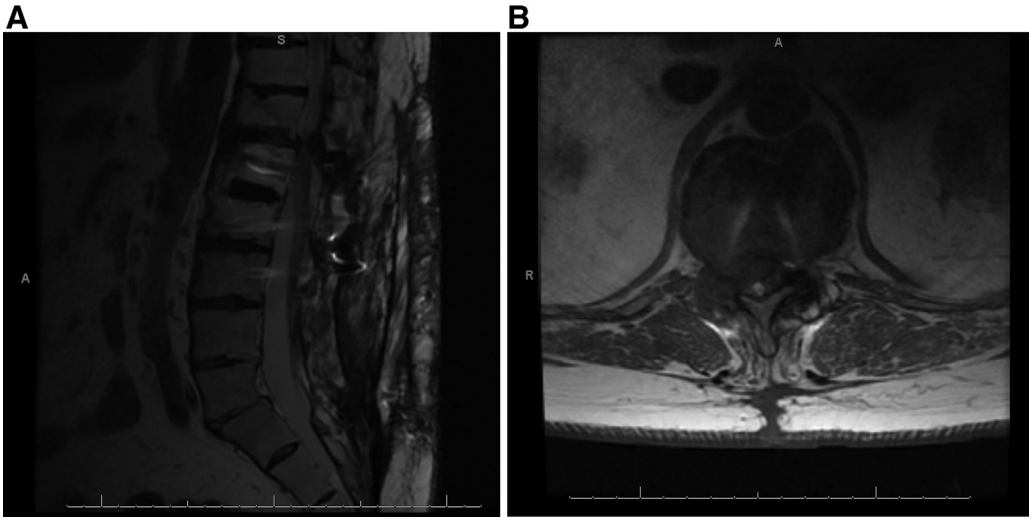

Fig. 6. A, 6B Sagittal (6A) and Axial (6B) view MRI of the lumbar spine without contrast demonstrating a large facet cyst at T12–L1 with lateral recess stenosis and compression of the spinal cord.

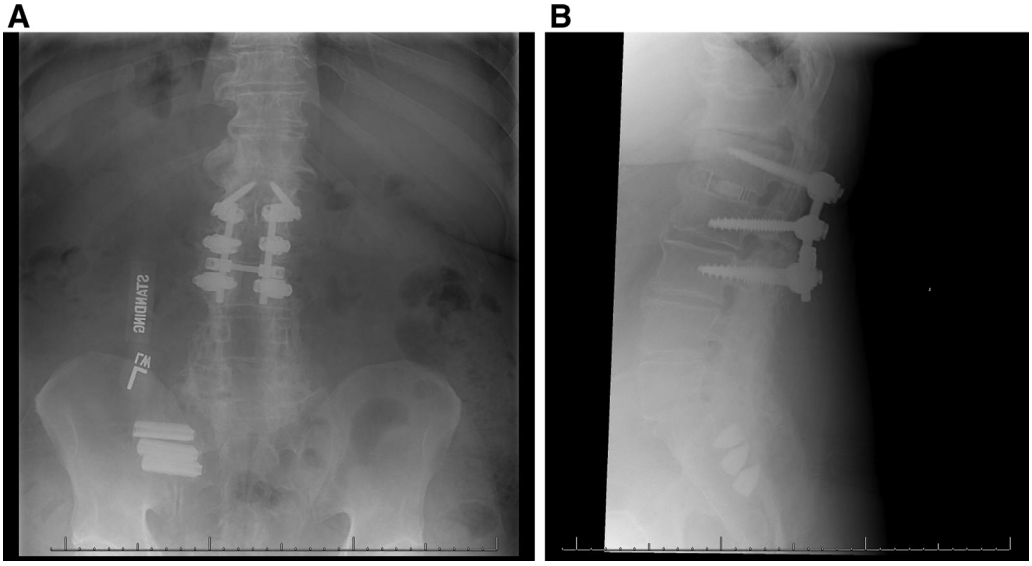

Fig. 7. A, 7B Post-operative AP (7A) and Lateral (7B) view radiographs demonstrating intact hardware without evidence of failure.

Declarations of Competing Interests

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Patient Informed Consent Statement

The authors declare that informed patient consent was taken from all the patients.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.xnsj.2020.100026.

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