Retro-Odontoid Calcium Pyrophosphate Dehydrate Deposition: Surgical Management and Review of the Literature

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Abstract

Study Design   Case report and review of the literature.

Objective   A retro-odontoid mass is a rare cause of cervical compression and myelopathy. The differential diagnosis includes the following: metastatic disease, primary tumor, collagen disorder, or inflammatory disease. Calcium pyrophosphate dihydrate (CPPD) deposition has been referred to as "crowned dens syndrome" when there are periodontoideal calcifications. There are only a few reported cases where CPPD presents as a cystic retro-odontoid mass in the atlanto-dens interval. In previous descriptions of surgical intervention, transoral resection of the mass is associated with significant morbidity and usually requires stabilization. The objective of this article is to report a case of an unusual presentation of CPPD disease of C1/C2, where we used a novel, minimally invasive surgical technique for decompression without fusion.

Patients and Methods   An 83-year-old female patient presented with progressive cervical myelopathy over a 3-month period. Computed tomography and magnetic resonance imaging demonstrated a cystic odontoid mass with a separate retro-odontoid compressive mass. A novel, minimally invasive transoral aspiration was performed. Histologic confirmation of CPPD was obtained.

Results   Postop imaging showed satisfactory decompression, which was maintained at the 6-month follow-up. This correlated with clinical improvement postop and 6-month follow-up.

Conclusion   CPPD in the atlanto-dens interval may present as a cystic retro-odontoideal mass and should be included in the differential. We used a transoral minimally invasive approach to aspirate the cyst. This novel technique avoided the need for a stabilization procedure or morbid transoral resection and provided excellent results immediately and at 6 months.

Keywords

► crowned dens syndrome
► calcium pyrophosphate dihydrate
► retro-odontoid mass
► cervical myelopathy
► crystal deposition disease
► cervical decompression
► cervical mass aspiration

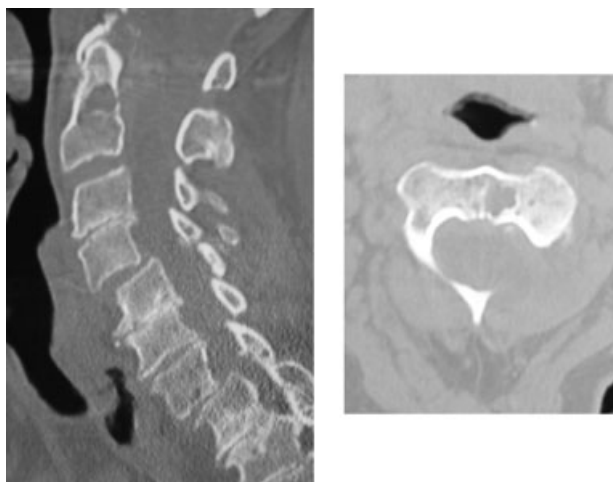© 2014 Georg Thieme Verlag KG
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Fig. 1 Sagittal and coronal computed tomography scan demonstrating cystic mass at the base of the dens, with multilevel cervical spondylosis.

Introduction

Calcium pyrophosphate dihydrate (CPPD) deposition in the cervical spine is an unusual cause of cervical spinal cord compression and myelopathy. Retro-odontoideal crystal deposits may lead to acute pseudogout and can be visualized as calcifications that surround the apex of the dens, and is therefore known as "crowned dens syndrome" (CDS).[1,2] This entity may manifest itself in a variety of ways, but it usually presents as acute neck pain and stiffness.[3,4] With disease progression, it may present as a mass-occupying lesion, causing progressive cervical spinal cord compression and myelopathy. In general, noninfectious, nontumorous retro-odontoid cystic masses are exceedingly rare.[5] They have mainly been described in the literature as synovial cysts, ganglion cysts, transverse ligament degeneration, or herniated discs.[5,6]

We present a rare case of retro-odontoid CPPD resulting in cervical myelopathy that required surgical intervention. Our patient is an 83-year-old woman with a cystic lesion within the body of the dens, as well as a retro-odontoid mass, causing cervical spinal cord compression. She had no history of pseudogout, or other predisposing factors. We successfully decompressed her lesion using a minimally invasive transoral decompression, under fluoroscopic visualization. The literature on cervical CPPD was reviewed, and our surgical approach discussed.

Case Report

An 83-year-old female patient presented to our emergency department (ED) with progressive difficulty with balance and upper extremity clumsiness. While the balance difficulty had been progressive for some time, it had worsened drastically in the few weeks before presentation in the ED. Medical history is negative for diabetes or rheumatoid disease. There was no history for joint swelling or inflammation in any of her major joints.

Physical examination revealed hyperreflexia in her upper and lower extremities with bilateral positive Hoffman and pathological clonus. She had diffuse weakness in her upper and lower extremities suggestive of neural compression. She also had diffuse paresthesias that was nondermatomal.

Computed tomography (CT) and magnetic resonance imaging (MRI) scans were completed to determine the etiology of her myelopathy. The CT scan revealed multilevel spondylosis without evidence of acute fracture or instability, and with a cystic lesion located within the body of the dens (►**Fig. 1**). This lesion was noted to be hyperintense on T2-weighted MRI and to be associated with a large extradural mass. The mass caused significant cord compression, and was also associated with a bilobulated cystic mass that resulted in additional anterior compression, best appreciated on the axial MRI (►**Fig. 2**).
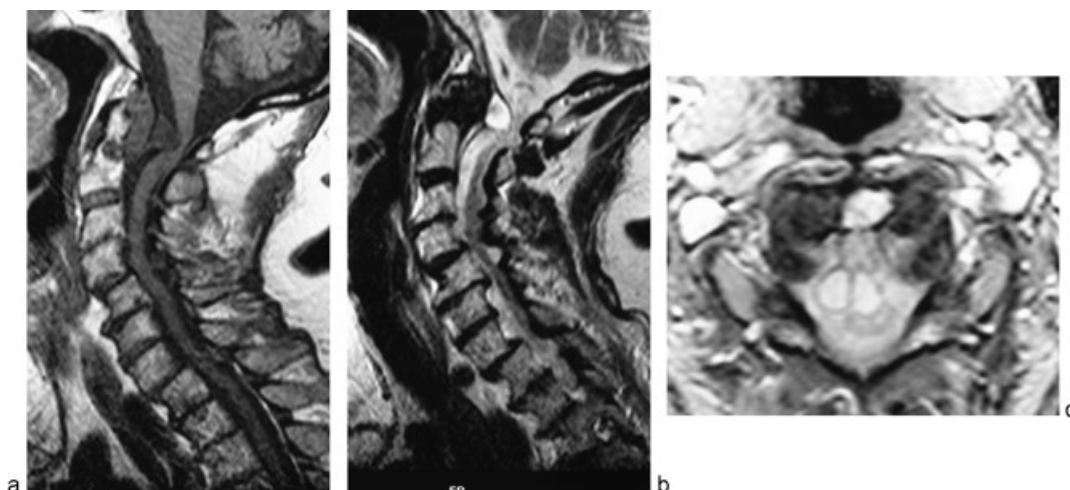

Fig. 2 Magnetic resonance imaging (T1 (a) and T2 (b, c) sequences) demonstrating a retro-odontoid compressive mass with additional bilobulated cystic compression, with myelomalacia noted in the cord.
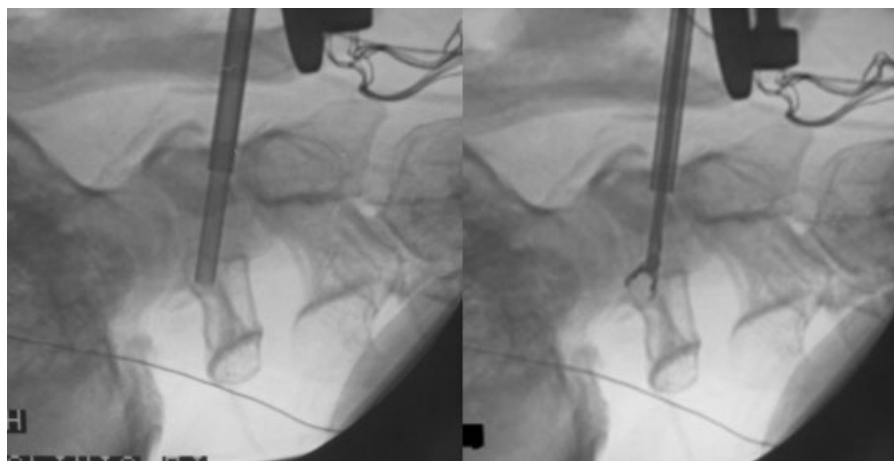
Fig. 3 Intraoperative fluoroscopic images of the sleeved trocar placed through the retro-odontoid mass and into the cyst, and removal of the cystic material.

As our patient had progressive neurologic decline, we felt that surgical decompression was indicated. As this was an elderly 83-year-old patient, we were concerned about the morbidity of either a posterior fusion followed by an anterior transoral approach or a posterior fusion alone. After a lengthy discussion with the patient and her family, we elected to proceed with an anterior minimally invasive transoral aspiration of the cystic lesion to determine the etiology and to decompress the spinal cord within the canal.

Technique

The patient was positioned and intubated supine and we used the Crockard Transoral Instrumentation System to maintain the exposure (Codman & Shurtleff, Inc., Raynham, MA, United States). The uvula was reflected out of the way using a suture and red rubber catheter through the nasopharyngeal cavity. The retropharyngeal soft tissue was visualized, and using computer-assisted navigation, the correct trajectory and entry point was selected. A small 5-mm incision was made into the retropharyngeal tissue anterior to the body of C2. Then using fluoroscopy, a trochar was advanced into and through the body of C2 into the retro-odontoid pannus (►**Fig. 3**). The two cysts were then sequentially aspirated. Two gelatinous aspirates were removed and sent for pathology (►**Fig. 4**). Histology revealed inflammatory cells with positively birefringent rhomboid crystals, consistent with the diagnosis of CPPD. The trochar was removed and the retropharynx closed primarily. Immediate postoperative MRI revealed successful cyst aspiration (►**Fig. 5**).

The patient's postoperative course was complicated by respiratory distress that required reintubation. This resolved with supportive care and the patient was discharged home in stable condition, with resolution of her myelopathy including improved strength and sensation. At 6-month follow-up, the patient was walking with a cane only, and showed continued functional improvement. A repeat MRI at that time revealed complete resolution of the cysts and improved spinal canal diameter (►**Fig. 6**).

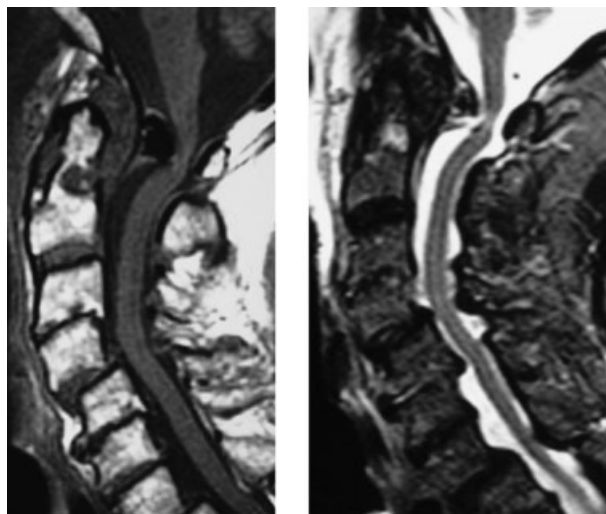**Fig. 4** Gross specimen: gelatinous material removed from cyst.

Fig. 5 Immediate postoperative magnetic resonance imaging (MRI), demonstrating successful cyst aspiration. Note the absence of signal on T2-weighted MRI (right).
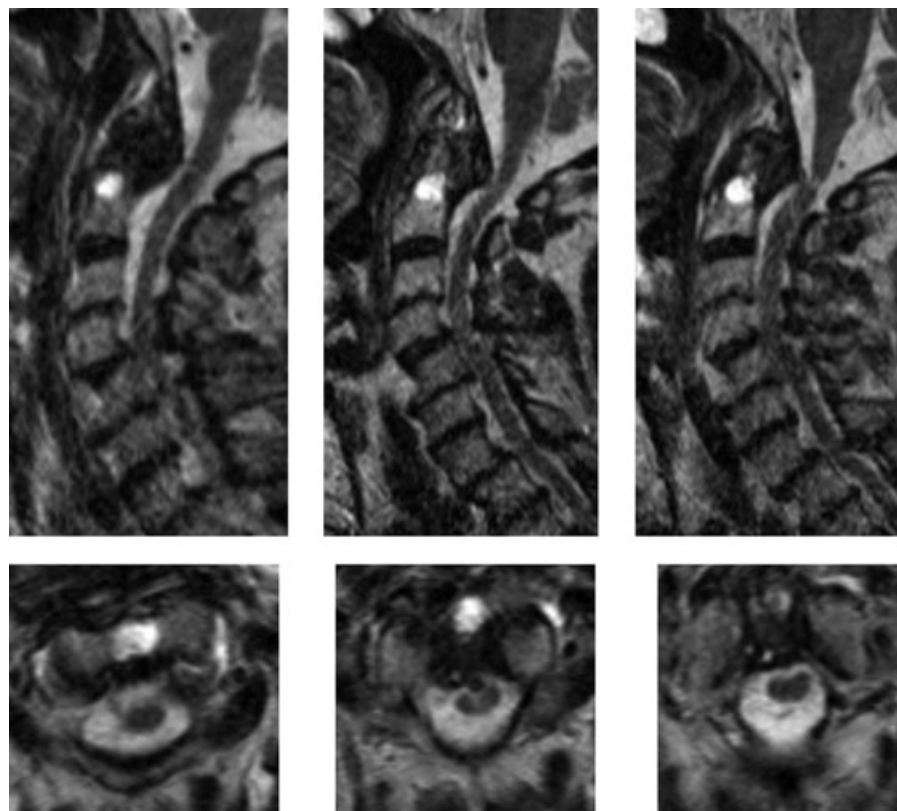
Fig. 6 Six-month postoperative magnetic resonance imaging, axial and sagittal T2 sequences. Note the absence of cyst on images, and significant improvement in cord compression.

Discussion

Cervical myelopathy that occurs as a result of compression at the craniocervical junction is uncommon and may be difficult to diagnose. The etiology of the compression may include rheumatic disease, craniocervical tumors, synovial cyst and CPPD deposition. There are increasing numbers of case reports regarding CPPD deposition at the C1/2 articulation. Although more likely at advanced ages, in 2009, Unlu et al described a case of idiopathic CDS in a young male, causing severe neck pain.[7,8] Clearly, the diagnosis of CPPD should be considered in all patient populations, and may occur in the absence of significant risk factors.

Most cases of CDS arise in patients with known CPPD disease. In 2001, Ryan et al noted that there was asymptomatic calcification of the periodontoid region in half of patients with a known diagnosis of chondrocalcinosis, "suggesting a high incidence of involvement of these structures in CPPD disease."[9] Interestingly, they noted that this deposition did not cause clinical disease in any of their patients. A 2004 case-control study by Finckh et al described that cervical calcifications were found in 69% of CPPD patients (24 of 35), compared with only 11% in controls. In addition, these patients were five times more likely to report neck pain.[10] In 2008, Salaffi et al had similar results with 51% (25 of 49) having periodontoid calcifications in known CPPD. They continued further to describe subchondral cysts or erosion in 40% (10 of 25) of those patients.[3] When clinically significant, the crystal deposition can present either acutely or insidiously, and mimics an inflammatory process.[6] The correct diagnosis may be difficult, as CPPD deposition can occur at any location in the spine. Resnick et al conducted a postmortem study of more than 1,000 spines and found CPPD deposition in the intervertebral discs, interspinous and supraspinous ligaments, ligamentum flavum, posterior longitudinal ligament, sacroiliac joint, transverse atlas ligament, and posterior median atlantoaxial joint.[10,11] There are numerous case reports showing CPPD in various locations in the spine, supporting this.

Atlantoaxial CPPD deposition is best visualized on CT and plain film.[1,12] It was initially described by Bouvet et al as the "crowned dens" in 1985.[13] CT is generally considered the most sensitive modality of visualizing periodontoid calcifications, and the existing literature suggests that diagnosis of CPPD deposition is largely radiographic.[14,15] The largest review of this syndrome was reported in *The Journal of Bone and Joint Surgery* in 2007 by Goto et al.[16] They reviewed 40 patients retrospectively that had characteristic calcium deposits visualized on CT scan; however, none of these patients had histologic confirmation, and none displayed symptomatic disease.

In the English literature, a limited number of cases of CDS have had their diagnosis confirmed. In our case, the CPPD diagnosis was verified via histology, which was especially important due to the absence of significant calcium deposition on CT scan. A cystic odontoid mass, while also previously documented in CPPD, usually lends itself to an alternate diagnosis, including synovial or ganglionic cysts, and transverse ligament degeneration.[3] We attained confirmation with the classic weakly positively birefringent rhomboid crystals

on histological analysis. Our MRI findings were similar to others, with the compressing mass roughly isointense on T1-weighted, and very hyperintense on T2-weighted.[17–20] As with any abnormal presentation, we recommend thorough radiographic studies to rule out other diagnoses.

Treatment for CPPD is symptomatic, and certainly nonsurgical modalities, including medications, should be attempted before surgical intervention when possible, with management tailored to both disease and patient. Of the existing literature on cervical CPPD disease, only a small fraction of cases required surgical intervention due to either pain, myelopathy, or type-2 odontoid fracture.[19,21] Retro-odontoid masses are difficult to reach safely with traditional surgical techniques. Posterior approaches are familiar but require manipulation of the vulnerable spinal cord. Whiteside approach allows access to the dens, but requires significant bony resection, and does not easily allow for instrumentation. A transoral approach allows for direct access from the anterior clivus down to C3 or C4. It also reduces manipulation of the nearby neurovascular structures, compared with posterior and lateral approaches. However, the approach introduces oral flora into the surgical field, increasing the risk of infection. Other complications include upper airway obstruction secondary to edema, velopalatine insufficiency, dental injury, tongue necrosis/edema, odynophagia, dysphagia, meningitis, pharyngeal cellulitis, and TMJ syndrome.[22–24] All of these approaches have significant morbidity and may require surgical stabilization before or after anterior decompression. At the very least, they require a period of modified nutritional support and attention to aspiration and possible swallowing difficulties, as well as observation for potential infection. In 1996, Zünkeler et al described seven cases of histology-confirmed CPPD masses that were posterior to the odontoid process. These patients underwent a transoral–transpharyngeal resection of the anterior arch of C1, odontoid process, and the compressing mass.[25] However, six of the seven patients required posterior fixation at a later time to treat instability.[22]

To minimize complications, different techniques have been used to reduce pharyngeal dissection, such as endoscopic-assisted and minimally invasive approaches. Our technique utilized a cannula to assist with decompression of the retro-odontoid space. This minimizes the risks associated with a transoral approach, with the advantage of direct decompression of the mass. It also allows for resection of the mass with minimal bone resection, which may prevent late instability. The limitations are that this approach is technically demanding, is reliant on ideal intraoperative imaging, and requires a mass that can be removed via the cannula. Despite the previously stated limitations, this approach has proven to be a viable alternative for direct surgical aspiration through the transoral approach, and avoids the need for surgical stabilization.

Conclusion

While CPPD is a rare cause of cervical spinal cord compression, deposition may occur at a variety of locations in the craniocervical junction, and should be included in the differential diagnosis of cervical inflammatory diseases.[26–31] Advanced imaging is required to correctly identify the etiology of the compression, and may offer some clues to the diagnosis, particularly if the dens is surrounded by calcifications. We present a case of CPPD in the retro-odontoid space that caused significant spinal cord compression. We utilized novel minimally invasive transoral aspiration to establish the diagnosis, and affect appropriate decompression with minimal morbidity.

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Commentary

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In this article, the authors report on their management of an elderly patient with a rare form of cervical compressive myelopathy secondary to retro-odontoid calcium pyrophosphate dehydrate (CPPD) deposition. With an overall goal of achieving decompression of the spinal cord, while minimizing the risk of serious morbidity to the patient, the authors chose to perform a transoral aspiration of the associated epidural cystic mass. Although the patient's immediate postoperative course was complicated by respiratory distress syndrome, the authors reported good clinical and radiological outcomes at follow-up.

The presented case presents a variety of challenges both at a disease and patient level. Although the deposition of CPPD is a common cause of peripheral joint arthropathy, it is a rare cause of cervical myelopathy.[1,2] As a result, in comparison to cervical spondylotic myelopathy or myelopathy secondary to ossification of the posterior longitudinal ligament, the natural history and ideal treatment approach to CPPD related myelopathy remains more poorly defined. With respect to patient factors, the advanced age of the discussed patient requires the surgeon to choose an operative approach that balances the desire for complete and permanent spinal cord decompression with the potential for complications and morbidity.

In accordance with these principles, the surgical approach selected effectively minimized the extent of potentially destabilizing tissue dissection and bony resection, obviating the need for posterior placed instrumentation. Furthermore, the cyst aspiration resulted in significant spinal cord decompression with concurrent improvement in symptoms. While ostensibly successful in this case, the described approach carries several potential limitations. First, advancing a trochar in the midline

through diseased bone toward the spinal canal carries the risk of iatrogenic spinal cord injury. An option would have been to undertake an endoscopic or miniopen microsurgical resection. In addition, the chosen method would be limited to those lesions with a major cystic component. Second, the durability of the chosen treatment approach remains in question. As seen on the postoperative imaging, while the epidural lesion is reduced in size as compared with preoperatively, it does remain present. Over time, potential reaccumulation of cyst contents resulting in recurrent symptoms of myelopathy would necessitate the revisiting of potential surgical options. Even if a similar, minimally invasive approach is used, this would expose an even older patient to the risks of repeat surgery and hospital admission. Moreover, CPPD is often associated with degenerative atlantoaxial instability, and continued motion at C1–C2 could also predispose the patient to recurrent cyst formation. That said, at 6 months follow-up, the patient remained clinically and radiologically stable, indicating that the chosen treatment approach may be sufficient throughout the long term.

In summary, the authors are to be congratulated for their innovative approach to the treatment of a rare form of myelopathy in an elderly patient. Although this approach may not be suitable for a younger individual, due to potential concerns surrounding durability, it achieved the goals of attaining cord decompression while minimizing patient harm. With the overall aging of the population, we are likely to see an expanded use of similar minimally invasive techniques when treating spinal disorders amongst the elderly in the coming years.

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Editorial Perspective

EBSJ welcomes the case presentation and the thoughtfully rendered concerns about the technique described by Drs. Wilson and Fehlings. The main purpose of including this article rested in the increasing number of elderly patients who present with disabilities secondary to severe spinal disorders, usually with considerable delays in recognition. The crowned dens syndrome is an example. By showing pertinent images of this case, recognition of this condition —or other storage diseases—will hopefully increase and treatment options will be discussed more knowingly. EBSJ does not endorse the technique described because no conclusions can be drawn from a single case, especially as disease recurrence is a very distinct possibility, as pointed out by Drs. Wilson and Fehlings. Furthermore, this patient quite evidently has diffuse subaxial spondylo-sis with likely contributions on healthy spinal cord func-tion, rendering determination of clinical success over time quite difficult.