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Diagnostic Challenge of a Tumor-mimicking Spinal Pseudogout Lesion: Case Report and Literature Review

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Abstract

Background/Aim: Retro-odontoid pseudotumor (ROP) is a non-neoplastic soft-tissue mass occurring around the C2 odontoid process and is most often associated with rheumatoid atlantoaxial instability. In rare instances, calcium pyrophosphate dihydrate (CPPD) crystal deposition (pseudogout) in the C1-C2 ligaments – a presentation known as "crowned dens syndrome" – can produce a periodontoid mass. Such CPPD-induced lesions usually present with acute neck pain and stiffness, with calcifications visible around the odontoid process on imaging. However, these masses can occasionally enlarge and compress the cervical spinal cord, mimicking a neoplasm and posing a diagnostic challenge.

Case Report: We describe a 77-year-old man with a 3-month history of neck pain radiating to the occiput, progressive hand numbness, and gait disturbance. Magnetic resonance imaging revealed an upper cervical intra-/extradural mass compressing the spinal cord. No atlantoaxial instability was evident on radiographs, and no calcification of a retro-odontoid mass was seen on computed tomography. A presumptive diagnosis of a spinal tumor (*e.g.*, meningioma or nerve sheath tumor) was made. The patient underwent posterior decompression and resection of the lesion. Intraoperative frozen-section pathology revealed birefringent calcium pyrophosphate crystals, consistent with a tophaceous pseudogout. Postoperatively, the patient's myelopathic symptoms improved significantly. By one month, he was ambulatory with a cane and had recovered normal motor strength with rehabilitation.

Conclusion: This case highlights CPPD crystal deposition that can produce a ROP leading to cervical myelopathy even in the absence of radiographically visible calcifications. Awareness of this entity is crucial to avoid misdiagnosis. In an elderly patient with an upper cervical mass, considering CPPD in the differential diagnosis can guide appropriate management and prevent unnecessary interventions.

Keywords: Spinal tumor, retro-odontoid pseudotumor, ROP, calcium pyrophosphate deposition, CPPD, pseudogout.

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Introduction

Retro-odontoid pseudotumor (ROP) is a benign, fibrous mass located posterior to the odontoid process of C2 that can cause spinal cord compression. Classically, ROP is associated with long-standing rheumatoid arthritis (RA), wherein chronic atlantoaxial instability and synovial inflammation lead to pannus formation around the odontoid. However, similar periodontoid masses have been described in patients without RA and attributed to other etiologies such as long-standing degenerative osteoarthritis with C1-C2 instability or various metabolic deposition diseases (1). For example, β2-microglobulin amyloid deposition in patients on long-term hemodialysis can cause a craniocervical pseudotumor at C1-C2 (1), and even rare proliferative synovial lesions such as pigmented villonodular synovitis of the atlantoaxial joint have been reported to mimic ROP (1, 2). These non-rheumatoid causes must be considered when evaluating a periodontoid mass. In addition, pseudotumor-like lesions associated with IgG4-related disease have been reported, and accurate diagnosis with prompt initiation of corticosteroid therapy can prevent unnecessary therapeutic interventions and the progression of irreversible fibrosis (3). Furthermore, due to the heterogeneity in clinical presentation and radiological features, IgG4-related disease is often difficult to differentiate from malignant lesions, and preoperative definitive diagnosis may not always be achievable (4). The diagnosis and treatment planning of spinal cord tumors can be challenging, and clinical decision-making may be particularly complex in certain cases (2-7).

One metabolic crystal arthropathy known to cause ROP is calcium pyrophosphate dihydrate (CPPD) deposition disease, widely recognized for causing pseudogout in peripheral joints. CPPD crystals can deposit in the atlantoaxial ligaments, a condition termed "crowned dens syndrome" when they form a crown-like calcific halo around the odontoid (8). Clinically, crowned dens syndrome typically manifests with acute neck pain, neck stiffness, and sometimes fever in elderly patients, often without significant neurological deficits (8). It is usually managed conservatively with anti-inflammatory medications as symptoms often improve once the acute crystal-induced inflammation subsides (8). In most cases of crowned dens syndrome, radiographic calcifications around the odontoid process on computed tomography (CT) are the hallmark finding, and serious cord compression is uncommon (8).

Rarely, however, CPPD crystal deposition can lead to an enlarging retro-odontoid mass that causes cervical myelopathy. These pseudotumors are non-infectious and non-neoplastic, but on magnetic resonance imaging (MRI) they may appear as soft-tissue masses that are difficult to distinguish from true spinal tumors (9-13). Typically, ROP lesions show an isointense or slightly hypointense signal on T1-weighted MRI and iso- to hyperintense signal on T2-weighted MRI, with variable contrast enhancement. CT can be very helpful in suggesting a CPPD etiology by revealing the characteristic calcifications around the odontoid, but importantly, calcification is not present in all cases. Given the potential for neurologic injury from cord compression, recognizing a CPPD-induced pseudotumor is critical (8). Misidentifying such a lesion as a neoplasm could lead to inappropriate management. We report a case of a CPPD-associated ROP in an elderly patient without RA and without the typical calcifications seen on CT, underlining the difficulties in diagnosis. We also review the relevant literature on this rare condition and discuss its management in light of previously reported cases (9-13).

Case Report

Clinical presentation. A 77-year-old Japanese man presented to our hospital with chronic neck pain and progressive neurological symptoms. He had a 3-month history of posterior neck pain radiating to the left occiput (greater occipital nerve distribution) that did not improve with oral analgesics and physical therapy. Over the same period, he noticed increasing numbness and tingling in both hands, clumsiness in fine finger movements, and a

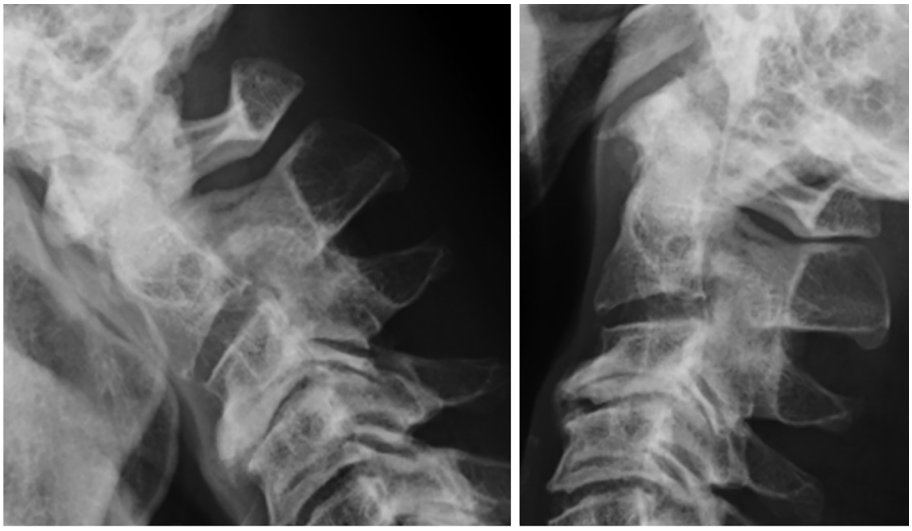

Figure 1. *Patient X-ray images. Plain cervical radiographs in flexion (left) and extension (right) showed normal alignment of C1 and C2, with no evidence of atlantoaxial instability (no C1-C2 subluxation on dynamic views).*

mild unsteadiness of gait. He denied any recent trauma. He also had no history of RA or any crystal deposition disease (gout or pseudogout). His past medical history was unremarkable for musculoskeletal or metabolic disorders. Routine blood tests showed no systemic inflammation or infection: C-reactive protein was 0.14 mg/dl (normal), and white blood cell count was 4,100/μl. Metabolic panels were also within normal limits.

On neurologic examination, the patient exhibited signs of cervical myelopathy. He had hyperreflexia in the lower extremities, and bilateral Hoffmann's signs were present. Finger dexterity was impaired (in a 10-second rapid grip-and-release test, only 13 hand grips were performed on each side, reflecting slowed fine motor function). Mild weakness was noted in the intrinsic hand muscles. His gait was broad-based and slightly ataxic, though he could walk unassisted for short distances. The Japanese Orthopedic Association score (JOA) for cervical myelopathy was 7.5 out of 17, indicating a severe myelopathic deficit.

Imaging studies. Plain cervical radiographs (dynamic flexion-extension views) showed normal C1-C2 alignment with no atlantoaxial instability (no evidence of C1-C2 subluxation) (Figure 1). MRI of the craniovertebral junction revealed an abnormal mass lesion dorsal to the C2 odontoid process that was causing marked compression of the upper cervical spinal cord. The mass was isointense relative to spinal cord on T1-weighted MRI and hyperintense on T2-weighted MRI, consistent with a soft-tissue lesion (Figure 2). The lesion measured approximately 12 mm in craniocaudal length and 10 mm in thickness and caused significant effacement of the subarachnoid space. Notably, there was no clear cerebrospinal fluid cleft around the mass on MRI, making it difficult to determine whether the lesion was intradural or extradural. To further evaluate the lesion, a high-resolution contrast-enhanced CT scan of C1-C2 was obtained, which showed a soft-tissue mass posterior to the odontoid, without significant contrast enhancement and without calcific densities or ossifications within the mass or around the odontoid process (Figure 3).

Based on these imaging findings, an upper cervical extradural tumor was considered the most likely diagnosis. The location and MRI characteristics raised the possibility of a benign neoplasm such as an atypical meningioma or a nerve sheath tumor of the C1 nerve root with an unusual extradural extension. An inflammatory pseudotumor was also included in the differential diagnosis, though the

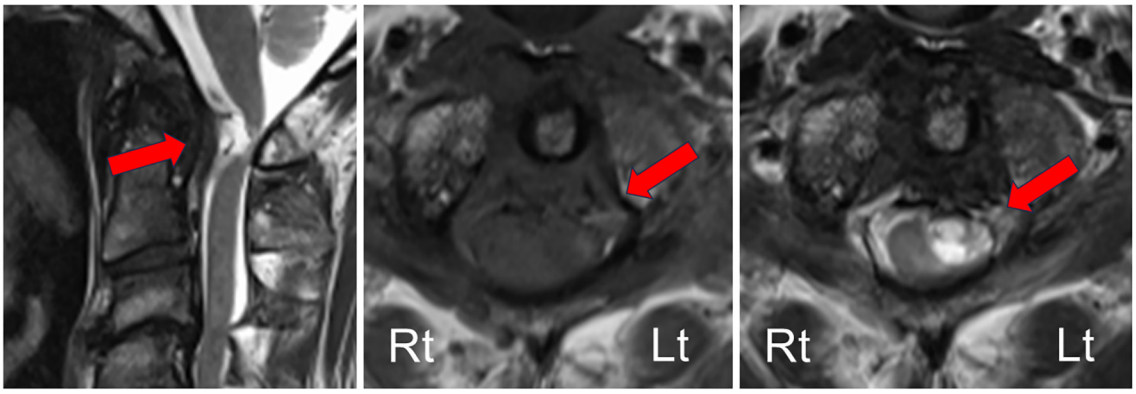

Figure 2. *Patient magnetic resonance imaging (MRI) images. Preoperative MRI showed the retro-odontoid pseudotumor (arrow) compressing the cervical medulla. Left: Sagittal T2-weighted image showed a hyperintense mass posterior to the C2 odontoid, causing marked indentation of the cord. Middle: The mass was isointense to spinal cord on T1-weighted MRI. Right: Hyperintense mass on axial T2-weighted image at the C1-C2 level showed that the lesion was compressing the spinal cord.*

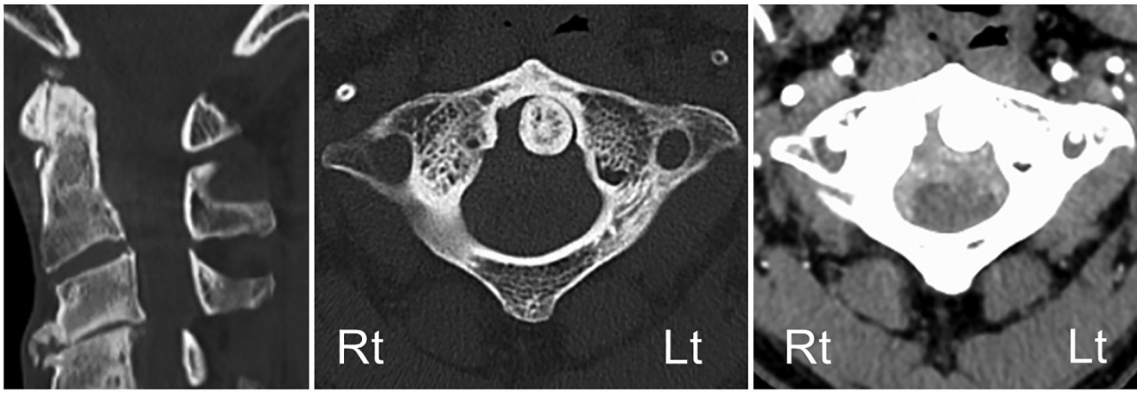

Figure 3. *Patient computed tomography (CT) scan. The scan showed no calcification, and a mass without significant contrast enhancement.*

absence of systemic inflammation and lack of response to initial conservative therapy made an active inflammatory process less likely. There were no radiologic signs of infection (no abscess, bone destruction, or disc-space involvement), and the patient was afebrile with normal inflammatory markers, arguing against an infectious process such as granulomatous pachymeningitis. Given the patient's progressive myelopathy and the uncertainty of the diagnosis, we elected to proceed with surgical intervention for decompression and tissue diagnosis.

Surgical intervention. A posterior surgical approach was undertaken to decompress the spinal cord and obtain tissue for diagnosis. Through a midline posterior cervical incision, a C1 laminectomy and partial C2 laminectomy were performed to expose the region of the compression. Upon opening the posterior dura mater, a soft, grayish-yellow mass was encountered ventral to the spinal cord at the C1-C2 level. The lesion was found to be an intradural-extramedullary pseudotumor originating from the inner (ventral) surface of the dura and compressing the cord anteriorly, as shown in Figure 4. It did not have the firm consistency of a typical fibrous tumor; instead, when a small longitudinal incision was made in the mass capsule, a gelatinous, turbid material was released (Figure 4). This suggested that the lesion was a granulomatous or tophaceous deposit rather than

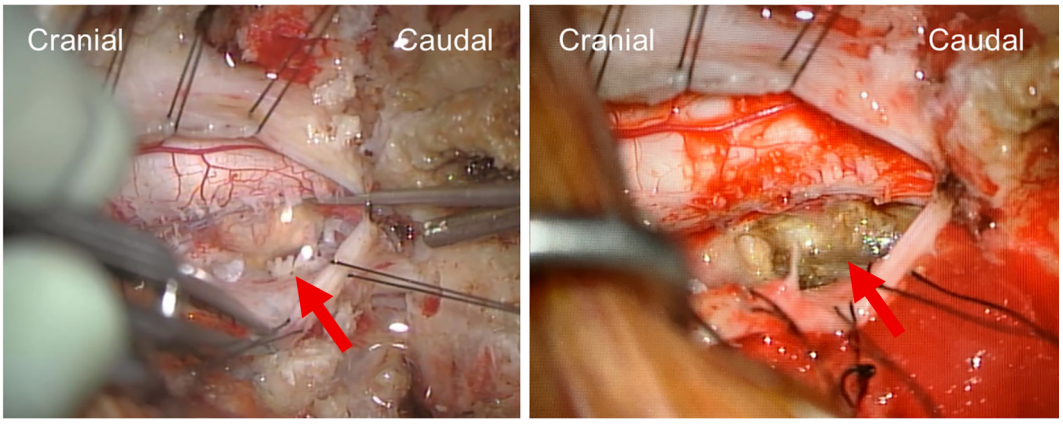

Figure 4. *Intraoperative findings. Left: After C1 laminectomy and dural opening, a yellow-gray pseudotumor (arrow) became visible on the ventral aspect of the dural sac, displacing the spinal cord posteriorly. Right: After incision of the pseudotumor's capsule, gelatinous material mixed with chalky deposits was removed (arrow indicates the released tophaceous material). The lesion, along with the involved inner layer of dura, was excised en bloc for histopathological analysis.*

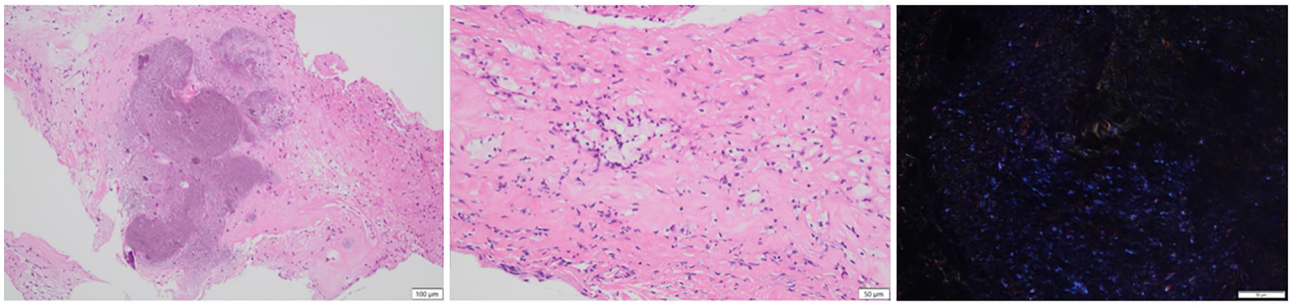

Figure 5. *Hematoxylin and eosin (H&E) staining. The lesion was confirmed to be a pseudogout (CPPD) pseudotumor. Left: Low-power H&E image (scale bar=100 μm) shows several purple-staining nodules of calcified material (CPPD crystal deposits) surrounded by fibrous stroma. Middle: Higher-power view (scale bar = 50 μm) shows chronic inflammatory changes with fibrosis and clusters of foamy (lipid-laden) macrophages around the crystal deposits. Right: Under polarized light microscopy, rhomboid-shaped or short rod-shaped CPPD crystals were observed, showing weak positive birefringence. The histopathological features are consistent with a tophaceous CPPD deposition.*

a solid neoplasm. We achieved gross total resection of the mass by carefully dissecting it away from the surrounding dura and spinal cord. The outer layer of the dura was preserved to maintain some structural integrity as the lesion appeared to arise from the inner dural layer. As no overt atlantoaxial instability was observed intraoperatively after decompression, we decided that immediate C1-C2 fusion was not required.

Histopathological findings. Intraoperative frozen section pathology was performed on the excised tissue, which showed dense fibrous and inflammatory tissue with no evidence of malignancy. Notably, when examined under polarized light, the specimen contained numerous positively birefringent, rhomboid-shaped crystal deposits. These microscopic findings were highly suggestive of CPPD crystal deposition, consistent with pseudogout (Figure 5). The final permanent histology sections (stained with hematoxylin and eosin) confirmed the diagnosis of a tophaceous pseudogout (CPPD) mass. The lesion was composed of nodular aggregates of pale basophilic crystal material surrounded by proliferative

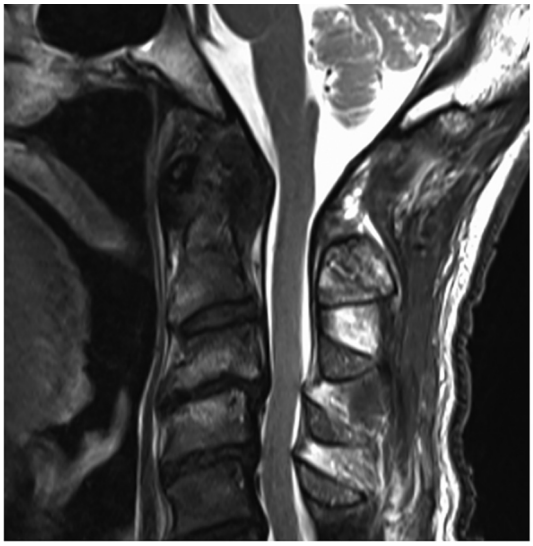

Figure 6. *Patient follow-up magnetic resonance imaging (MRI) scan. A follow-up MRI scan was performed after 3 months and showed effective relief of spinal cord compression.*

fibrous connective tissue and an infiltrate of chronic inflammatory cells, including many foamy macrophages and multinucleated giant cells. There was no evidence of infection (no organisms on special stains) and no neoplastic cells.

Postoperative course. With intensive physiotherapy, the patient's gait ataxia also improved; at two weeks, he was able to ambulate with a walker, and by one month, he walked independently with a cane for balance. His occipital headaches and neck pain resolved completely. There were no signs of cranial nerve dysfunction or delayed instability. A follow-up MRI at 3 months confirmed complete removal of the retro-odontoid mass and excellent spinal cord decompression, with no residual compression or signal changes in the cord (Figure 6). The postoperative JOA score for cervical myelopathy was 16 out of 17 and improved compared to before surgery.

Informed consent statement. Informed consent was obtained from the patient for the publication of this case report and accompanying images.

Discussion

Our case illustrates an uncommon cause of upper cervical cord compression: a CPPD-induced ROP without radiographically visible calcifications. This entity is part of the broader spectrum of ROPs, which are usually recognized on MRI as mass lesions adjacent to the odontoid. In the context of degenerative (non-rheumatoid) ROPs, these masses often consist of dense fibrocartilaginous tissue with minimal inflammation, and they typically appear iso- to low-intense on both T1- and T2-weighted MRI (occasionally with small regions of higher T2 signal) with none or minimal contrast enhancement. The primary treatment for a symptomatic degenerative ROP (not associated with infection or crystals) is to address atlantoaxial instability. Stabilization of C1-C2 with posterior fusion frequently leads to regression or even complete disappearance of the pseudotumor over time, as eliminating pathologic motion reduces the inflammatory stimulus for pannus formation (9). Several reports have documented that C1-C2 posterior fusion alone (without direct resection) can result in shrinkage of the mass and neurological improvement (9, 10). In cases in which the ROP stems from other causes such as dialysis-related β2-microglobulin amyloid deposition or chronic crystal deposition, stabilization may also help if instability is a contributing factor, but optimal management is less clear due to the rarity of such cases (1). It is important to note that not all periodontoid masses require fusion as the treatment must be tailored to the underlying cause and the presence or absence of atlantoaxial instability.

Among the metabolic causes of pseudotumor, CPPD crystal deposition disease is a recognized but infrequent culprit in the cervical spine. CPPD in the atlantoaxial region typically presents as crowned dens syndrome, which is characterized by episodic neck pain, stiffness, and fever rather than chronic myelopathy. Most patients with crowned dens syndrome respond well to conservative therapy (nonsteroidal anti-inflammatory drugs, colchicine, and/or corticosteroids), and surgical intervention is usually

unnecessary unless there is significant cord compression. In crowned dens syndrome, CT scans usually show calcifications in the transverse ligament or periodontoid tissues that form a "crown" around the dens. MRI findings in CPPD-related ROP can be non-specific, often showing an isointense or slightly hyperintense mass on T2-weighted images and isointense signal on T1, which can closely resemble other lesions. Yurube *et al.* (12) reviewed a case of upper cervical myelopathy due to combined degenerative changes and CPPD deposition; their patient's MRI showed a compressive retro-odontoid mass with mixed intensity, and CT clearly revealed calcifications, clinching the diagnosis of CPPD pseudotumor. In general, finding calcification on CT greatly increases suspicion of a crystal deposition pseudotumor (either CPPD or occasionally gout). Interestingly, a literature review by Klineberg *et al.* (13) noted only one prior case of CPPD-associated ROP that presented with pronounced T2 hyperintensity on MRI at the site of cord compression. Even in that case, the diagnosis was supported preoperatively by the presence of calcified deposits around the dens on CT. By contrast, our patient's CT imaging showed no calcification whatsoever in the periodontoid region, which obscured the true etiology until pathological analysis. This suggests that CPPD pseudotumor cannot be radiologically excluded by the absence of the classic calcifications – a critical point for clinicians to recognize.

In the present case, the lack of CT calcifications and the MRI appearance of the lesion led us to strongly consider an intradural extramedullary tumor (such as meningioma or nerve sheath tumor) in the differential diagnosis. Consequently, we proceeded with surgical resection to both decompress the cord and establish a diagnosis. Only after histopathological examination did the true nature of the lesion become evident as a CPPD-induced inflammatory pseudotumor. In hindsight, had CPPD crowned dens syndrome been suspected preoperatively, a trial of conservative treatment with anti-inflammatory medications might have been considered, potentially avoiding surgery. There have been reports of patients with CPPD periodontoid pannus and mild symptoms improving with medical management alone, particularly when the diagnosis is recognized early (8). However, in our patient's case, the severity of spinal cord compression and progressive myelopathy made surgical decompression a reasonable and ultimately successful approach. This case underscores the importance of including crystal deposition disease (pseudogout) in the differential diagnosis of retro-odontoid masses in elderly patients – especially when the patient has no history of RA and imaging findings are atypical. Awareness of this entity can prompt appropriate diagnostic tests (such as a careful CT review for calcifications or even a biopsy) and guide management. An accurate diagnosis of CPPD pseudotumor might favor conservative management if neurologic impairment is mild or inform the surgical strategy (for example, combining decompression with stabilization if instability is present or targeting the crystal-induced inflammation postoperatively).

Conclusion

We report a rare case of cervical myelopathy caused by a CPPD-induced ROP without the classic radiographic signs of crowned dens syndrome. This case highlights the diagnostic challenges when a periodontoid pseudogout mass lacks visible calcification on CT as it can be misinterpreted as other pathologies. Clinicians should maintain a high index of suspicion for metabolic crystal deposition disease in elderly patients presenting with an atlantoaxial mass lesion, even if imaging is not characteristic. Recognizing this condition is crucial as it can broaden the treatment options (conservative *versus* surgical) and prevent misdiagnosis. Early identification of CPPD-related pseudotumor may allow for medical management in appropriate cases, whereas surgery can lead to good outcomes in patients with significant myelopathy by achieving prompt neural decompression.

Conflicts of Interest

The Authors have no conflicts of interest to declare in relation to this study.

Authors' Contributions

I.T and K.F were involved in patient management and data acquisition. N.N and M.F contributed to radiological evaluation. H.T and H.S drafted the manuscript. T.S supervised the study and critically revised the manuscript. All Authors read and approved the final version of the manuscript.

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Artificial Intelligence (AI) Disclosure

During the preparation of this manuscript, a large language model (ChatGPT, OpenAI) was used solely for language editing and stylistic improvements in select paragraphs. No sections involving the generation, analysis, or interpretation of research data were produced by generative AI. All scientific content was created and verified by the authors. Furthermore, no figures or visual data were generated or modified using generative AI or machine learning–based image enhancement tools.

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