



Case report

Symptomatic retro-odontoid pseudotumor causing calcium pyrophosphate dihydrate deposition combined with multilevel cervical spondylotic myelopathy

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ABSTRACT

Introduction: Symptomatic retro-odontoid pseudotumor (ROP) caused by cervical compression and myelopathy is rare. Pathological diagnosis is recommended for differential diagnosis including the following: inflammatory disease, primary bone tumor, metastatic disease and calcium pyrophosphate dihydrate deposition (CPPD) also known as “crowned dens syndrome”. The authors report a rare case of ROP caused by CPPD deposition combined with multilevel cervical spondylotic myelopathy (MCSM) which was treated by tumor resection using a transoral approach combined with posterior decompression and fusion.

Case presentation: A 66-year-old male presented with progressive neck pain and spastic gait with no history of trauma. Radiographic imaging revealed degenerative change involving the atlanto-axial and atlanto-occipital joints with calcified enhancing soft tissue around the odontoid process causing cord compression and cervical instability at the C1-C2 level combined with MCSM and spinal cord compression at C3 to C7. Microscopic assisted transoral tumor resection combined with posterior decompression and fusion was performed at the occiput to T2. The pathology report describes a rhomboid-shaped crystal caused by calcium pyrophosphate dihydrate deposition (CPPD) disease. At the 6-month follow-up following the operation, the patient's neck pain and spastic gait were improved compared to the preoperative examination.

Discussion: Cervical compression and myelopathy from ROP causing CPPD combined with MCSM is rare. Pathology diagnosis and surgical management are highly recommended.

Conclusion: In this case, a combined surgical approach: tumor resection using a transoral approach and a posterior approach for decompression and fusion at occiput to T2 was an effective option for this condition.

1. Introduction and importance

Retro-odontoid pseudotumor (ROP) caused by cervical compression and myelopathy is uncommon [1]. ROP is a non-neoplastic lesion associated with calcium pyrophosphate dihydrate deposition (CPPD) with periodontoid calcification as known as “crowned dens syndrome” [1]. Degenerative cervical spondylotic myelopathy (CSM) is part of the aging process which results in degenerative changes in the elderly that can cause multi-level spinal cord compression [2]. ROP associated with CPPD deposition combined with multilevel cervical spondylotic myelopathy (MCSM) is a rare case. The microscopic assisted transoral approach is a minimally invasive tumor resection surgery for pathological diagnosis [1]. This report presents a case of ROP-associated

MCSM treated by microscopic assisted transoral tumor resection combined with posterior decompression and fusion performed at the occiput to T2. This work has been reported in line with SCARE criteria [3].

2. Case presentation

A 66-year-old male presented with complaints of progressive neck pain and spastic gait for 1 month. He had no history of rheumatoid arthritis, pseudogout, cervical spine trauma or other predisposing factors. The patient stated that he had no underlying disease, no smoking, no alcohol consumption or recreational drug use. Physical examination found his mental status and vital signs were normal. He had bilateral radicular neck pain without upper or lower extremity weakness (motor

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power grade V). His gait and balance were spastic and he was unable to walk toe-to-heel. Upper and lower extremity deep tendon reflexes were found hyperreflexic. Special examinations showed abnormalities in the inverted radial reflex, Hoffmann's sign, Lhermitte's sign and scapulohumeral reflex (Shimizu test). Rectal examination found normal sphincter tone.

Radiographic imaging showed severe cervical spondylosis with periodontoid calcifications (Fig. 1A, B). Patient's differential diagnosis included the following: inflammatory disease, primary tumor or metastatic disease. Magnetic resonance imaging (MRI) (Fig. 1C, D) revealed degenerative change involving the atlanto-axial and left atlanto-occipital joints with calcification enhancing soft tissue around the odontoid process causing spinal cord compression with radiographic instability and compressive myelopathy at the C1-C2 level. Degenerative changes of the remaining cervical spine with moderate C3-C4 and C6-C7 spinal cord compression, severe bilateral C3-C4 to C6-C7 neural foramina. MRI results suggested a retro-odontoid mass as well as a less likely diagnosis of CPPD or crowned dens syndrome. Based on that finding, an anterior transoral approach for resectioning the retro-odontoid mass and a posterior approach for occipito-cervico-thoracic fixation and fusion with decompressive laminectomy of C3 to T2 were chosen. The patient was receiving under general anesthesia via nasotracheal tube. He was placed in the supine position and a special retractor was applied. An orogastric tube (5-French feeding tube) was inserted through the nasopharyngeal cavity and sutured to the uvula. The retropharyngeal soft tissue (Fig. 2A) was visualized after orogastric tube upward traction upward traction had been applied to the orogastric tube.

After dissecting the anterior longitudinal ligament (ALL) (Fig. 2B) and the anterior arch of C1, intra-operative findings included soft tissue inflammation around the anterior odontoid (Fig. 2C). Resection of the retro-odontoid mass was performed under microscopic assistance (Fig. 2D) and tissue was sent for histopathology diagnosis. The patient was placed in the prone position for occipito-cervico-thoracic fixation and fusion with decompressive laminectomy of C3 to T2 (Fig. 3A, B) by an experienced spine surgeon (SP).

Postoperative radiographic films showed cervical alignment, rods and screws were in good position (Fig. 3C, D). The pathology report included rhomboid-shaped crystals caused by calcium pyrophosphate dihydrate deposition disease (CPPD) (Fig. 4). There were no postoperative complications. The patient was given colchicine to prevent further attacks and was started on a rehabilitation program. At the 6-month follow-up, the patient's neck pain and spastic gait were improved compared to the preoperative examination. The patient was satisfied with the outcome of the treatment.

3. Clinical discussion

A review of the literature found that CPPD is the most common form of crystal-associated arthropathy in the elderly [2,4]. However, CPPD of the cervical spine is asymptomatic especially in the atlantoaxial and periodontoid areas [1]. Bouvet et al. reported visualizing periodontoid calcifications and described it as "crowned dens" in 1985 [5]. The standard diagnostic tools for detecting and visualizing CPPD include plain film, computed tomography (CT) scans and MRI. Pathology diagnosis is recommended for differential diagnoses, including inflammatory disease, primary bone tumor, metastatic disease and CPP deposition [1,6]. Histology diagnosis of CPPD can reveal positively birefringent rhomboid crystals. Zünkeler et al. reported the outcomes of seven patients who had undergone a transoral resection of the anterior arch of C1, the odontoid process and the compressing mass for pathological diagnosis [7]. However, all those cases required a second operation, posterior fixation, due to spinal instability.

The study subject had symptomatic ROP causing CPPD concomitant with MCSM, cervical compression and myelopathy, which is a very rare condition. Surgical management of retro-odontoid masses is challenging in terms of both preoperative planning and of surgical techniques. Posterior approaches are less complicated but require manipulation of the vulnerable spinal cord for resection of the periodontoid mass which can increase the risk of spinal cord damage [1,7]. A transoral approach provides direct access to the anterior of the atlantoaxial and odontoid for removal of the tumor mass, but it introduces the risk of oral flora infection in the surgical field [1]. However, this case involved ROP with MCSM C3 to C7 that required adequate decompression and fusion. We used a combined transoral and posterior approach with microscopic assistance and adequate intravenous antibiotics to prevent oral bacterial infection.

Case of an ROP patient with MCSM C3 to C7 are extremely rare involves challenging preoperative surgical treatment. In this case, the treatment resulted in a successful outcome. Using a microscopic-assisted transoral tumor resection for anterior decompression and pathological diagnosis combined with a posterior approach for decompression (cervical laminectomy) and fusion (occiput to T2) is associated with good results and outcome.

4. Conclusions

Cervical instability and spinal cord compression caused by a retro-odontoid mass combined with MCSM is rare. Combined approach including tumor resection using a transoral approach and posterior approach for decompression and fusion at occiput to T2 is an effective treatment that should be an option for this condition.

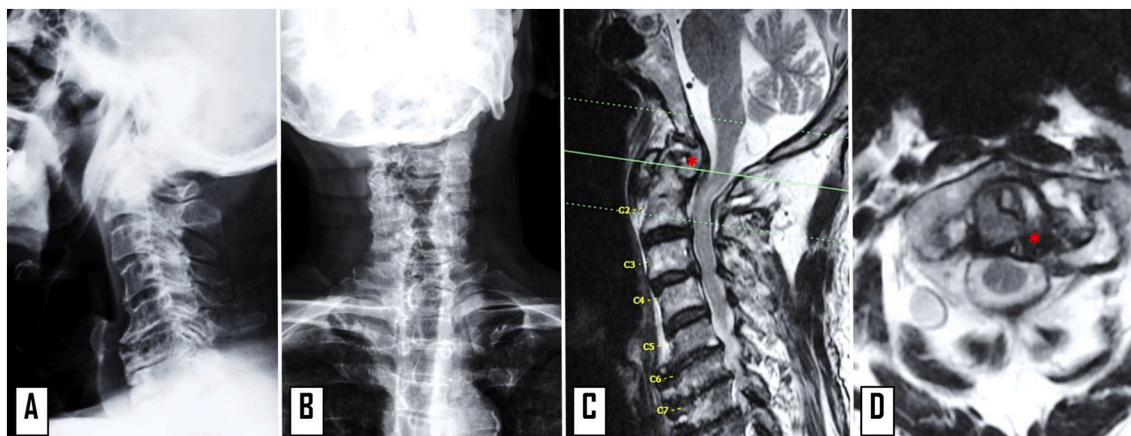


Fig. 1. Pre-operative radiographic X-ray lateral- (A) and AP-view (B), retro-odontoid mass (red asterisk) in MRI sagittal view (C) and axial view (D).

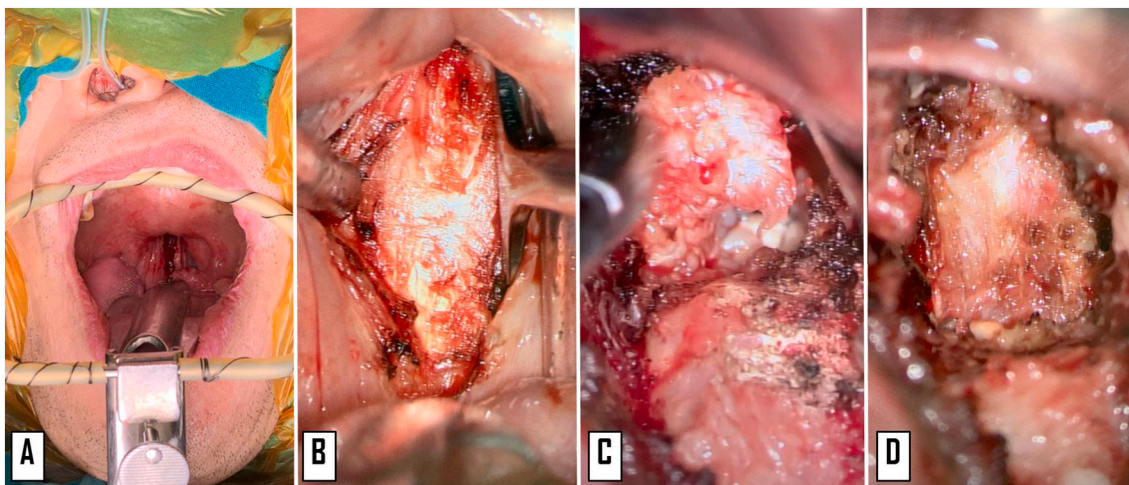


Fig. 2. Intra-operative transoral approach (A), anterior longitudinal ligament (ALL) (B), peri-odontoid tissue (C) and resection (D).

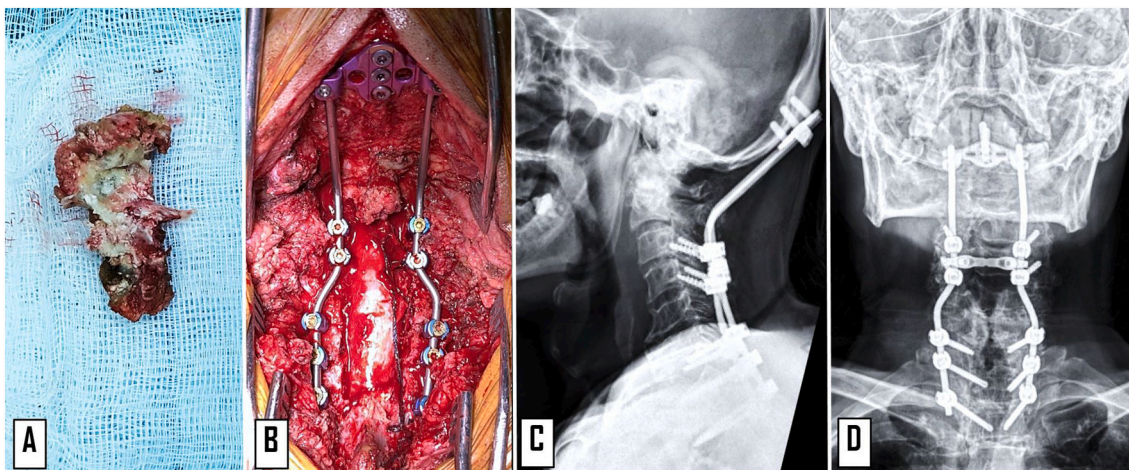


Fig. 3. Intra-operative findings of abnormal soft tissue (A), posterior occipito-cervico-thoracic instrumentation and fusion with compressive laminectomy C3 to T2 (B). Post-operative radiographic X-ray lateral view (C) and AP-view (D).

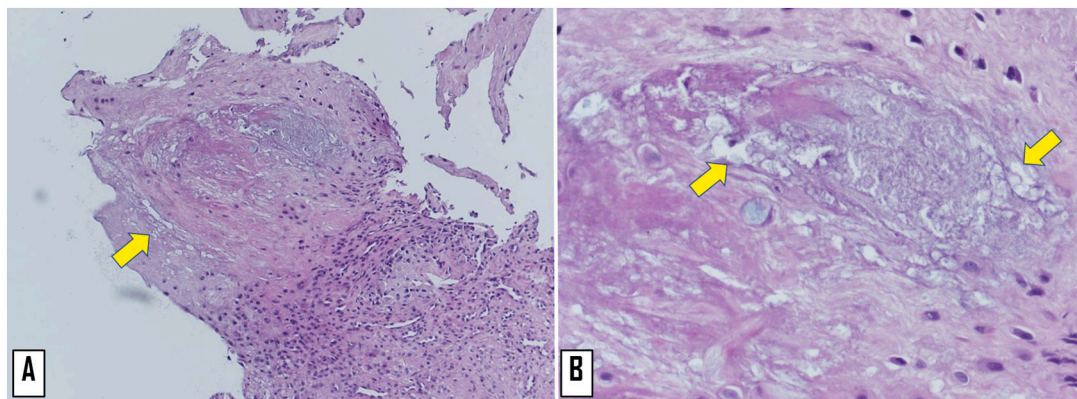


Fig. 4. Histological findings: hematoxylin and eosin (H&E) stain ($\times 100$) showed chondroid metaplasia (arrows) (A), H&E stain ($\times 400$) showed multiple tiny foci of rhomboid shape crystals (arrow) caused by calcium pyrophosphate dihydrate deposition disease (CPPD) (B).

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Ethical approval

This case report was approved by the Institutional Review Board,

Faculty of Medicine, Chiang Mai University.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in Chief of this journal upon request.

Author contributions

Suthipas Pongmanee (SP): Resources, Data curation, Review & editing, Supervision.

Sitthikorn Kaensuk (SK): Resources, Data curation.

Worapat Suppagornmongkol (WS): Resources, Data curation.

Wongthawat Liawrungrueang (WL): Review literature, Conceptualization, Methodology, Visualization, Writing-original draft, Editing and revision the final version for publication.

Registration of research studies

None.

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Declaration of competing interest

None.

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