CASE REPORT

Recovery After Cervical Decompression Surgery for the Treatment of Crowned Dens Syndrome Causing Progressive Neurological Decline: A Case Report

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Introduction

Traumatic injury to the spinal cord can be divided into complete and incomplete subtypes, based on complete or partial absence of neurologic function below the level of injury. Incomplete spinal cord injury has been shown to occur following cervical hyperextension or flexion injury in the setting of preexisting cervical spinal stenosis. The broad spectrum of symptoms from incomplete spinal cord injury includes weakness and sensory deficits potentially affecting upper and lower extremities, as well as bowel, bladder, and sexual dysfunction in severe cases. The most common type of incomplete spinal cord injury is central cord syndrome, which is characterized by a more severely impaired motor function in the upper than the lower extremities [1, 8, 10, 13, 16]. Progressive neurological deficit has also been reported to occur due to C1–C2 pannus formation in the setting of rheumatic disease [4, 12, 17, 19, 20].

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Crowned dens syndrome (CDS) is a rare but known cause of decreased cervical range of motion and severe axial neck pain, in the setting of calcium deposition around the odontoid, for which medical treatment with nonsteroidal anti-inflammatory drugs and/or steroids has been reported [2, 7]. Patients with CDS may present with symptoms of cervical myelopathy caused by a mass containing calcium pyrophosphate dihydrate (CPPD) at the atlantoaxial articulation and/or in the transverse ligament and/or in the ligamentum flavum [3, 9, 11, 14, 15]. According to the findings of Goto et al. evaluating 2,023 patients with neck pain as the chief complaint, 40 patients were identified with CDS, indicating a prevalence of 2.0% [7]. Chang et al. reported an overall prevalence of radiographically evident atlantoaxial CPPD deposition of 12.5%, which was increasing with age [5].

However, severe and gradually progressive neurological deterioration due to an isolated large mass consisting of calcium pyrophosphate at the C1–C2 level causing functional quadriplegia is rare, and, therefore, the optimal treatment remains to be determined. Surgical decompression and stabilization may alleviate the compression on the cervical spinal cord [11], yet the potential for neurological recovery remains to be further elucidated. We report the case of an 87-year-old male diagnosed with quadriplegia resulting from CDS who made an excellent recovery following surgical decompression.

Case Report

An 87-year-old male patient presented in a wheelchair with a 2-year history of gradual decline in mobility and symptoms of cervical spondylotic myelopathy. Subsequently, he experienced a more rapid decline in neurological function over a 4-month period after hitting the side of his head sustained during a low-energy fall from standing. Interestingly, the patient was discharged from the hospital neurologically at baseline, but in the ensuing days to weeks, he developed progressive neurological deficits. Four months after this

Table 1 Motor strength examination at initial office consultation

Upper Extremity	Right	Left
Deltoid	0/5	0/5
Biceps	1/5	4-/5
Triceps	0/5	3/5
Wrist extension	0/5	4/5
Wrist flexion	0/5	4/5
Finger flexion	2/5	4-/5
Interossei	1/5	2/5
Lower Extremity	Right	Left
Hip flexion	2/5	4-/5
Quadriceps	3/5	4-/5
Hamstring	3/5	3/5
Extensor hallucis longus	0/5	3/5
Tibialis anterior	0/5	3/5
Gastrosoleus	2/5	3/5

low-energy fall, the patient presented to our institution with hypoesthesia to the knee bilaterally and complete dense paraesthesias in both hands. Furthermore, motor testing revealed markedly decreased strength in all major muscle groups of the upper and lower extremities, consistent with function quadriplegia (Table 1). The patient reported increased urinary frequency and urgency, but denied any fecal or urinary incontinence. Apart from hypertension, the review of systems was unremarkable.

Plain radiographic imaging studies of the cervical spine demonstrated loss of lordosis, multilevel spondylosis, as well as degenerative spondylolisthesis (Fig. 1). The computed tomography (CT) exams depicted a large calcified retrodental mass anterior to the thecal sac along with calcification in the ligamentum flavum (Fig. 2). Magnetic resonance imaging studies demonstrated a large retrodental mass anterior to the thecal sac and a fluid-containing complex mass posterior to the thecal sac at the level of C1–C2 both compressing the cervical spinal cord. Also noted were

Fig. 1. Lateral X-ray performed at first office consultation, depicting loss of cervical lordosis, multilevel spondylosis, anterolisthesis at C3/C4 and C6/C7, as well as minimal retrolisthesis at C5/C6. The sagittal alignment at C1/C2 is normal.

Fig. 2. Preoperative sagittal CT image depicts calcification within a large retrodental mass and the ligamentum flavum.

fluid superior and posterior to the dens, severe multilevel cervical spinal stenosis from C1–C2 to C5–C6, high T2 signal intensity within the spinal cord at the level of C2, and acute angular cord compression at C1–C2 (Fig. 3). Preoperative CT angiography was prohibited by patient's baseline chronic renal insufficiency; however, preoperative magnetic resonance angiography demonstrated absence of the left vertebral artery (Fig. 4).

Due to the severity and further progression of the patient's symptoms in the face of cervical cord compression, surgical decompression was performed. Access to the posterior cervical laminae (C1 to C7) was gained via midline posterior cervical incision. Following exposure from the occiput to C7, the patient underwent posterior decompression (C1 to C7), lateral mass fixation at C1, stabilization with C2 pedicle screws, and lateral mass fixation from C3 to C7 (Fig. 5). Following laminectomy of C2, crystalline

Fig. 3. Preoperative MRI T2 sagittal image demonstrates a large retrodental mass anterior to the thecal sac and a fluid-containing complex mass posterior to the thecal sac at the level of C1/C2 both compressing the cervical spinal cord, fluid superior and posterior to the dens, severe multilevel cervical spinal stenosis from C1/C2 to C5/C6, high T2 signal intensity within the spinal cord at the level of C2, and acute angular cord compression at C1–C2.

Fig. 4. Preoperative magnetic resonance angiogram depicting absence of the left vertebral artery. The *arrow* points to the right vertebral artery.

deposits were visible at the dorsal aspect of the thecal sac between C1 and C2. After resection of the posterior arch of C1, a large mass with deposits of chalky white material approximately 2.5 cm in diameter expanded dorsally around the thecal sac (Fig. 6). Histopathologic examination of the resected mass revealed low-cellularity collagenous tissue containing large deposits of positively birefringent rhomboid crystals with morphologic features of calcium pyrophosphate indicating CPPD disease (Fig. 7a), in addition to perivascular deposits of dense acellular material which stained positive for Congo red and showed apple green birefringence under polarized light (Fig. 7b, c).

In the immediate postoperative setting, the patient presented with marked improvement to his prior baseline of motor strength and a restored ability to ambulate with a walker. At an office consultation at 9-months postoperatively, the patient stated great improvement, in addition to being

Fig. 5. Intraoperative lateral X-ray, illustrating posterior arthrodesis from C1 to C7.

able to use the computer and feed himself since the time of surgery. The patient described intermittent left-sided neck pain as well as frontal headache, which was not present before the surgery. His neurological examination revealed a 5/5 motor strength in his arms with the exception of the right wrist extensor and right grip (4/5). Apart from his baseline right foot drop, the patient demonstrated a 5/5 lower extremity motor strength bilaterally.

Discussion

We present a unique case of an octogenarian with a progressive neurological decline resulting in functional quadriplegia caused by isolated calcium pyrophosphate deposition at the C1–C2 level, who achieved marked neurological recovery following late decompression and stabilization.

Incomplete spinal cord injury has been shown to occur following hyperextension or flexion trauma in the context of preexisting cervical stenosis [1, 8, 10, 13, 16]. As previously reported by Ishida and Tominaga, nonsurgical treatment of patients with acute central cervical spinal cord injury with motor and sensory impairment involving only the upper extremities (ACCSCI-U) resulted in nearly full recovery within a period of 6 weeks after the spinal cord injury had been diagnosed [8]. Interestingly, in patients with central cord syndrome requiring surgical intervention, Chen et al. demonstrated an absence of significant association between the surgical timing or approach with the degree of neurological improvement. The authors further concluded that despite marked postoperative neurological improvement, graded according to the ASIA impairment scale motor score, 32.7% of patients were dissatisfied with their clinical outcome [6].

Moreover, neurological deterioration has also been reported to occur in the setting of rheumatic disease due to inflammatory involvement of the atlantoaxial articulation and retrodental pannus formation [4, 12, 17, 19, 20]. Although the cervical spine is a common site of rheumatoid disease manifestation, there is paucity of literature on isolated C1–C2 involvement, especially in the elderly population without evidence of previous rheumatic disease. Additionally, isolated deposition of CPPD at the atlantoaxial joint, as observed in CDS, is a rare entity potentially causing progressive neurological decline [2, 3, 7, 9, 11, 14, 15].

In this case, gradually progressive neurological deterioration resulting in functional quadriplegia was caused by a large retrodental mass consisting of crystalline calcium pyrophosphate and reactive fibrous tissue admixed to deposits of amyloid. In fact, localized deposition of amyloid at the retro-odontoid region, as opposed to systemic disease, has previously been reported by Werner et al. [18] In the present case, systemic amyloidosis was excluded based on the patient's clinical presentation with absence of systemic findings such as lymphadenopathy, unexplained fever, chills, weight loss, cardiac, pulmonary, or hepatic disease. However, due to the fact that the mass was composed of fibrous

Fig. 6. **a, b** Intraoperative images of the surgical site showing a component of the retrodental mass. *Arrows* point at crystalline deposits within the mass. **c** Resected specimen sent for histopathologic examination.

tissue, in addition to both amyloid and CPPD, the diagnosis of crowned dens syndrome rather than primary cervical amyloidoma was made. Nevertheless, most of the compressive effect of the retrodental mass on the cervical spinal cord was caused by the formation of reactive fibrous tissue admixed to deposits of amyloid. We believe that the compressive lesion at the C1–C2 articulation was causing repetitive microtrauma within the spinal cord during cervical motion, resulting in a progressive neurological decline over the time course of weeks to months. In case of a single traumatic event causing incomplete spinal cord injury, one would expect a neurological deficit immediately after the

trauma followed by a gradual recovery [8], rather than progressive worsening over a period of weeks to months, after a symptom-free interval.

To the authors' best of knowledge, this is the first report of progressive neurological deterioration resulting in functional quadriplegia caused by isolated deposition of CPPD at C1–C2 with marked neurological improvement following late surgical decompression and stabilization. We suggest that crystalline arthropathy of the atlantoaxial joint should be considered in the differential diagnoses of isolated C1–C2 retrodental mass causing progressive neurological deterioration due to cervical stenosis and spinal cord compression.

Fig. 7. Histopathologic analysis of the resected retrodental mass illustrating positively birefringent rhomboid crystal deposits with morphologic features of calcium pyrophosphate (**a**; hematoxylin and eosin stain under polarized light) and Congo red positive perivascular deposits of amyloid (**b**) with apple green birefringence under polarized light (**c**).

Disclosures

Conflict of Interest: Alexander Aichmair, MD, Richard J. Herzog, MD, Giorgio Perino, MD, and Darren R. Lebl, MD have declared that they have no conflict of interest.

Human/Animal Rights: All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008 (5).

Informed Consent: Informed consent was waived from all patients for being included in the study.

Required Author Forms Disclosure forms provided by the authors are available with the online version of this article.

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