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Unilateral Biportal Endoscopic Resection of the Posterior Arch of the Atlas for Crowned Dens Syndrome

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Conflict of interest: None declared

Patient: Female, 86-year-old

Final Diagnosis: Crowned dens syndrome

Symptoms: Neck pain exacerbated by rotation • gait instability • "walking on cotton" sensation • thoracoabdominal girdle tightness • impaired fine motor skills

Clinical Procedure: UBE posterior cervical atlantal posterior arch resection and decompression

Specialty: Orthopedics and Traumatology

Objective: Rare disease

Background: Crowned dens syndrome (CDS) is a rare disorder caused by calcium pyrophosphate deposition around the odontoid process. Although most patients respond favorably to conservative management, some develop refractory myelopathy requiring surgical intervention. We present a minimally invasive surgical technique for CDS complicated by spinal cord compression.

Case Report: An 86-year-old woman with CDS exhibited progressive myelopathy, including thoracoabdominal girdle sensation, gait disturbance ("walking on cotton"), and impaired fine motor function in the upper extremities. Her symptoms were refractory to 9 months of conservative treatment. Computed tomography and magnetic resonance imaging demonstrated calcification of the transverse and alar ligaments at the craniovertebral junction with associated compression of the medulla oblongata. The patient underwent unilateral biportal endoscopic resection of the posterior arch of the atlas with spinal canal decompression. Intraoperatively, a midline bony defect was identified. Approximately 8 mm of the posterior arch was resected bilaterally using a high-speed drill; partial excision of the posterior atlanto-occipital membrane and posterior atlantoaxial ligament was performed. Postoperatively, neck pain and girdle sensation were greatly alleviated. At the 1-year follow-up, gait stability and fine motor function had substantially improved, the Japanese Orthopaedic Association score increased from 9 to 12, and no procedure-related complications were observed.

Conclusions: Unilateral biportal endoscopic posterior arch resection of the atlas with decompression represents a feasible and minimally invasive alternative to conventional open surgery for selected patients with CDS displaying myelopathy, particularly high-risk older individuals. This case underscores the importance of surgical intervention in patients with CDS and spinal cord compression.

Keywords: Atlas • Endoscopy • Orthopedics • Surgical Procedures, Operative

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Introduction

Crowned dens syndrome (CDS) is a rare cervical disorder caused by calcium pyrophosphate crystal deposition in the ligaments surrounding the odontoid process [1]. First described in 1985, CDS predominantly affects older individuals. Its typical presentation includes acute neck pain, fever, and elevated inflammatory markers; however, the condition frequently poses diagnostic challenges due to its clinical overlap with meningitis, cervical spondylosis, and other inflammatory disorders [2]. Although most patients respond favorably to conservative management, some may develop refractory symptoms or progressive spinal cord compression.

Management of these refractory cases remains challenging, and surgical intervention is rarely reported in the literature; most available reports describe open decompression or fusion procedures [3]. Here, we report a case of CDS successfully managed with unilateral biportal endoscopic (UBE) posterior cervical decompression. To our knowledge, the application of UBE in CDS has not previously been described. This case is clinically significant because it demonstrates the feasibility of a minimally invasive endoscopic approach for upper cervical decompression in a patient with CDS, providing a potential alternative to conventional open surgery. By highlighting this novel surgical strategy, we seek to expand therapeutic options for refractory CDS and inform clinical practice regarding the management of this rare but clinically challenging condition.

Case Report

An 86-year-old woman presented with a 9-month history of neck pain and unsteady gait. Her symptoms included neck pain

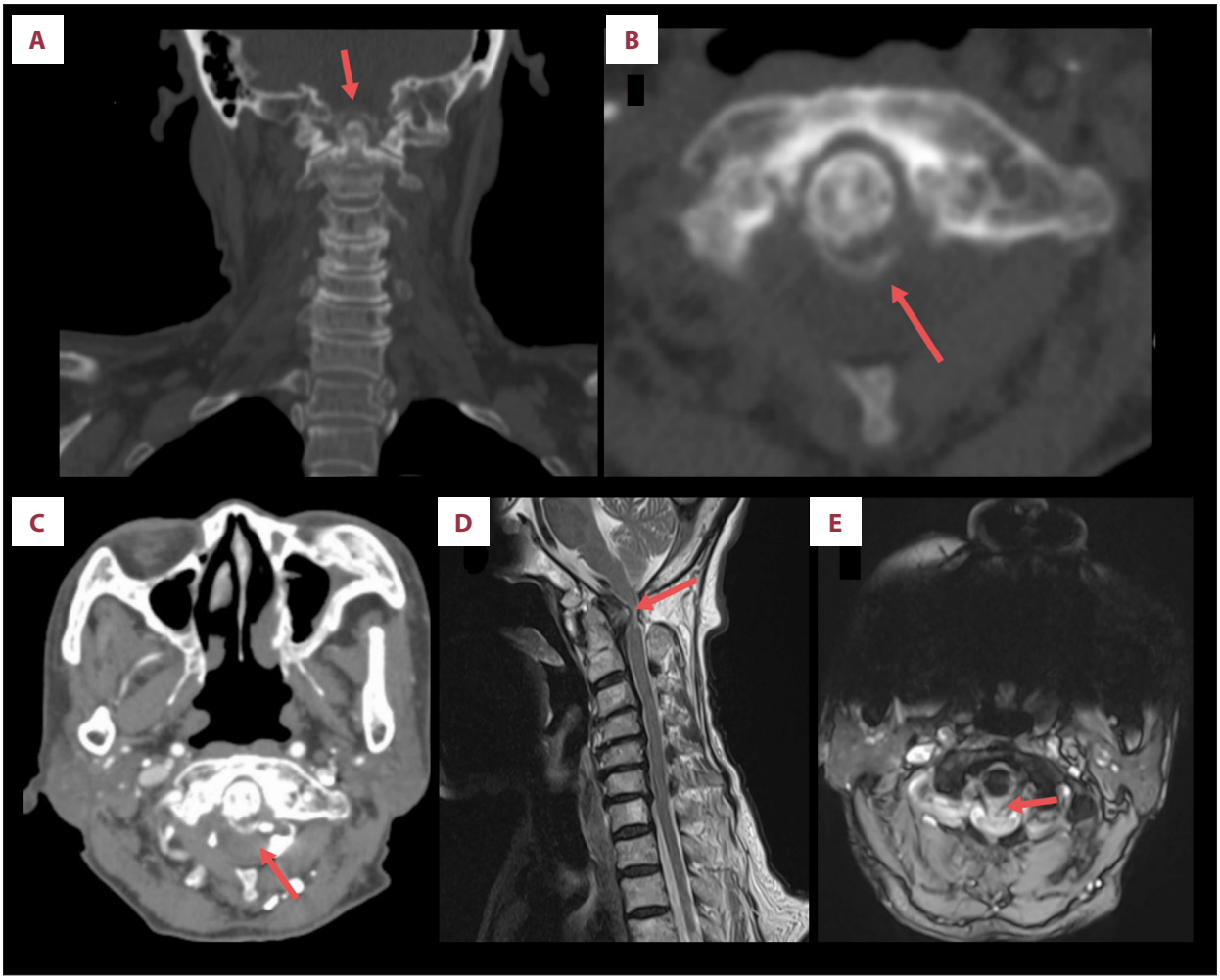

Figure 1. (**A**) Coronal computed tomography (CT) scan showing transverse ligament calcification around the odontoid process. (**B**) Axial CT scan (bone window) confirming calcification. (**C**) Axial CT scan (soft-tissue window) demonstrating the same finding. (**D**) Sagittal magnetic resonance imaging (MRI) revealing transverse and alar ligament calcification compressing the medulla oblongata. (**E**) Axial MRI confirming ligament calcification with medullary compression.

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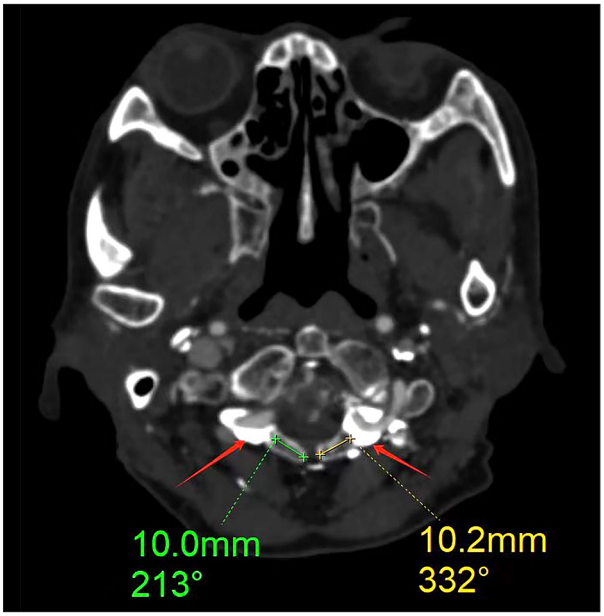

Figure 2. Axial computed tomography scan (2024) showing the surgical plan for posterior arch resection of the atlas.

exacerbated by neck rotation, gait instability, a sensation of "walking on cotton," thoracoabdominal girdle tightness, and impaired fine motor skills. Computed tomography (CT) revealed calcification of the transverse ligament surrounding the odontoid process of the atlas (**Figure 1A-1C**). During the 9 months after symptom onset, the patient had received treatment with analgesics, nonsteroidal anti-inflammatory drugs, corticosteroids, neurotrophic agents, physical therapy (including ultrasound and intermediate-frequency electrical stimulation), and acupuncture; she had experienced poor clinical response to all treatments. Progressive symptoms and signs of cervical spondylotic myelopathy prompted hospital admission.

The patient had a 4-year history of hypertension and type 2 diabetes mellitus, both managed with regular medication. Physical examination revealed a body temperature of 36.7°C—supporting exclusion of the acute phases of meningitis and CDS—limited neck flexion and extension, no clinically significant abnormalities in muscle strength or tone, decreased sensation in the posterior aspect of the left lower limb, negative bilateral Hoffmann and Babinski signs, and no radiating pain or numbness in the upper or lower limbs. The Japanese Orthopaedic Association score was 9. Somatosensory evoked potential testing demonstrated abnormal findings. Laboratory tests revealed a white blood cell count of 6.25×10^9/L, erythrocyte sedimentation rate of 20 mm/hour, and C-reactive protein level of 0.4 mg/L. Serum calcium, magnesium, rheumatoid factor, uric acid, and tumor marker levels were within normal limits.

Magnetic resonance imaging (MRI) and CT scans of the head and neck confirmed calcification of the transverse and alar ligaments at the craniovertebral junction, as well as compression of the medulla oblongata (**Figure 1C-1E**). Based on the patient's history, physical examination findings, and imaging results, the final diagnosis was CDS with high cervical spondylotic myelopathy [4].

Given the progressive worsening of neurological symptoms despite 9 months of conservative treatment, conservative management was considered unsuccessful, and surgical intervention was planned. Considering the patient's advanced age and multiple comorbidities, as well as the substantial surgical trauma associated with conventional open decompression procedures, spinal endoscopic posterior arch resection of the atlas was selected following departmental discussion. After completion of preoperative preparation (**Figure 2**) and exclusion of surgical contraindications, the patient underwent UBE posterior cervical decompression via resection of the posterior arch of the atlas (**Figure 3**) [5,6].

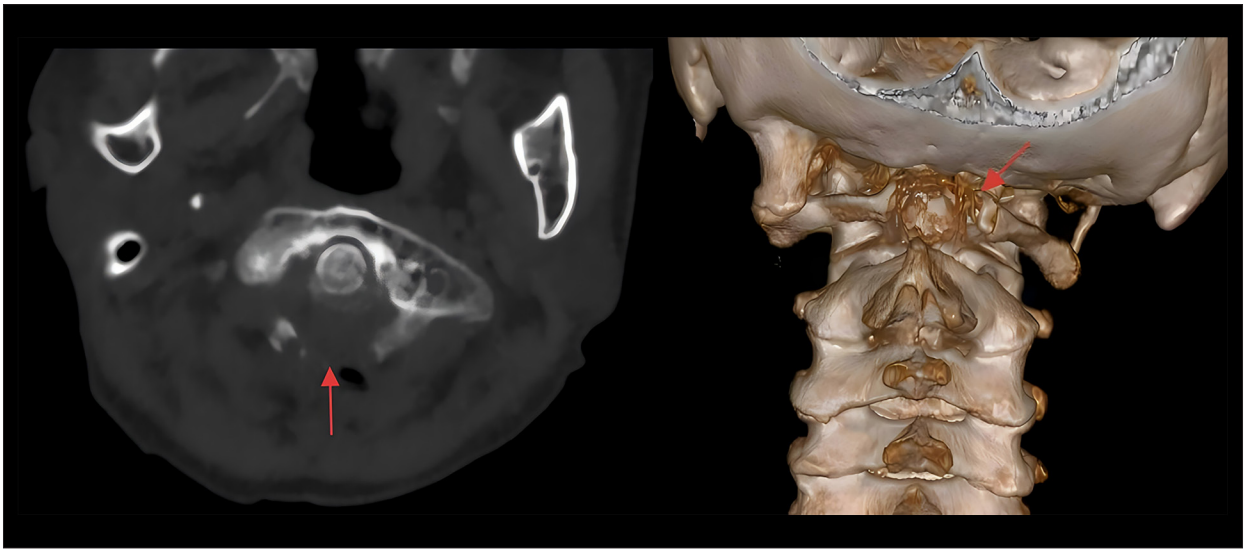

Figure 3. Postoperative computed tomography scan (2024) showing resection of the posterior arch of the atlas.

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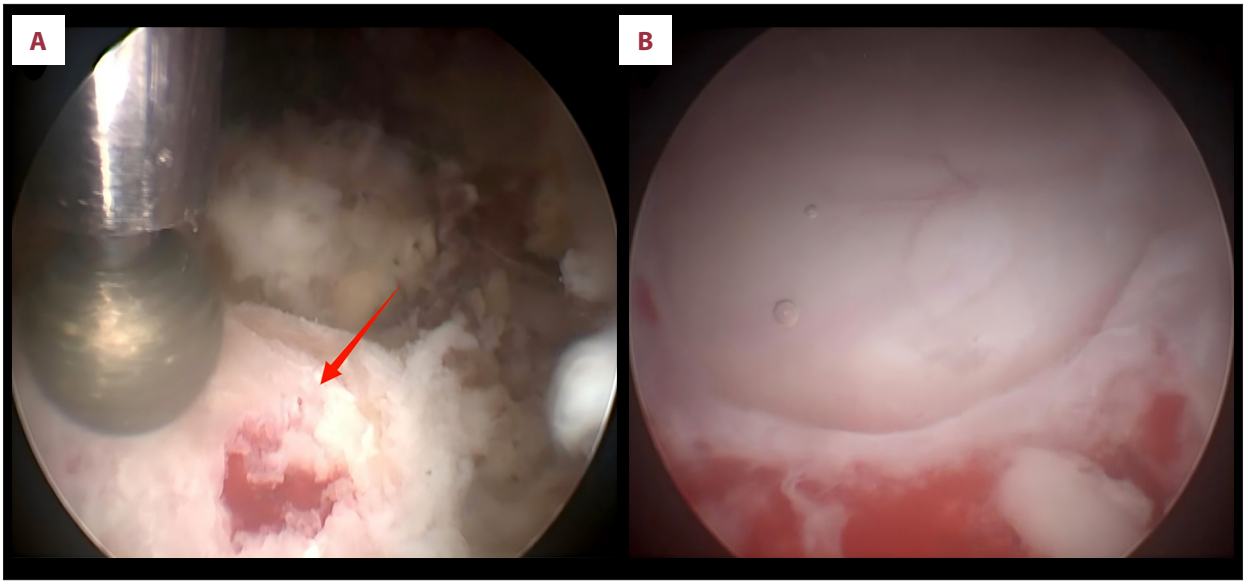

Figure 4. (**A**) Intraoperative endoscopic image showing drilling of the left posterior arch of the atlas. Red arrow: left posterior arch of the atlas. (**B**) Intraoperative endoscopic image demonstrating successful decompression.

Intraoperatively, a midline bony defect measuring approximately 4 mm was identified in the posterior arch of the atlas (**Figure 4**). Beginning at this defect, approximately 8 mm of the posterior arch was resected bilaterally using a high-speed drill (**Figure 4A**). The posterior atlanto-occipital membrane and posterior atlantoaxial ligament were then partially excised. Adequate decompression was confirmed by satisfactory bulging and pulsation of the dural sac (**Figure 4B**).

Postoperatively, the patient's neck pain and thoracoabdominal girdle sensation greatly improved. At the 1-year follow-up, symptoms had not recurred, the sensation of "walking on cotton" had substantially improved, and upper-limb fine motor function had recovered. The Japanese Orthopaedic Association score improved to 12. Follow-up MRI at 1 year demonstrated relief of spinal cord compression and gradual resolution of postoperative edema (**Figure 5**). The surgical outcome was favorable and resulted in a pronounced improvement in the patient's quality of life.

Discussion

CDS is a rare disorder primarily caused by calcium pyrophosphate deposition. Early clinical manifestations typically include acute neck pain, stiffness, and restricted range of motion. Most patients present with fever and elevated inflammatory markers; late-stage manifestations may include signs of spinal cord compression [7,8]. CT is considered the gold standard for diagnosis and typically demonstrates calcifications surrounding the odontoid process. On fat-suppressed T2-weighted MRI, these lesions may manifest as areas of high signal intensity [9,10].

Clinically, due to its symptoms and elevated inflammatory markers, CDS should be distinguished from conditions such as meningitis, epidural abscess, rheumatoid arthritis, giant cell arteritis, cervical spondylosis, and metastatic bone tumors [7,8]. However, some patients may not exhibit obvious systemic or inflammatory features [11]. Thus, CT evaluation of the atlantoaxial joint is crucial for diagnosing CDS because it may prevent unnecessary invasive procedures and inappropriate pharmacologic treatment.

Although most patients respond well to nonsteroidal anti-inflammatory drugs or corticosteroids [3,11], surgical intervention is required for patients with refractory symptoms or progressive neurological deficits [12]. In the present case, failure of conservative treatment was defined by the following criteria: (1) persistence or worsening of myelopathic signs, such as gait disturbance, hand clumsiness, and hyperreflexia, despite 9 months of adequate pharmacologic treatment with nonsteroidal anti-inflammatory drugs, corticosteroids, and neurotrophic agents; (2) radiologic progression of spinal cord compression on follow-up MRI; and (3) lack of improvement with nonoperative interventions, including physical therapy and acupuncture. Because CDS primarily affects older individuals and is associated with increased surgical risk, minimally invasive techniques may offer particular advantages. Our patient was 86 years old and had multiple comorbidities, including hypertension and type 2 diabetes mellitus, making conventional open decompression more likely to result in greater blood loss, prolonged recovery, and increased perioperative morbidity. After multidisciplinary discussion, the UBE approach was selected because it provided a less invasive alternative while achieving adequate decompression.

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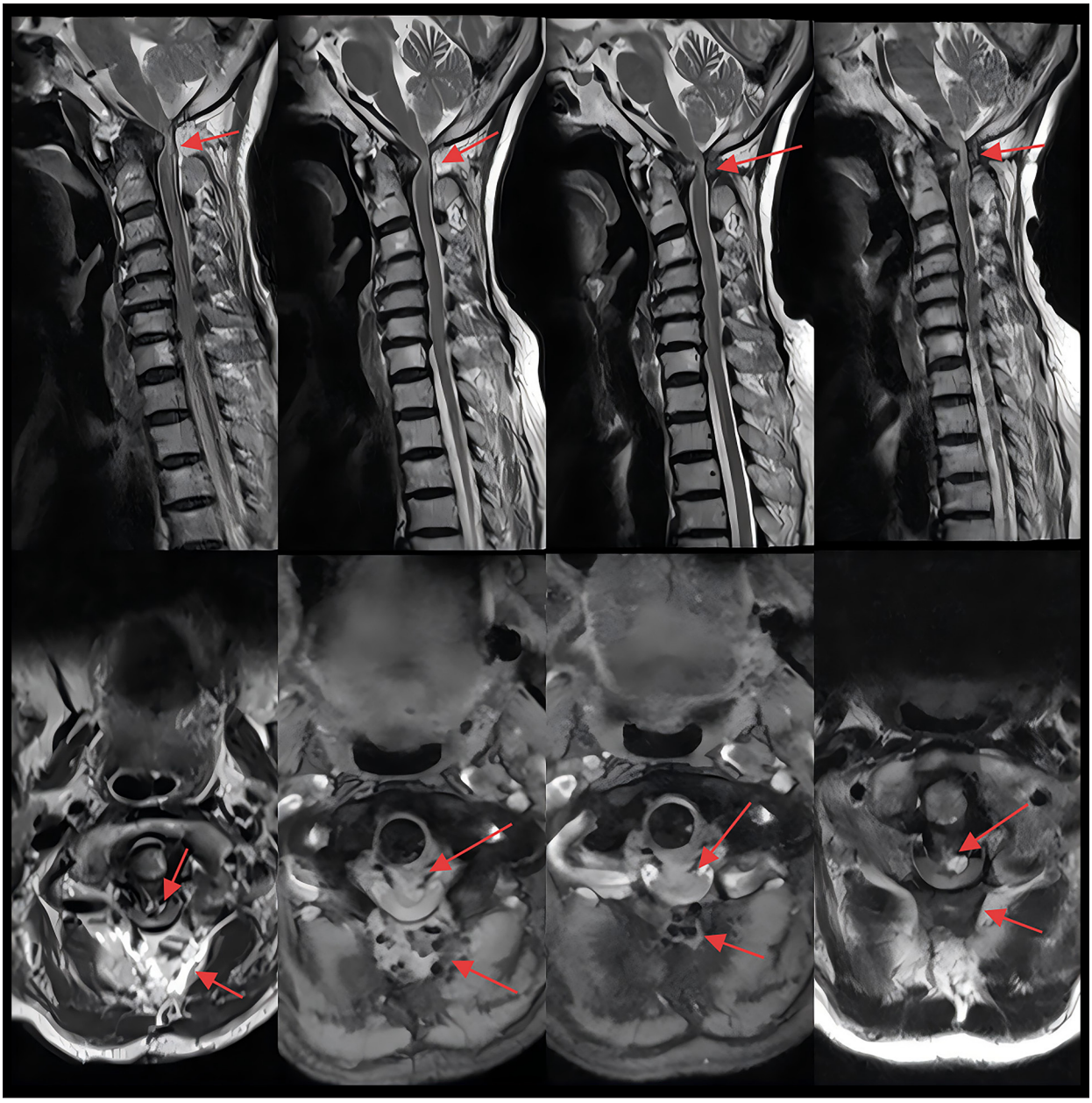

Figure 5. Serial postoperative magnetic resonance examinations (2024-2025) demonstrating relief of spinal cord compression and gradual resolution of postoperative edema.

Various surgical options have been reported for refractory CDS, including posterior open decompression (such as laminectomy or posterior arch resection of the atlas) and posterior fusion with or without instrumentation [13]. Compared with open decompression, the UBE technique used in our case offered several potential advantages: (1) enhanced visualization of the compressed spinal cord through a magnified endoscopic view; (2) reduced paraspinal muscle trauma and blood loss due to smaller incisions; (3) greater preservation of posterior ligamentous and bony structures; and (4) faster postoperative recovery and shorter hospital stay [14,15]. However, UBE also has limitations. The steep learning curve requires advanced endoscopic skills and familiarity with upper cervical anatomy. Moreover, the working space in the atlantoaxial region is inherently narrow, and intraoperative bleeding may compromise visualization more readily than in open surgery. Unlike posterior fusion, UBE does not provide immediate segmental stability and is thus best suited for patients without clinically significant atlantoaxial instability or subluxation. Preoperative X-ray examination of our patient showed only degenerative changes in the cervical spine without indicating cervical instability; thus, UBE decompression was a reasonable treatment

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option. These comparative considerations have not been systematically addressed in the CDS literature. Our report helps address this gap by providing a balanced evaluation of UBE relative to conventional surgical approaches.

The patient in this case report presented with late-stage CDS symptoms, was refractory to initial conservative treatment, and had developed clinical manifestations of cervical spondylotic myelopathy, indicating the need for surgical intervention. Considering the patient's advanced age, the elevated risks associated with open surgery, and previous experience with UBE cervical decompression, posterior endoscopic resection of the posterior arch of the atlas was selected for decompression. This case demonstrates successful application of the UBE technique for atlantoaxial decompression. In addition to reporting a novel surgical approach, this case provides several unique contributions to the understanding and management of CDS. First, it challenges the conventional view that CDS is uniformly a benign, self-limiting condition by documenting a patient with chronic progressive myelopathy requiring surgical intervention. Second, it introduces UBE as a feasible minimally invasive option for upper cervical decompression in CDS, thereby expanding the therapeutic options for refractory cases. Third, it provides preliminary evidence that endoscopic decompression alone, without fusion, may be sufficient in selected patients who lack instability, potentially avoiding the morbidity associated with fusion procedures. Finally, the sustained clinical improvement observed at the 12-month follow-up suggests that the benefits of UBE decompression may be durable. Collectively, these findings advance the field by emphasizing the importance of recognizing surgical indications in CDS, supporting a less invasive treatment strategy, and encouraging further investigation of endoscopic approaches for craniocervical junction pathologies [16,17].

Conclusions

Due to its nonspecific presentation, CDS is frequently misdiagnosed; therefore, early diagnosis is essential. Surgical intervention is indicated in a minority of patients who are refractory to medical treatment and present with myelopathy. In the present case, the patient exhibited progressive myelopathic signs over 9 months despite adequate conservative therapy, and CT/MRI confirmed spinal cord compression. Among the available surgical options—including posterior open decompression, posterior fusion, and minimally invasive endoscopic approaches—we selected UBE posterior cervical decompression. This decision was supported by several considerations: (1) the patient's advanced age (86 years) and multiple comorbidities (hypertension and type 2 diabetes mellitus) increased the risks associated with open decompression, including greater blood loss and delayed recovery; and (2) intraoperative findings demonstrated that UBE provided a clear, magnified view of the compressed spinal cord and enabled precise resection of the posterior arch of the atlas with minimal paraspinal muscle injury. Compared with previously reported open decompression procedures, our patient experienced a shorter hospital stay (12 days) and more rapid return to daily activities. Relative to posterior fusion, UBE preserved segmental motion and avoided fusion-related complications such as adjacent segment degeneration. Thus, our case supports UBE posterior cervical decompression through posterior arch resection of the atlas as a viable and effective minimally invasive surgical option for carefully selected patients with advanced CDS [18,19].

Department and Institution Where Work Was Done

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Patient Consent

The patient provided written informed consent for publication, including acquisition of clinical data and images.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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