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CASE REPORT

Complex cervical spondylotic myelopathy: a report of two cases and literature review

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Abstract
Study Design A report of two cases with complex cervical spondylotic myelopathy (CSM) and review of the literature.
Objective To describe two unique patients with complex CSM due to simultaneous anomalies as anteroposterior compressions of the spinal cord in both upper and lower cervical spine, caused by hypertrophic transverse ligament of atlas (TLA), dysplasia of the posterior arch of atlas, disc herniation, hypertrophic ligamentum flavum and osteophytes.
Methods We present such two cases with clinical, imageological presentations, and describe the surgical procedure, to which both patients responded favorably.
Results The neurological functions of both patients gradually improved according to the JOA scores and VAS scores in preoperative clumsiness and gait disturbance during the mean follow-up period lasted for 18 months. The latest plain radiographs and computed tomography (CT) revealed good fusion without instrumental failure and magnetic resonance imaging (MRI) showed good decompression of C1–7 spinal cord of both patients. Both patients are progressively followed-up.
Conclusion Posterior surgical approach as C1–7 laminectomy with fixations or occipital-cervical fusions may obtain better reconstructions of the cervical spine and good neurological recovery for the patients with complex CSM we present. However, the incidence and ethnic predisposition for the patients with complex CSM are still unclear.

Keywords Complex · Hypertrophy · Transverse ligament of atlas · Dysplasia · Posterior arch of atlas · Cervical spondylotic myelopathy · Laminectomy · Fixation

Introduction

Complex cervical spondylotic myelopathy (CSM) due to simultaneous anomalies involving both upper and lower cervical spine and anteroposterior compression of the spinal cord by hypertrophic transverse ligament of atlas (TLA), dysplasia of the posterior arch of atlas, disc herniation, hypertrophic ligamentum flavum and osteophytes is rarely encountered by spine surgeons and has not been reported earlier. Consequently, we present such two unique cases and describe the clinical, imaging presentations and surgical procedure, to which both patients responded favorably.

Case report

A 74-year-old female (patient A) experienced progressive pain of neck and bilateral upper limbs along with hand clumsiness in May 2012. In August 2012, the symptoms deteriorated and she also noticed gait disturbance and began to use a cane. She underwent conservative therapy with poor outcomes in another hospital in February 2013. Then, she visited our clinic and was admitted for surgical treatment in March 2013.

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The patient had no history of traumatic injury of the head or neck or rheumatoid diseases, no particular diseases in her family were noticed either.

Upon admission, neurological examination showed reduction of superficial sensation of arms and hands, grade 3+ muscle strength of bilateral deltoid, biceps, triceps, wrist and finger extensor and flexor, quadriceps, anterior tibialis, gastrocnemius and extensor hallucis longus, and grade 3 of bilateral iliopsoas. Hyporeflexia of bilateral biceps tendon, triceps tendon and radioperiosteal reflexes as well as hyperreflexia of bilateral patellar tendon were detected. Bilateral Hoffmann and Babinski signs were positive. Preoperative imaging diagnosis indicated loss of intervertebral height on C3–6 levels, instability on C2–3 level, hypertrophic TLA combined with dysplasia of the posterior arch of atlas and intervertebral disc herniation combined with hypertrophic ligamentum flavum at the levels of C3–7, osteophyte on C3–6, which caused severely anteroposterior compression of C1 and C3–7 spinal cord and stenosis of upper and lower cervical canal (Figs. 1, 2, 3, 4).

In March 2013, she underwent C1–7 laminectomy with bilateral C1, C2, C7 pedicle screws and C3–6 lateral mass screws fixation and fusion. Initially, she was placed in the prone position after general anesthesia, and the head was fixed firmly in head frame. A vertical skin incision approximately 12 cm was made along C1–T1 spinous process midline with the neck in slight flexion. Subcutaneous tissue and nuchal ligament were subsequently incised, and paravertebral muscles around the spinous process were bilaterally dissected. The laminaes and facet joints from C1–7 were exposed. The bilateral pedicles on C1, C2 and C7 levels were identified and proper pedicle screws were placed and lateral mass screws were inserted into C3–6 bilateral lateral masses; rods of suitable length were fixed with the screws after viewing the screws were in optimum position by intraoperative fluoroscopy. Thereafter, laminectomy on C1–7 levels was performed with the medial quarter of bilateral C3–7 facet joints were excised without injury to the spinal cord that was exposed (Fig. 5). Facet joint cartilages on C1–7 levels were cut with an osteotome, and the gap between the facet joints was fused. The wound was irrigated and the spinal cord was covered with a biological membrane. A drainage tube was placed inside the incision and stepwise suturing was performed. The incision was finally covered with sterile dressing.

The other 78-year-old female (patient B) experienced progressive neck pain and numbness of bilateral upper limbs in April 2012. The symptoms worsened with hand clumsiness and gait disturbance in October 2012. She also underwent conservative therapy with poor outcomes in another hospital in January 2013. Then she visited our hospital and underwent cervical spine surgery in April 2013.

Inpatient physical examination showed reduction of superficial sensation of both hands and legs, grade 3+ muscle strength of bilateral deltoid, biceps, triceps,

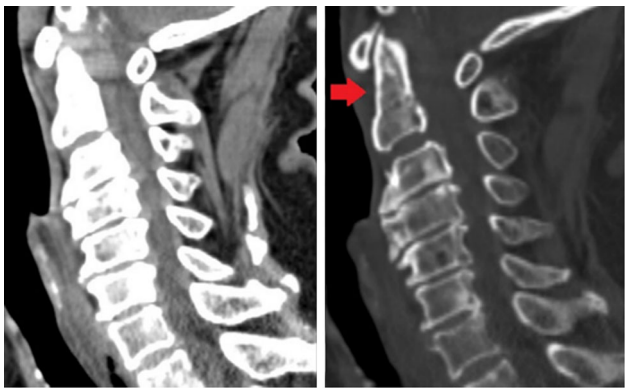

Fig. 2 Preoperative sagittal CT of patient A showed osteophytes on C3–6 and no dens lesion was observed (*arrow*)

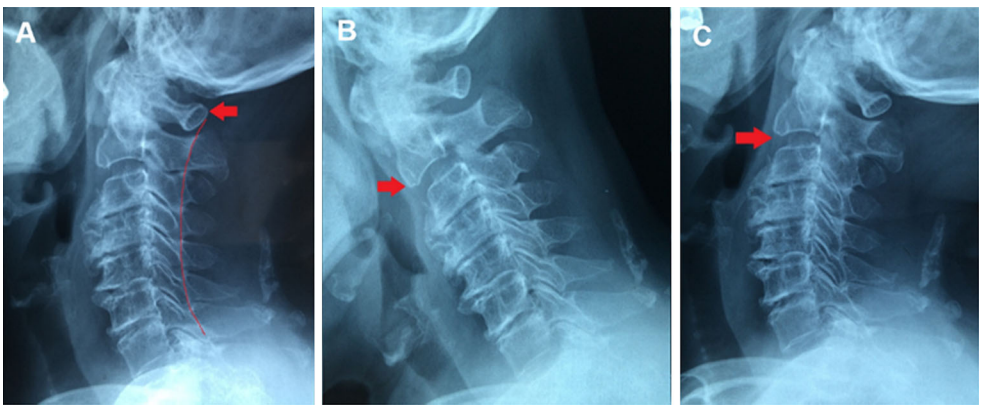

Fig. 1 **a** Preoperative neutral X-ray of patient A showed dysplasia of the posterior arch of atlas (*arrow*) which translated ventrally (*arc*). **b** and **c** Instability on C2–3 level (*arrow*) was observed in dynamic views

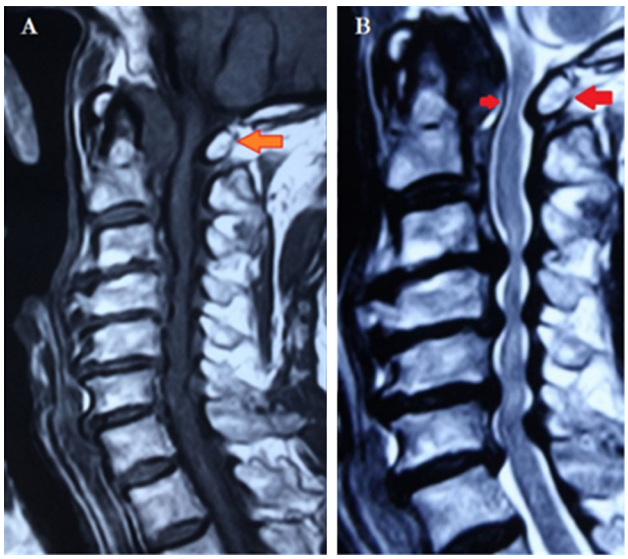

Fig. 3 **a** Preoperative T1-weighted MRI of patient A showed hypertrophic TLA and dysplasia of the posterior arch of atlas (*arrow*). **b** T2-weighted MRI revealed anteroposterior compression of the C1 and C3–7 spinal cord (*arrow*) with intramedullary high signal areas on C1 and C3–6 levels

wrist and finger extensor and flexor, iliopsoas and quadriceps, grade 4− muscle strength of bilateral anterior tibialis, gastrocnemius and extensor hallucis longus. Hyperreflexia of bilateral triceps tendon and patellar tendon as well as hyporeflexia of bilateral Achilles tendon were detected. Bilateral Hoffmann signs were positive. Preoperative imaging examination demonstrated instability on C5–6 level, hypertrophic TLA combined with dysplasia of the posterior arch of atlas and intervertebral disc herniation combined with hypertrophic ligamentum flavum at the levels of C3–7, which also caused anteroposterior compression of C1, C3–7 spinal cord.

In April 2013, she underwent occipito-cervical fusion and C1–7 laminectomy with C2–7 unilateral pedicle screws and lateral mass screws fixation. The initial surgical procedure was similar to that of the first patient. After the occiput and C1–7 laminae with facet joints were exposed, proper pedicle screws were inserted into right side of C2 and C7 pedicles along with lateral mass screws into right side of C3–6 lateral masses, and then two U-shaped screws were inserted into the occiput and a suitable rod was fixed with the screws. Thereafter, laminectomy on C1–7 levels was performed.

Both patients received postoperative rehabilitation therapy gradually. The drainage tubes were taken out 3 days after operation. Soft neck collars were applied for 1 month. The two patients were followed up at 19 and 18 months, respectively. At the latest follow-up, although residual symptoms were noticed and intramedullary alterations were still observed, the neurological functions of both patients gradually improved according to the JOA scores and VAS scores in preoperative hand clumsiness and gait disturbance during the mean follow-up period that lasted for 18 months (Tables 1, 2).

At the final follow-up, grade 4 muscle strength of bilateral deltoid, biceps, triceps, wrist and finger extensor and flexor, quadriceps, anterior tibialis, gastrocnemius and extensor hallucis longus and grade 4− of bilateral iliopsoas were detected on patient A, as well as grade 4 of such muscles was detected on patient B. The latest imaging examination revealed good position of the screws without penetration into vertebral foramen or transverse foramen and the total cervical spinal cord was effectively decompressed (Figs. 6, 7, 8, 9).

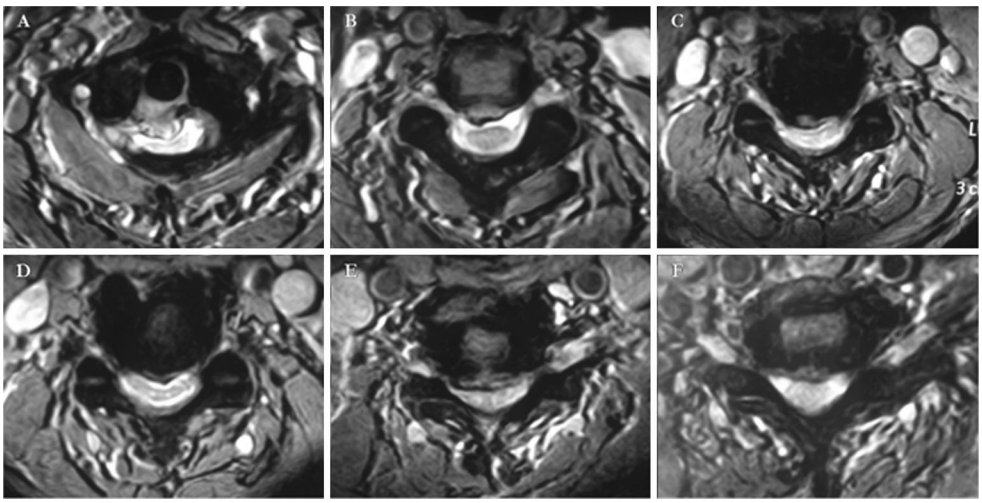

Fig. 4 Preoperative axial MRI of patient A showed compressed spinal cord on C1, C3–7 levels

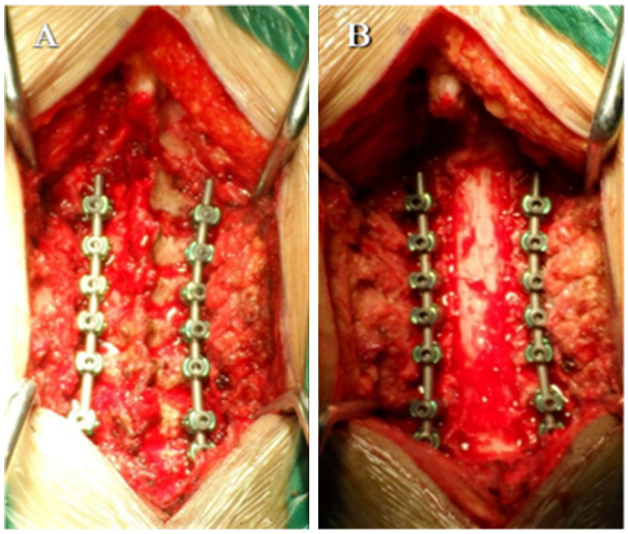

Fig. 5 **a** Fixation instrumentations of patient A were in satisfactory position. **b** Total laminectomy on C1–7 levels was performed and the dural sac was exposed

Table 1 JOA scores

	Patient A	Patient B
Preoperative	5	6
6 months after surgery	7	9
12 months after surgery	10	11
18 months after surgery	12	13

Recovery rate = (post − pre)/(17 − pre) × 100 %

Recovery rate patient A = 58.3 %

Recovery rate patient B = 63.6 %

Table 2 VAS scores

	Patient A		Patient B
	Neck pain	Arm pain	Neck pain
Preoperative	9	9	7
6 months after surgery	5	5	4
12 months after surgery	2	3	2
18 months after surgery	1	2	1

Discussion

The causes of the complex CSM in our cases include hypertrophic TLA, dysplasia of the posterior arch of atlas, disc herniation, hypertrophic ligamentum flavum and osteophytes. All these reasons resulted in anteroposterior compressions of the spinal cord in both upper and lower cervical spine in both patients, and it has been rarely reported yet. Several factors include dysplasia of anterior or posterior arch of atlas, ossification of TLA, hypertrophic

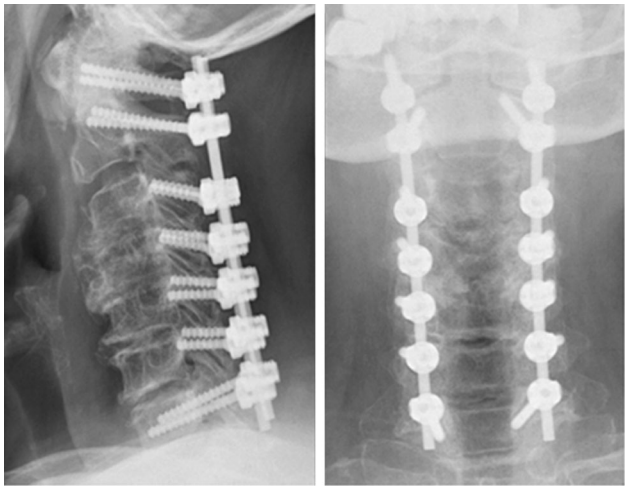

Fig. 6 The latest radiographs in lateral and anteroposterior views of patient A showed good position of the screws 18 months postoperatively

dens and os odontoideum are rare cases causing CSM reported singly or in combination only by sporadic case reports in the medical literature, however, few studies reported the complex CSM due to the reasons we mentioned in our cases [1–9]. Regarding the concomitance of upper and lower with anteroposterior cervical spinal cord compression with multi-factors, the complex CSM was considered consequently.

Stenosis of atlantal canal is rarely encountered due to the particularly protective anatomy against compression [6]. Although the primary anomalies at the level of atlas reported before are defects of either anterior or posterior arch of atlas [1], these infrequently result in compression of atlantal spinal cord, since they usually either increase the canal dimension or leave it changeless with a few exceptions [8]. Compared to these anomalies, hypertrophic TLA decreases the dimension of the atlantal canal. Tatsuro Sasaji, etc., reported a rare case of CSM due to ossification of TLA with a degenerated and hypertrophic dens [5]. Nonetheless, the TLA was not ossified but rather a hugely hypertrophic mass in our cases, which was observed by MRI (Fig. 3) and the dens were intact without retro-odontoid pseudotumor [16, 17] (Fig. 2). However, the mechanism of hypertrophic TLA is still unclear.

Dysplasia of the posterior arch of atlas was considered in our cases due to the ventrally translated arch that was observed according to the sagittal neutral radiograph and CT. Klimo P Jr, etc., reported a 17-year-old patient with cervical myelopathy caused by congenital anomaly of the posterior arch of atlas [18].

Atlantal anomalies also show evidence of ethnic predisposition, which believed to be a disease exclusive to the far East [1, 9], but a recently report was Caucasian, demonstrating that although primarily affecting Asians, it

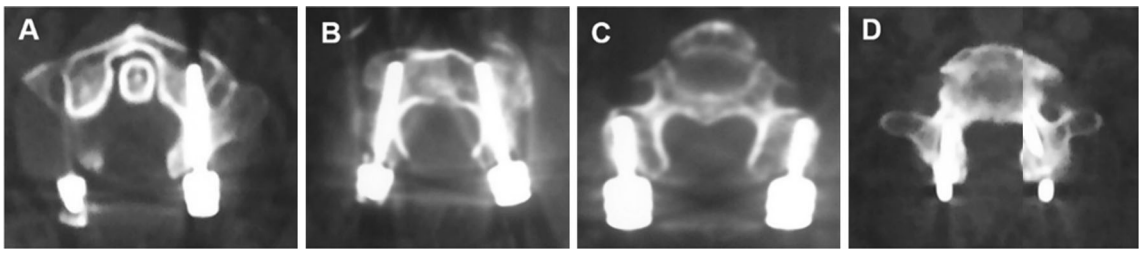

Fig. 7 **a–d** The latest axial CT on C1, C2, C3 and C7 of patient A showed the screws were in correct positions

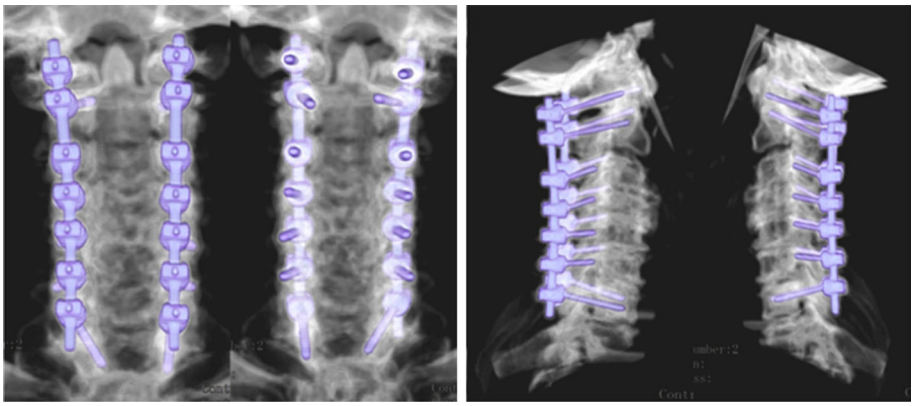

Fig. 8 The latest reconstructed CT of patient A showed good instrumental placements

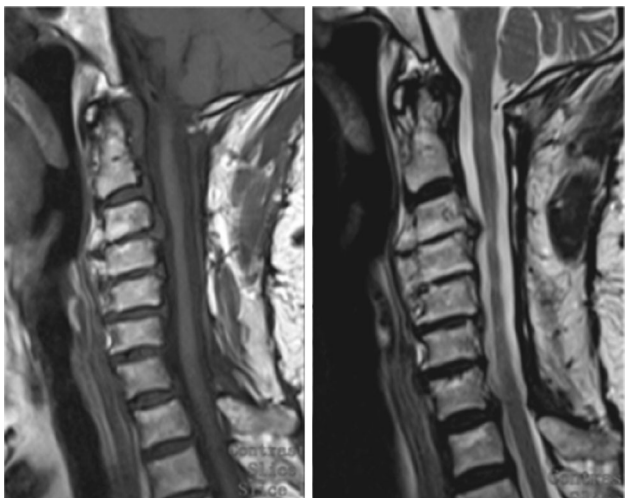

Fig. 9 The latest T1 and T2-weighted MRI of patient A demonstrated effective decompression of C1–7 spinal cord

is not exclusive to a specific race or sex and it is still not clear whether the disease we mentioned is correlated to female Asians [2, 8].

Treatment of multilevel CSM has been a popular topic among most spine surgeons. In our cases, coexistence of anteroposterior compression of C1 and C3–7 spinal cord due to hypertrophic TLA, dysplasia of the posterior arch of atlas, disc herniation, hypertrophic ligamentum flavum and osteophytes along with noticed symptoms were observed. Therefore, posterior laminoplasty or laminectomy with fixation were the indication for these patients [10–15]. However, C1–7 laminectomy with bilateral C1, C2, and C7 pedicle screws and C3–6 lateral mass screws on patient A; occipital-cervical fusion and C1–7 laminectomy with unilateral C2, C7 pedicle screws and C3–6 lateral mass screws on patient B were performed because: (1) laminoplasty was suitable for lower cervical cord lesions [11–15]; (2) the medial quarter of bilateral/unilateral facet joints on C3–7 levels of both patients were removed for thorough decompression during laminectomy, this may result in iatrogenic instability and screw fixation can solve the problem; (3) primary instabilities were observed on both patients; (4) the narrow posterior arch of atlas and hemorrhage of left side of the venous sinus below the arch were detected on the second patient intraoperatively, both of which prevented the insertion of C1 pedicle screws, thus, occipital-cervical fusion and C1–7 laminectomy with unilateral C2–7 fixations were performed on patient B, which can reconstruct the biomechanical stability of cervical spine and present an effective alternative technique in treatment of CSM [19]; (5) pedicle screws were placed on C1, C2, C7 levels, since they can offer the necessary biomechanical strength, and lateral mass screws were placed on C3–6 levels, since they facilitate the manipulation of placing the screws with less complication incidence

as injuries of vertebral artery, nerve root or spinal cord [10–14].

Both patients are progressively followed-up.

Conclusion

Complex CSM due to simultaneous lesions involving both upper and lower cervical spine and anteroposterior compression of the spinal cord by hypertrophic TLA, dysplasia of the posterior arch of atlas, disc herniation, hypertrophic ligamentum flavum and osteophytes is a rare event. The incidence and ethnic predisposition are still unclear so far. Posterior surgical approach as C1–7 laminectomy with fixations or occipital-cervical fusions may obtain better reconstructions of the cervical spine and good neurological recovery for the patients.

Conflict of interest None of the authors has any potential conflict of interest.

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