Clinical Case Report

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Ossification of the posterior atlantoaxial membrane associated with atlas hypoplasia

A case report

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Abstract

Rationale: Hypoplasia with an intact posterior arch of the atlas and ossification of the posterior atlantoaxial membrane (PAAM) are individually rare.

Patient concerns: The patient presented with a 6-month history of progressive weakness and paresthesia of his lower extremities.

Diagnoses: Cervical myelopathy resulting from atlas hypoplasia and ossification of the posterior atlantoaxial membrane.

Interventions: Laminectomy of the atlas with duroplasty.

Outcomes: Preoperative symptoms were alleviated.

Lessons: In most reported cases, either atlas hypoplasia or ossification of the PAAM is responsible for patients' myelopathy. The case illustrated here, to the best of our knowledge, is the first one with coexistent atlas hypoplasia and ossification of the PAAM. And laminectomy of the atlas with duroplasty provided satisfied outcome.

Abbreviations: CT = computed tomography, MR = magnetic resonance, PAAM = posterior atlantoaxial membrane, SAC = space available for the spinal cord.

Keywords: atlas hypoplasia, case report, ossification, posterior atlantoaxial membrane

1. Introduction

Cervical spinal stenosis is a well described pathomechanism that causes cervical myelopathy. It generally occurs below the level of C3 and rarely occurs in the upper cervical region. In literature, 16 cases of upper cervical myelopathy resulting from atlas hypoplasia and 6 cases resulting from ossification of the posterior atlantoaxial membrane (PAAM) have been reported.[1–21] There is no study showing association between atlas hypoplasia and ossification of the PAAM. Here, we present a case involving both hypoplasia with a complete posterior arch of the atlas and ossification of the PAAM which cause upper cervical myelopathy.

2. Case report

A 39-year-old male presented with a 6-month history of progressive weakness and paresthesia of his lower extremities. He also described progressive difficulty using chopsticks. In addition, he had intermittent urinary incontinence and the frequency increased in the last 2 months. On admission, he was unable to walk with assistance. He had no respiratory dysfunction. Physical examination demonstrated markedly decreased muscle power (2/5) in the lower extremities, mildly decreased muscle power (4/5) in the upper extremities, hyperactive deep tendon reflexes in all extremities with positive Babinski and Hoffmann signs bilaterally. No significant past medical history or preceding trauma was noted. Routine laboratory tests were normal.

Lateral cervical X-rays revealed narrowing of the spinal canal at the atlas level, with space available for the spinal cord (SAC) of 10 mm (Fig. 1A). And SAC decreased to 6 mm with flexion of the neck (Fig. 1B). Open-mouth view showed mild instability of atlantoaxial joint (Fig. 1C). Cervical computed tomography (CT) scan revealed severe cervical canal stenosis, ossification of the PAAM and a hypoplastic atlas with inner sagittal diameter of 23 mm (Fig. 2). Magnetic resonance (MR) scan revealed severe spinal cord compression with a dural sac sagittal diameter of 5.7 mm in the atlantoaxial region (Fig. 3). The T2-weighted imaging confirmed constriction of the dural sac and an intramedullary high intensity area. Therefore, atlas hypoplasia and ossification of the PAAM were thought to cause myelopathy together.

He underwent decompressive surgery of the atlas and axis via a posterior approach (Fig. 4A). Intraoperatively, ossified PAAM was found between the inferior border of the posterior arch of C1 and superior border of the lamina of C2. No adhesion was noted between the PAAM and the dura mater. The lower half part of

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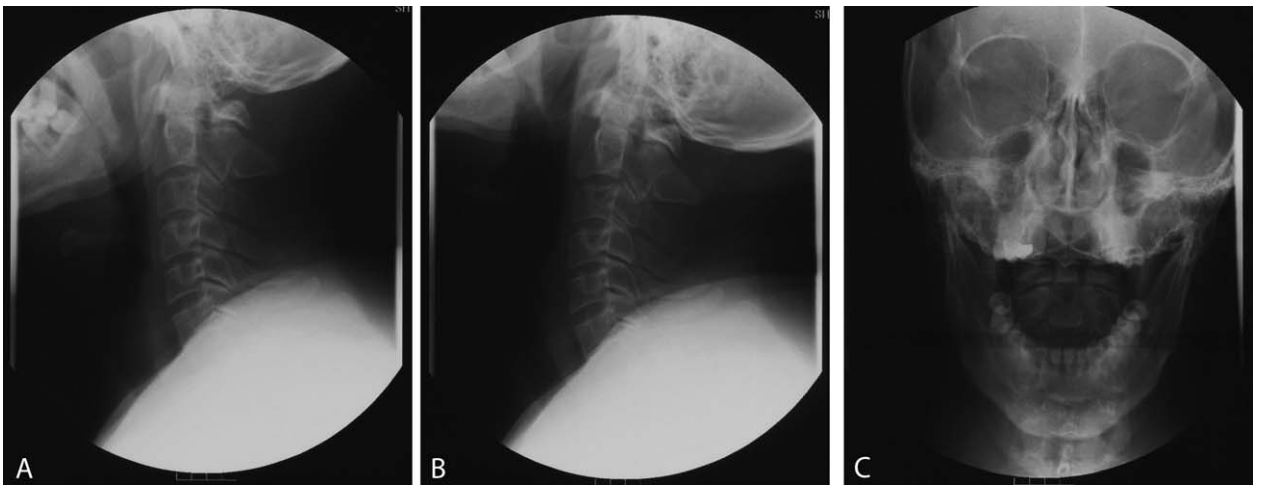

Figure 1. Plain radiograph of the cervical spine. (A) The space available for the spinal cord is10 mm in neutral view. (B) SAC decreases to 6 mm in flexion view. (C) There is mild instability of the atlantoaxial joint in open mouth view.

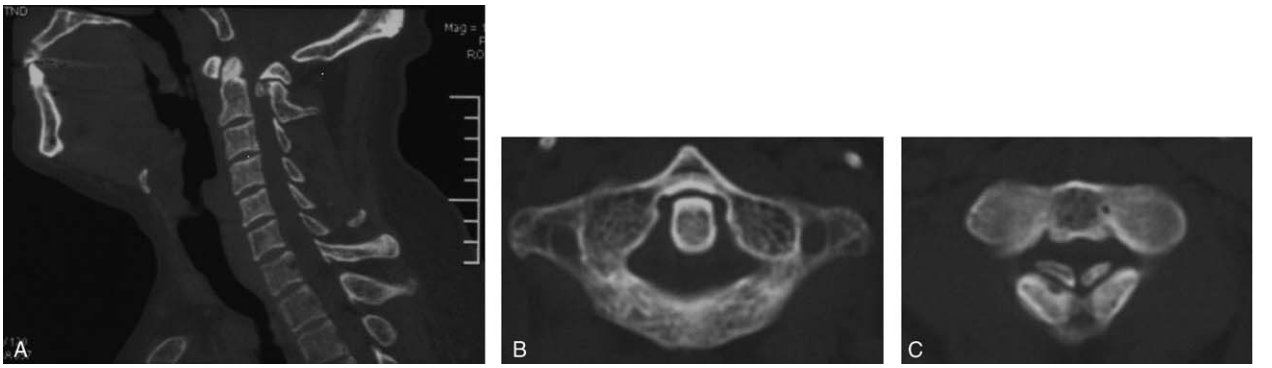

Figure 2. CT of the cervical spine. (A) Sagittal CT demonstrates osseous mass exists between the inferior border of the posterior arch of C1 and superior border of the lamina of C2. (B) Axial CT at the level of atlas shows a small inner sagittal diameter of 23 mm. (C) Axial CT at the atlantoaxial level shows ossified PAAM causing cervical canal stenosis.

posterior arch of C1, upper half part of lamina of C2, and ossified PAAM were resected meticulously using a high-speed drill and bone rongeurs. After decompression, the dural sac expanded to its normal size and good pulsation was confirmed. C1–C2 fusion was performed with autologous iliac crest bone graft. Postoperative CT scan demonstrated complete decompression at the atlantoaxial level (Fig. 4B), whereas MR T2-weighted imaging showed a narrowed cord with a diameter of 2.3 mm and

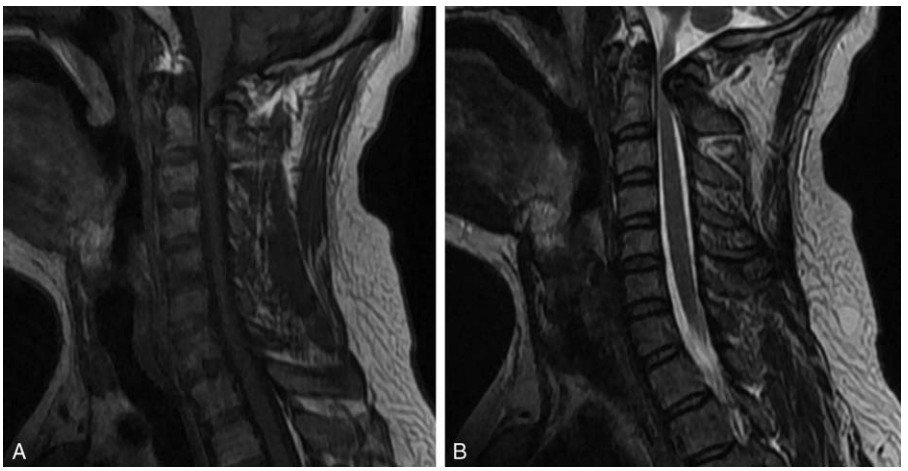

Figure 3. MRI of the cervical spine. (A) T1-weighted MRI shows ossified PAAM compressing the spinal cord. (B) T2-weighted MRI confirms constriction of the dural sac and an intramedullary high intensity area.

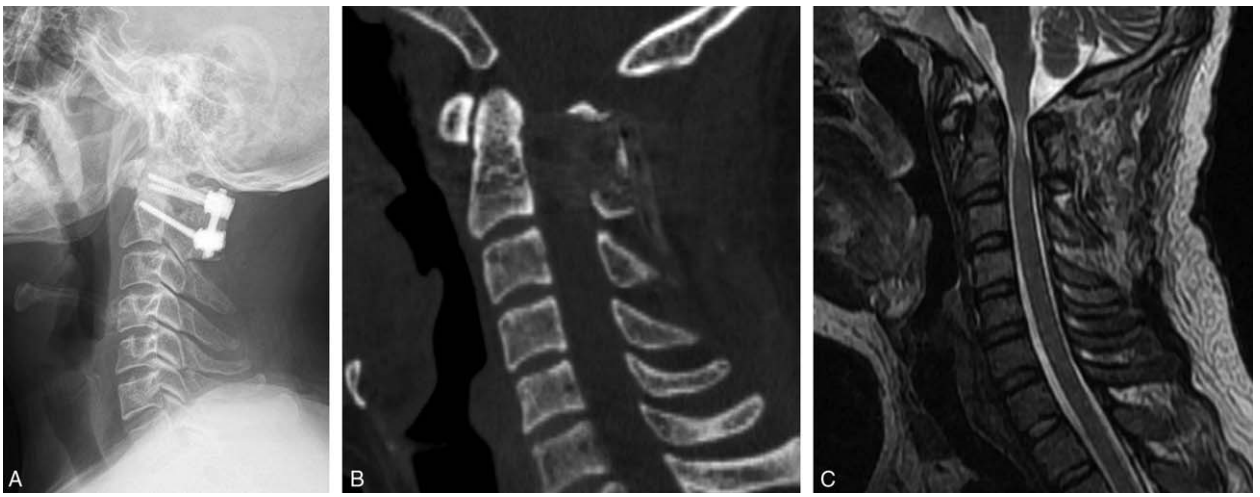

Figure 4. Postoperative images of the cervical spine. (A) Plain radiograph of the cervical spine. (B) CT of the spine reveals enough decompression of the atlantoaxial region. (C) T2-weighted MRI shows a narrowed cord and persisting intramedullary high intensity change at the previous region.

persisting intramedullary high intensity change at the previous region (Fig. 4C).

Postoperative course was uneventful. The patient received 3-month systematic physiotherapy. He could walk 1000 feet with a cane at 6-month follow-up, but still complained of intermittent urinary incontinence.

3. Discussion

We experienced an extremely rare case of the patient who had myelopathy resulting from both atlas hypoplasia and ossification of the PAAM.

The majority of anomalies at the level of atlas reported in the literature are various clefts or aplasias of anterior and posterior arches of atlas and os odontoideum, both of which can result in myelopathy.[12] Atlas hypoplasia with an intact posterior arch is quite rare compared to these anomalies. Symptomatic atlas hypoplasia was first reported in 1974 and 16 further cases have been reported (Table 1).[22] Atlas hypoplasia is defined by Kelly et al who conduct a cadaveric study by measuring sagittal and coronal data of 543 cervical spine specimens. They conclude that atlas with an inner sagittal diameter of 26 mm or less can be described as hypoplastic.[23] Besides, it generally is agreed that a

Table 1

Patients with hypoplasia of the atlas.

Study	Sex/age, y	Type	Ethnicity	C1/2 instability	Canal diameter at atlas level	Dural sac diameter at atlas level	Coexisting ossification	Treatment	Outcome
Sawada et al[2]	Male/38	Hypoplasia of the posterior arch	Asian	No	7 mm	—	No	Laminectomy of the atlas	Improved
Tokiyoshi et al[1]	Male/55	Hypoplasia of the posterior arch	Asian	No	8 mm	—	No	Decompressive suboccipital craniectomy; removal of the posterior arch of the atlas with duroplasty	Improved
Okamoto et al[4]	Male/77	Hypoplasia of the posterior arch	Asian	No	11 mm	—	No	Laminectomy of the atlas and occipitocervical sublaminar wiring	Improved
Phan et al[5]	Male/80	Hypoplasia of the posterior arch	Asian	No	8 mm	—	No	Suboccipital decompression of foramen magnum; laminectomy of the atlas; C2 partial laminectomy	Improved
	Male/75	Hypoplasia of the posterior arch	Asian	No	7 mm	—	No	Laminectomy of the atlas	Improved
Connor et al[7]	Female/8	Hypoplasia of the posterior arch	Caucasian	No	10 mm	—	No	Laminectomy of the atlas	Improved
Urasaki et al[8]	Male/14	Atlas with asymmetrical posterior arch	Asian	No	10 mm	—	No	Laminectomy of the atlas	Improved
Nishikawa et al[6]	Male/82	Hypoplasia of the posterior arch	Asian	No	12 mm	7 mm	No	Laminectomy of the atlas	Improved
	Male/72	Hypoplasia of the posterior arch	Asian	No	11 mm	6 mm	No	Laminectomy of the atlas	Improved
	Female/22	Hypoplasia of the posterior arch	Asian	No	9 mm	—	No	Laminectomy of the atlas	Improved
Tsuruta et al[14]	Female/79	Markedly anterior position of the posterior arch	Asian	No	—	—	Transverse ligament	Laminectomy of the atlas	Improved
Tubbs et al[9]	Female/9	Idiopathic growth hormone deficiency and Klippel–Feil syndrome	Caucasian	No	<10 mm	—	No	Removal of the posterior arch of the atlas and suboccipital craniectomy	Improved
Hsu et al[10]	Male/38	Incurving of the posterior arch	Asian	No	12.46 mm	6.23 mm	No	Removal of the posterior arch of the atlas with duroplasty	Improved
Sabuncuoglu et al[15]	Male/26	Partial absence of the posterior arch	Asian	No	—	—	No	Conservative therapy and a cervical collar	Improved
Tang et al[12]	Female/52	Hypoplasia of the posterior arch	Asian	No	—	5.5 mm	Transverse ligament	Laminectomy of the atlas	Improved
Bokhari and Baeesa[13]	Female/68	Hypoplasia of the posterior arch	Asian	No	—	—	Transverse ligament	Laminectomy of the atlas	Improved

Table 2

Patients with ossification of the posterior atlantoaxial membrane.

Study	Sex/age, y	Type	Ethnicity	C1/2 instability	Canal diameter at atlas level	Dural sac diameter at atlas level	Coexisting ossification	Treatment	Outcome
Yamaguchi et al[21]	Male/46	Bilateral	Asian	—	—	—	No	Laminectomy of the atlas	Improved
Kimura et al[19]	Female/55	Unilateral	Asian	—	—	—	No	Laminectomy of the atlas	Improved
Harimaya et al[18]	Male/52	Bilateral	Asian	No	—	—	Anterior longitudinal ligament	Laminectomy of the atlas	Improved
Nadkarni et al[20]	Male/30	Bilateral	Asian	No	—	—	No	Laminectomy of the atlas	Improved
Shoda et al[17]	Male/70	Unilateral	Asian	No	—	—	Transverse ligament	Laminectomy of the atlas	Improved
Ohya et al[16]	Female/46	Tongue	Asian	Yes	—	—	No	Laminectomy of the atlas, atlantoaxial stabilization	Improved
Present case	Male/39	Bilateral and hypoplasia of the posterior arch	Asian	Yes	10 mm	5.7 mm	No	Laminectomy of the atlas with duroplasty	Improved

reduction of the SAC to 13 mm or less may be associated with neurologic problems.[24] The patient in our case has an atlas sagittal diameter of 23 mm, indicating a hypoplastic atlas.

Ossification of the cervical ligamentum flavum causing myelopathy was first reported by Koizumi in 1962.[25] PAAM has a different character to the ligamenta flava and ossification of the PAAM is a quite rare cause of spinal cord compression.[26] Only 6 cases have been reported so far (Table 2). The exact pathomechanism for ossification of the PAAM still remains unclear. Chronic mechanical stress which may induce osteogenic differentiation of the ligament cells have been suggested to be an etiological factor of ossification of the ligamentum flavum.[27,28] As hypoplastic atlas presented in our patient, extension and flexion of the neck may exert extra stress on the PAAM between the arch of the atlas and the lamina of the axis and thus promote the osteogenic process.

Myelopathy due to combination of hypoplasia with an intact posterior arch of the atlas and ossification of the PAAM has not been reported previously. We suggest that in our case, compression of the spinal cord was mainly caused by ossification of the PAAM secondary to atlas hypoplasia. Resection of the posterior arch of the atlas and lamina of C2 achieved satisfactory decompressive results. C1–C2 pedicle screw fixation was also employed for the complication of mild atlantoaxial instability in this case.[29]

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