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Case Report

Concurrent Ossification of the Posterior Atlantoaxial Interlaminar Membrane and Atlas Hypoplasia: A Case Report

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An onset of cervical myelopathy due to ossification of the posterior atlantoaxial membrane (PAAM) is extremely rare in older patients, and its clinical characteristics are still unclear. We report an onset of ossification of PAAM with congenital atlas hypoplasia in an 81-year-old man who presented with a 2-year history of progressive cervical myelopathy. Cervical computed tomography (CT) revealed canal stenosis secondary to a hypoplastic posterior arch of the atlas with a diameter of 20.3 mm between the anterior and posterior process. Magnetic resonance imaging showed marked spinal cord compression at the level of C1–2 secondary to atlas hypoplasia as well as ossification of PAAM. The patient underwent laminectomy of C1 and partial C2, as well as removal of the ossification, without atlantoaxial fusion. His neurological status improved 1 year postoperatively. In older patients, cervical myelopathy secondary to PAAM ossification, in the absence of trauma and atlantoaxial instability, may be induced by age-related pathophysiology associated with congenital atlas hypoplasia.

Keywords: atlas hypoplasia, cervical spinal cord, myelopathy, ossification of the posterior atlantoaxial membrane

Introduction

Upper cervical myelopathy is a rare pathology. To date, myelopathy secondary to ossification of posterior atlantoaxial membrane (PAAM) has been reported in only seven patients.[1–7] Additionally, atlas hypoplasia is a rare congenital anomaly. A congenitally narrow cervical canal, with restricted available space, is considered a risk factor for developing upper cervical myelopathy; however, the pathophysiology of the related ossification remains uncertain. We report a patient with upper cervical myelopathy secondary to concurrent ossification of PAAM, and a hypoplastic posterior arch of the atlas. We discuss that the pathophysiology of PAAM ossification with atlas hypoplasia could lead to symptomatic onset in older patients.

Case Report

An 81-year-old man presented with a 2-year history of progressive numbness and weakness of his upper and lower extremities and gait disturbance. Spastic quadriparesis and slow gait had progressed in the previous month, and on admission, he had worsened to the point of needing a wheelchair. Physical examination revealed decreased sensation in his upper extremities, motor weakness in both his upper extremities (grade 4/5) and lower extremities (3/5), increased deep tendon reflexes in his lower limbs, and finger dexterity movement disorder. No bladder or bowel dysfunction was observed.

Lateral cervical spine radiography revealed a narrow canal diameter of 9.0 mm at the level of the atlas without instability, and degenerative cervical spondylosis below the level of C2 (Fig. 1). Computed tomography (CT) showed a narrow sagittal canal diameter of 20.3 mm between the anterior and posterior processes, and ossification of PAAM and marked secondary spinal cord compression at the level of C1–2 (Fig. 2). Magnetic resonance imaging showed that the ossified lesion caused severe spinal cord compression in the atlantoaxial area with intramedullary hyperintense signals on T2-weighted imaging. The diameter of the sagittal dural sac was 5.2 mm at the level of atlas.

The patient underwent decompressive resection of the posterior arch of the atlas involving total resection of an ossified lesion and C2 partial laminectomy via a posterior approach (Fig. 3). There was no adhesion between the dura mater and the ossified lesion, intraoperatively. Immediately after the resection, the dural sac just below the ossification expanded, and satisfactory pulsation was observed. We did not perform atlantoaxial fusion. No decompression at middle and lower cervical level was performed because of no significant cord compression.

Pathological findings revealed the proliferation of chondrocytes and collagenous tissue with elastic fibers, indicating ossification within PAAM. The patient's motor weakness recovered gradually, postoperatively. Two weeks after surgery, he began gait training with some assistance and was transferred to a rehabilitation facility. At the 1-year postoperative follow-up, he had improved to crutch walking, and no atlantoaxial instability was observed.

Discussion

We described a rare patient with concurrent ossification of PAAM and atlas hypoplasia causing upper cervical myelopathy. An onset of cervical myelopathy secondary to ossification of PAAM is extremely rare in older patients, and the pathophysiology of the ossification remains unclear. PAAM bridges a space between the posterior arch of the atlas and the cranial edge of the axis. The roles of PAAM are equivalent to those of the ligamentum flavum, maintaining stability and mobility of the spine.[8]

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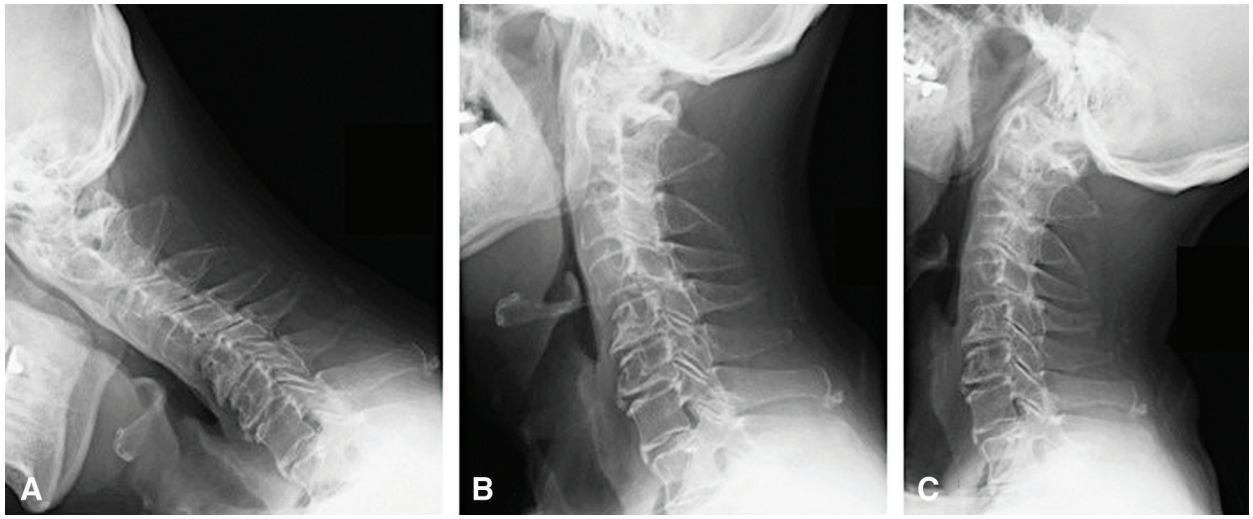

Fig. 1 Cervical spine radiography. (**B**) Lateral view showing a narrowed canal diameter of 9.0 mm at the level of the atlas as well as mild degenerative changes below C4. (**A**, **C**) Flexion–extension lateral radiographs showing no atlantoaxial instability.

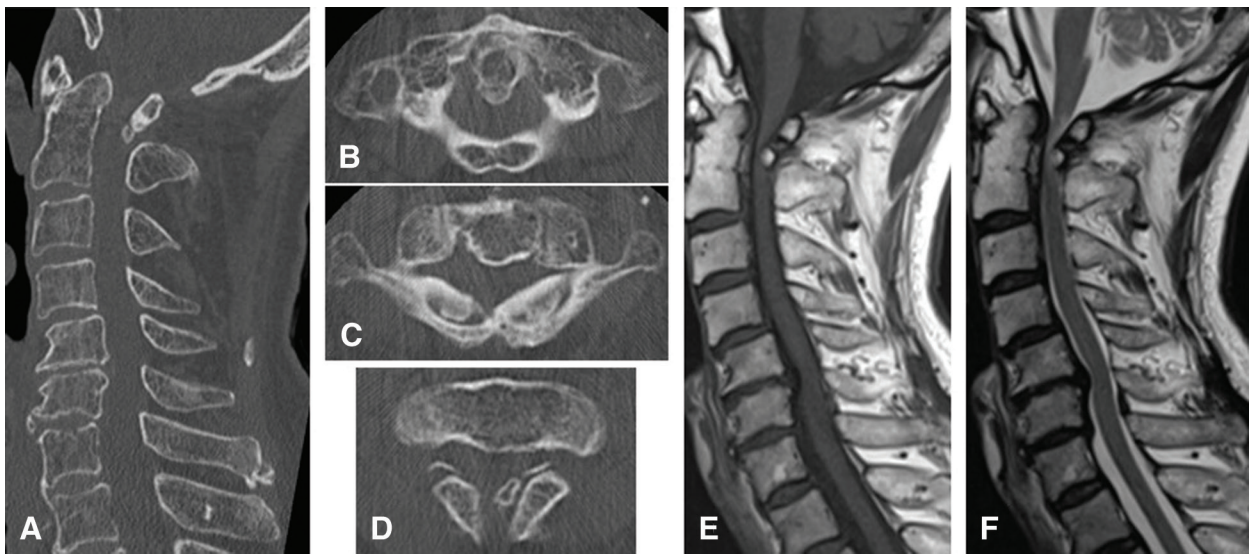

Fig. 2 Preoperative cervical spinal CT at the level of C1–2. (**A**) Lateral view showing atlantoaxial canal stenosis and mild degenerative changes below C4. (**B**) Spinal canal narrowing at the level of the atlas is seen. The narrowest width of the canal was 20.3 mm (inner anteroposterior diameter of the atlas). (**C**) The hypoplastic posterior arch curved inward and was adhered on the midline, with ridge formation inwards. The inner part of the posterior arch was complicated with centripetal hypertrophic ossification. (**D**) Axial CT at the atlantoaxial level showing unilateral ossification of PAAM. MRI of the cervical spine. (**E**) T1-weighted MRI showing marked cord compression with an ossified PAAM at the atlantoaxial level. (**F**) T2-weighted MRI showing severe atlantoaxial canal stenosis (dural sac diameter: 5.2 mm) with intramedullary high signal intensity change. CT: computed tomography, MRI: magnetic resonance imaging, PAAM: posterior atlantoaxial membrane.

We reviewed the literature in PubMed to identify previous reports of PAAM ossification, which identified eight reports, including our patient[1–7] (Table 1). All reported cases occurred in patients from Asia, and all presented with progressive myelopathy. According to these findings, the ossification of PAAM may be similar to that of the ligamentum flavum, which mostly occurs with gradual worsening in Asian. Two patients experienced onset at > 70 years of age (Shoda et al.[5] and our patient). These two patients exhibited slowly progressive symptoms for 2–6 years. The remaining six patients were younger, and the interval between symptom onset and diagnosis was shorter (1–6 months). Symptoms in the six younger patients were related to atlantoaxial instability (including os odontoideum) or post-traumatic conditions. Given these findings, the pathophysiology may differ between patients with older vs. younger onset of symptoms. Dynamic, chronic, and excessive mechanical stress, which may induce osteogenic differentiation of the ligament cells, is a suggested etiological factor in ossification of the ligamentum flavum, including ossification of PAAM. In younger

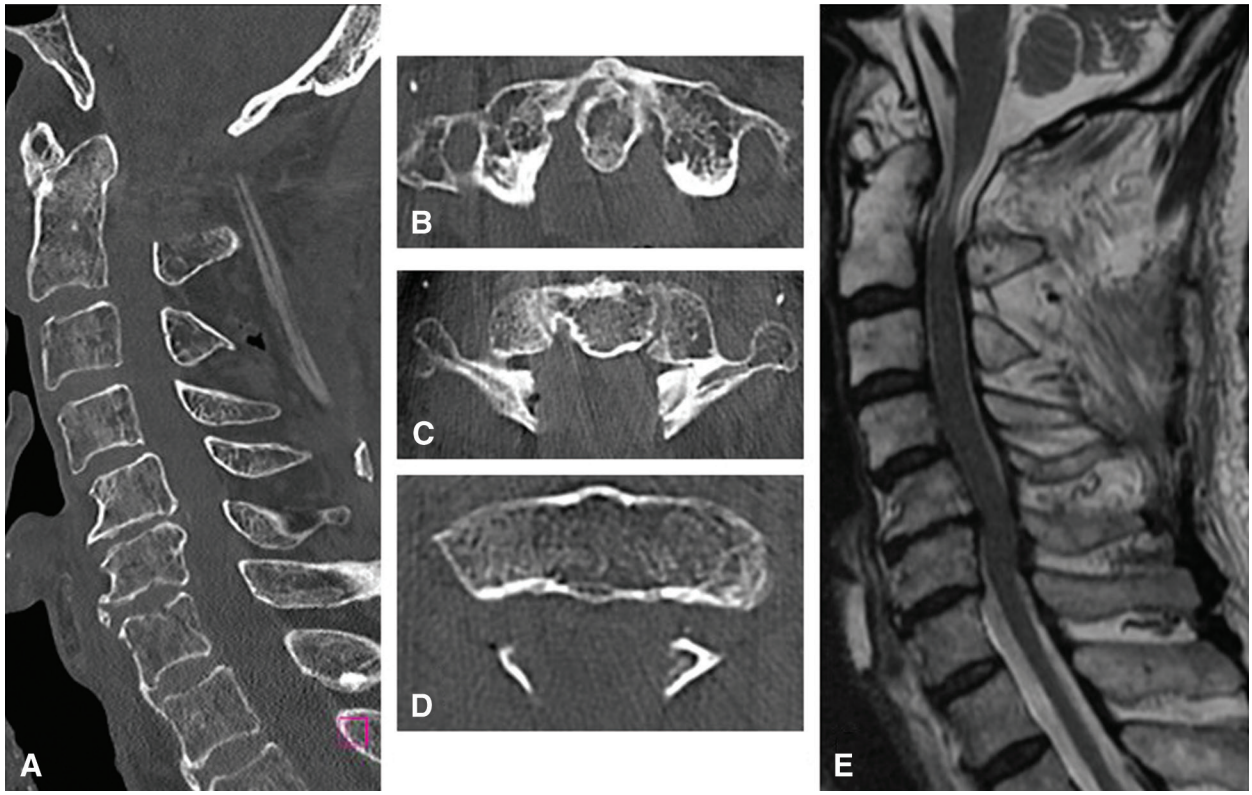

Fig. 3 Postoperative CT and MRI images. (**A–D**) Operative resection of the posterior arch of the atlas, involving total resection of an ossified lesion and C2 partial laminectomy. The postoperative CT image shows the decompressed spinal cord. (**E**) T2-weighted MRI showing upper cervical cord atrophy and persistent intramedullary high signal intensity change. CT: computed tomography, MRI: magnetic resonance imaging.

group, atlantoaxial instability,[7] os odontoideum,[6] and post-traumatic changes[4] could support this hypothesis, according to apparent mechanical stress. Systemic hyperostosis, endocrine abnormalities, and rheumatoid arthritis may also be responsible.

Concurrent ossification of PAAM with atlas hypoplasia has been reported in only one patient, previously,[7] and older onset of cervical myelopathy, as in our patient, has not been reported; atlas hypoplasia is also rare. Kelly et al.[9] first reported the concept of atlas hypoplasia, in a cadaveric study, and described an atlas with an inner sagittal diameter of ≤26 mm as "hypoplasia of the atlas." Hypoplasia of the atlas could be secondary to premature fusion of the cartilaginous neurocentral synchondrosis that occurs between the 6th week and the 4th month of gestation.[10,11] Only 22 patients with symptomatic atlas hypoplasia have been reported, to date. Almost all previous patients developed symptoms as adults, indicating that clinical changes occur in adulthood in addition to the congenital canal stenosis at the level of the atlas. Yamahata et al.[12] proposed a "two-hit pathophysiology," stating that several pathologies such as ossification of the ligament, retro-odontoid pseudotumor disorders, atlantoaxial subluxation, and os odontoideum may contribute to symptom onset, in addition to the congenital small atlas size. Previous reports also stated that spinal cord compression can be suspected if the sagittal canal diameter is less than 14 mm.[13,14] In our patient, the congenitally narrowed atlas, itself, would not have caused symptoms, unless complicated by age-related degenerative changes. Degenerative lower cervical spondylosis decreased the patient's range of motion at the same level, which increased the range of motion of the atlas. Subsequently, long-term and subtle mechanical stress might have led to symptomatic ossification of PAAM, despite the lack of apparent atlantoaxial instability. Similar to the patient with concurrent PAAM ossification and an ossified transverse ligament described by Shoda et al.,[5] secondary factors, namely, PAAM ossification, triggered our patient's symptoms, in addition to the narrowed spinal canal in the atlas secondary to the ossified transverse ligament.

All eight reported patients underwent laminectomy of the atlas; two patients with instability underwent atlantoaxial fusion, and all eight had good outcomes following decompression (Table 1). Atlantoaxial fusion should be performed to prevent iatrogenic instability and correct a primary pathogenesis; however, the risk of vertebral arterial injury must be considered. The postoperative risk of symptomatic progression of instability following decompression has not achieved consensus. We did not perform atlantoaxial fusion in our patient, and no postoperative worsening was seen during follow-up. Patients with apparent preoperative instability should undergo atlantoaxial fusion.

Table 1 Previous reports of patients with ossification of the posterior atlantoaxial membrane

Author	Age/sex	Ethnicity	C1/2 instability	Concurrent disease	Interval between symptom and diagnosis	Coexisting ossification	Treatment (all cases underwent ossified lesion resection)	Adhesion between dura and PAAM	Outcome
Yamaguchi et al.[1]	46 / M	Asian	N / A	N / A	N / A	No	C1 Laminectomy	N / A	Improved
Kimura et al.[2]	55 / F	Asian	N /A	None	5 months	No	C1 Laminectomy	N / A	Improved
Hariyama et al. [3]	52 / M	Asian	No	None	3 months	Anterior longitudinal ligament	C1 Laminectomy	No	Improved
Nadkarni et al. [4]	30 / M	Asian	No	None	6 months	No	C1 Laminectomy	No	Improved
Shoda et al.[5]	70 / M	Asian	No	None	6 years	Transverse ligament	C1 Laminectomy	No	Improved
Ohya et al.[6]	46 / F	Asian	Yes	Os odontoideum	1 month	No	C1 Laminectomy, C1-2 fixation	No	Improved
Meng et al.[7]	39 / M	Asian	Yes	Atlas hypoplasia	6 months	No	C1 Laminectomy, C1-2 fixation	No	Improved
Present case	81 / M	Asian	No	Atlas hypoplasia	2 years	No	C1 Laminectomy, C2 Partial Laminectomy	No	Improved

C: cervical vertebra, F: female, M: male, PAAM: posterior atlantoaxial membrane.

Conclusion

We reported an older patient with cervical myelopathy secondary to a unique interaction between PAAM ossification and atlas hypoplasia. Even without instability, age-related degenerative changes carry the risk of worsening myelopathy, with a narrowed spinal canal and stenosis at the level of the atlas.

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Conflicts of Interest Disclosure

All authors declare no conflict of interest.

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