Myelopathy Due to Multilevel Cervical Canal Stenosis With Forestier Disease
—Case Report—

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Abstract
A 56-year-old woman presented with multilevel myelopathy associated with Forestier disease (FD). The patient was hospitalized for dysphagia, bilateral shoulder pain, and progressive gait disturbance. The diagnosis was confirmed by radiography and magnetic resonance imaging which demonstrated coalescent anterior osteophyte formation extending from C2 to C7 with atlanto-axial dislocation and persistence of mobility at C3-4, and a hyperintense area in the spinal cord at the C1 and C3-4 levels on T2-weighted images. Dynamic radiography showed no instability at the C1 and C3-4 levels. Decompressive laminectomy of the atlas, dome-like laminectomy, and facet fusion at C3-4 were performed, resulting in symptomatic improvement. Myelopathy is very rare in patients with FD and is due to mechanical stress at the level where mobility persists. Strategic intervention should be considered based on evaluation of mobile levels and stenotic lesions.

Key words: Forestier disease, atlanto-axial dislocation, cervical myelopathy, ankylosing spinal hyperostosis

Introduction
Forestier disease (FD) is a progressive skeletal disorder characterized by massive ossification of the anterior longitudinal ligament (ALL) that forms a bridge on the anterior border of the spine. FD was originally described in 1950,6) and is also known as diffuse idiopathic skeletal hyperostosis,12) hyperostotic spondylosis, or ankylosing spinal hyperostosis. The major symptom is dysphagia. The etiology and genetic background of FD remain unknown.
but mechanical factors, drugs, fluoride levels in both water and air, and metabolic conditions have been reported as possible causes.

Here, we describe a case of FD presenting with multilevel myelopathy.

Case Report

A 56-year-old woman was referred to our hospital with headache and pain radiating to the bilateral upper arms. She had a history of unexpected neck rotation. She also had progressive gait disturbance and dysphagia that had developed 3 days after the onset of headache. Past medical and familial histories were unremarkable.

On admission, the patient was alert and oriented. Neurological examination revealed mild dysphagia with no other cranial nerve abnormalities, mild numbness, clumsiness of the bilateral fingers, and gait disturbance due to spastic paresis. Pyramidal signs with brisk deep tendon reflexes in all four extremities and bilateral pathologic reflexes were evident. Markedly decreased distal joint and vibratory sensations were noted in both the upper and the lower extremities. Pinprick and temperature sensations were normal. The active daily living criteria for cervical myelopathy score of the Japanese Orthopedic Association (JOA score) was 7.5 of a possible 17 points. Clinical laboratory data were within normal ranges.

Lateral radiography of the cervical spine on admission demonstrated hyper-ossification of the ALL from C2 to C7, with persistence of mobility at C3-4 (Fig. 1A–C). Dynamic radiography showed no instabilities at the cranio-cervical junction or the atlanto-axial junction (Fig. 1B, C). Power’s ratios were 0.78, 0.96, and 0.73 at the neutral, ante-flexion, and retro-flexion positions, respectively. Initial serial high resolution computed tomography of the cervical spine demonstrated canal stenosis at the C1 level due to atlantoaxial dislocation (AAD). The atlanto-dental interval was 3.5 mm and space available for the spinal cord (SAC), measured as the distance from the posterior surface of the dens to the anterior surface of the arcus dorsalis, was 9.5 mm. The intervertebral disk spaces of the cervical spine were comparatively preserved from C1 to C7. Sagittal T2-weighted magnetic resonance (MR) imaging of the cervical spine also showed canal stenosis at the C1 and C3-4 levels with cord compression and concomitant intramedullary hyperintensity (Fig. 2A). Lateral radiography of the thoraco-lumbar spine demonstrated a mild osteophyte protruding in the ventral direction whereas the sacroiliac joint appeared normal. The diagnosis was AAD with FD, and multilevel myelopathy caused by FD at the C1 and C3-4 levels.

Laminectomy was performed at the C1 and dome-like laminectomy with facet fusion at the C3-4 level. There were no complications during the operation or the postoperative period. Neurological function improved remarkably after the surgical intervention. Dysphagia, mild numbness, clumsiness of the bilateral fingers, and spastic gait were remarkably improved, and she could perform daily living activities without assistance. Postoperative sagittal T2-weighted MR imaging of the cervical spine demonstrated an enlarged vertebral canal width at the C1 and C3-4 levels (Fig. 2B). Her JOA score was 14/17 and quality improvement ratio was 68% at 2 weeks after surgery. She was discharged 15 days after the operation. At 2 years follow up, no deterioration of neurological findings was detected and dynamic radiography revealed no instability at the operative site (Fig. 1D–F).

Discussion

The roentgenographic criteria for the diagnosis of FD include: calcification and ossification along the anterolateral aspect of at least four contiguous vertebral bodies; relative preservation of intervertebral disk height; and absence of apophyseal joint bone ankylosis and sacroiliac
The most frequent clinical manifestation of FD in the cervical spine is dysphagia.\(^1\,^{10}\) Other major symptoms are neck pain, back pain, and mild gait disturbance. FD is generally diagnosed at the time of cervical cord injury or vertebral fracture caused by trauma. Neurological symptoms associated with FD are unusual. The neurological deficit is attributable to myelopathy in the lower cervical spine\(^4\) associated with stenosis of the spinal canal. Reported cases of FD in the craniocervical junction are very rare,\(^9\) and are associated with either AAD or myelopathy caused by posterior compression due to pseudoarthrosis between the posterior tubercule of the atlas and spinous process of the axis.\(^7\) Only three cases of myelopathy were caused by AAD\(^2,7,10\) and one of basilar impression\(^11\) related to FD.

In the present case, the cause of myelopathy at the C3-4 level is speculated to be mechanical stress involving increased use at the C3-4 level where mobility persisted, resulting in secondary bulging of the degenerated intervertebral disk and hypertrophy of the ligamenta flava.\(^9\) Furthermore, the myelopathy at the craniocervical junction where mobility also persisted was apparently due to the association of AAD with narrowing of C1 and the dens of C2. Surgical indications for the present case were considered to include progressive clinical symptoms, JOA score of less than 10 out of a possible 17, and SAC less than 13 mm. Simultaneous surgical intervention for fusion and decompression was recently recommended in the case of FD.\(^{10}\) Single intervention, fusion or decompression, might lead to increased mechanical stress because of the complicated etiology of FD.\(^9,\,^{14}\) Dynamic radiography showed no instabilities at the craniocervical junction or the atlantoaxial junction, so neither occipitocervical fixation nor stabilization of the atlantoaxial subluxation with posterior instruments was indicated in the present case. Thus, decompressive laminectomy at C1 and domelike laminectomy at C3-4 were performed to achieve posterior decompression of the spinal canal. In addition, facet fusion at the C3-4 level was performed to stabilize this level postoperatively.

Ankylosing spondylitis must be considered in the differential diagnosis of FD and is recognized as a disorder frequently associated with cervical spine fractures. Complication with fractures in FD occurs in less than 1% with 20% bone involvement, so dispersion of the external force is caused by the presence of discontinuous compartments affected by bony ankylosis and the occurrence of partial flexibility of the spine due to intact facet joints. Therefore, the external force is thought to be concentrated in these discontinuous compartments in the patient with FD. Based on these features of FD, cervical myelopathy may readily develop in a discontinuous compartment of ALL exposed to external force from trauma, and even chronic mechanical stress can cause cervical myelopathy in the patients with FD.

Symptomatic FD cases are very rare, and prompt and appropriate treatment is important. In the preoperative state, discontinuities of ossification of the ALL in adjacent intervertebral spaces must be evaluated. Strategic intervention should be considered not only at the involved cervical level, i.e. the origin of symptoms, but also at other levels which may develop problems in the future in adjacent intervertebral spaces and mobilities due to discontinuities of ossification of the ALL. Even if there are no neurological symptoms at the time of diagnosis, we should thoroughly evaluate the possibility of myelopathy developing at the level of discontinuities of the ALL. Furthermore, the optimal treatment must be considered for each patient.

References


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