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**A CASE OF CERVICAL MYELOPATHY  
WITH DEVELOPMENTAL CANAL STENOSIS  
AT THE LEVEL OF THE ATLAS  
A CASE REPORT**

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**INDEXING WORDS**

cervical myelopathy; developmental canal stenosis; atlas; craniocervical junction

**SYNOPSIS**

The craniocervical junction is one of the most common sites of malformations.<sup>7,12,14)</sup> Only three cases of myelopathy due to hypoplasia of atlas have been reported previously. Among these malformations, the hypoplasia of atlas was first described by Wackenheim in 1974.<sup>15)</sup> Although developmental canal stenosis due to the hypoplasia of atlas seems to have a tendency of causing the cervical myelopathy, only three cases of cervical myelopathy due to this condition have been reported previously. A 77-year-old man with severe canal stenosis at the level of the atlas is reported. The clinical manifestations were 20-year history of progressive gait disturbance and paresis of both upper and lower extremities. The spinal cord was markedly compressed at the level of the atlas. The clinical manifestations improved after a resection of posterior arch of the atlas.

**INTRODUCTION**

Developmental cervical canal stenosis as congenital anomaly usually causes the cervical myelopathy with the existence of spondylotic changes, vertebral instability, or hyperextension injury. But the cases of the cervical myelopathy due to only the narrow canal itself without any other condition have been reported. The clinical manifestations by the developmental canal stenosis are either the one of myelopathy or the one of radiculopathy.<sup>4)</sup> Developmental canal stenosis occurs

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usually below the C3 vertebral level but rarely above C2 vertebral level.<sup>1,3,9)</sup> Three cases at the level of the atlas<sup>6,11,13)</sup> and one at the axis<sup>10)</sup> have been reported previously. We describe a case of developmental cervical canal stenosis at the level of the atlas, presenting cervical myelopathy.

### CASE REPORT

This 77-year-old man had a 20-year history of progressive gait disturbance and paresthesia of all four extremities. During the year before admission, the paresthesia had become continuous, with exacerbation upon hyperextension of the neck. There was no history of trauma or loss of consciousness.

There was a slight motor weakness of all four extremities, especially proximally, and paresis of all four limbs. The lower limbs were spastic; the deep tendon reflexes were exaggerated in both upper and lower limbs. Hoffmann's signs and Babinski's signs were positive bilaterally.

The limitation of ADL in this patient was evaluated as four points followed by the criteria for evaluation of the operative results of patients with cervical myelopathy by the Japanese Orthopaedic Association (JOA score).<sup>5)</sup>



a.

b.

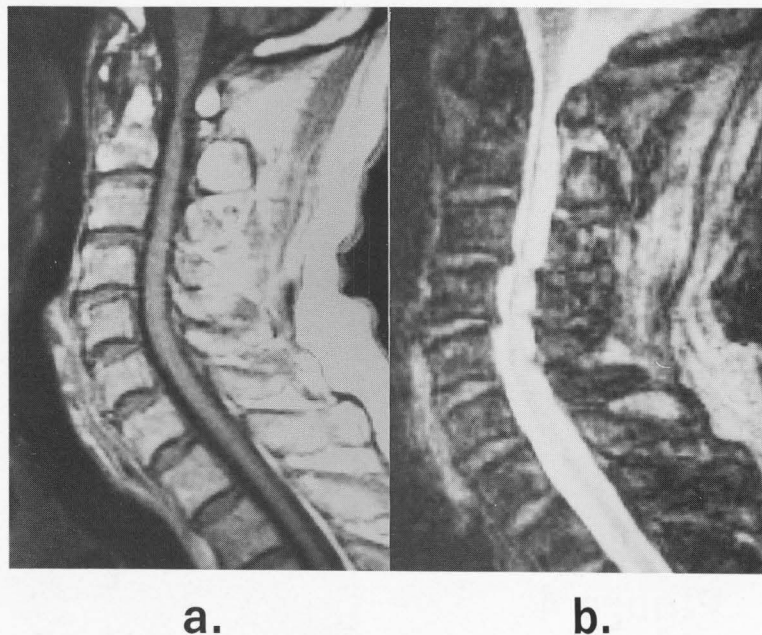
**Figure 1.**

Figure 1. a. Plain X-ray film, lateral view, of the cervical spine showing the marked narrowing of the spinal canal at the level of the atlas. b. The sagittal canal diameter between the posterior margin of the dens and posterior arch of the atlas was 11 mm on cervical tomographs.

## CANAL STENOSIS AT THE LEVEL OF THE ATLAS

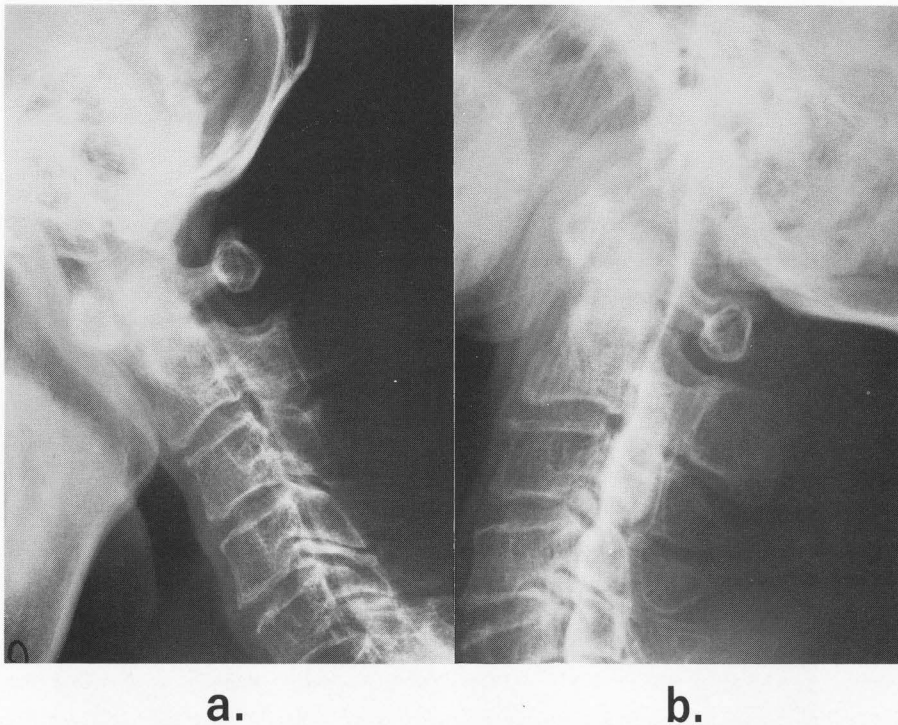
Plain X-ray films of the cervical spine revealed a markedly narrow canal at the level of the atlas. The sagittal canal diameter between the posterior margin of the dense and posterior arch of the atlas was 11 mm on cervical tomographs (Figure 1). Magnetic resonance imaging (MRI) showed narrowing of the canal at the level of the atlas and spinal cord compression (Figure 2). In addition, mild spondylosis was seen at the level of C4 and C5 on MRI. Cervical myelography demonstrated the spinal cord attenuated by the posterior arch of the atlas (Figure 3). Computed tomographic (CT) myelography revealed compressed cord by the posterior arch of the atlas without dislocation from the dense or spondylosis (Figure 4).

Resection of the posterior arch of the atlas and occipito-cervical fixation by the sublaminar wiring were performed (Figure 5). The posterior arch of the atlas was found to encroach on the spinal cord through the dura membrane. After the removal of the posterior arch, the constriction band of the dural membrane was confirmed. On 6 months after the surgery, JOA score is improved to be five points. Although the improvement of neurological findings is not found objectively, the improvement of gait disturbance and fine movement is recognised subjectively.



**Figure 2.**

Figure 2. a. Magnetic resonance T1-weighted image showing focal constriction of the dural sac at the level of the atlas. b. Magnetic resonance T2-weighted image showing spinal cord compression at the level of the atlas.



**Figure 3.**

Figure 3. a. b. Cervical myelograph demonstrating the spinal cord attenuated by the posterior arch of the atlas. Myelograph of flexion/extension film confirming stability at the C1, C2 level.



**Figure 4.**

Figure 4. CT myelograph showing spinal cord compression at the level of the atlas.

Figure 5. Occipito-cervical fixation by the sublaminar wiring .



**Figure 5.**

## CANAL STENOSIS AT THE LEVEL OF THE ATLAS

### DISCUSSION

Cervical myelopathy evoked by developmental canal stenosis at the level of the atlas has been described in the only three reports.<sup>6,11,13)</sup> In our case, the sagittal canal diameter at the level of the atlas is 11 mm, that indicates a milder narrow canal compared with other three cases. But symptoms have been gradually revealed during the period of 20 years, which is the longest period of time in comparison with other three cases.

In embryology, various types of congenital anomalies occur at the craniocervical junction, which cause cervical myelopathy or occipitalgia occasionally. Developmental canal stenosis is one of these anomalies, that can be complicated by spondylosis, hyperextension injury or ligament hypertrophy, presenting myelopathy.

The average normal sagittal canal diameter has been reported to range from 17 to 25 mm at the level of the atlas and the average of the cord diameter ranges from 10 to 12 mm.<sup>3)</sup> Although the compression of the spinal cord is reported to be found with 14mm or less of the sagittal diameter of the spinal canal,<sup>2)</sup> the clinical manifestations started to be recognized with less than 10mm of the canal diameter.<sup>7)</sup> In our case, the sagittal canal diameter at the level of the atlas is 11mm and the clinical manifestations appeared during adulthood even without instability or an obvious history of injury. The gradual appearance of the clinical symptoms of cervical myelopathy during many years like this reported case tends to delay the clinical diagnosis. Therefore, once the patient shows himself to the clinic, his myelopathy ends up to be the severe one, which makes his surgical result worse because of the highly damaged spinal cord.

The developmental canal stenosis at the level of the atlas without any instability in addition to atlantoaxial subluxation, basilar invagination, and other common abnormalities should be considered to evoke the cervical myelopathy.

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