Atlas hypoplasia as a cause of high cervical myelopathy

Case report

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✓ A high cervical myelopathy due to atlas hypoplasia is described in a 56-year-old man; the condition caused marked segmental compression of the spinal cord. A remarkable neurological recovery followed decompressive laminectomy of the atlas and adjacent regions. The authors discuss the embryology and etiology of this anomaly.

KEY WORDS • atlas • hypoplasia • cervical myelopathy • cervical spine

THE craniovertebral junction is one of the most common sites of malformations.1,2 One such malformation, hypoplasia of the atlas, was first described by Wackenheim3 in 1974. Atlas hypoplasia rarely causes myelopathy; the neurological and neuro-radiological findings and treatment have been reported for only one patient with this condition.3 This paper describes a severe case of high cervical myelopathy caused by atlas hypoplasia.

Case Report

This 56-year-old man was admitted to Kensei General Hospital after he hit his forehead during a fall. The patient had an 11-year history of progressive gait disturbance and spasticity of the lower extremities as well as weakness of the upper extremities; he was otherwise healthy. On admission, he was unable to stand and his hand coordination was impaired. Urination was also difficult.

Examination. Physical examination revealed a moderate weakness of the musculature in both arms, especially distally, and paresis of both lower limbs. No fasciculations were observed, but there was evidence of atrophy of all four extremities. Muscle tone was increased markedly in all four limbs, and there was diminution of all sensory modalities, including joint position sense and vibratory appreciation. He had hyperreflexia with bilateral Babinski signs.

Plain radiographs of the craniovertebral junction showed marked narrowing of the spinal canal at the atlas level; the retrodental space was only 7.7 mm (Fig. 1). The sagittal diameter of the spinal canal ranged from 11.7 to 12.6 mm at the other cervical spine levels. There was no evidence of atlantoaxial subluxation or basilar invagination. Magnetic resonance imaging

Fig. 1. Plain x-ray film, lateral view, of the craniovertebral junction showing marked stenosis of the spinal canal at the level of the atlas.
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Fig. 2. Magnetic resonance T1-weighted image showing gross focal constriction of the dural sac at the level of the atlas.

showed gross focal constriction of the dural sac at the atlas level due to a hypoplastic posterior arch; however, no abnormal intensity was demonstrated in the spinal cord (Fig. 2). Computerized tomographic myelography revealed hypoplasia of the atlas and a seriously compressed spinal cord (Fig. 3), as well as some thickening of the soft tissue behind the odontoid and displacement of the odontoid between the lateral masses of the atlas.

Operation. The patient underwent laminectomy of the atlas, decompressive laminoplasty of the axis, and occipital decompression of the foramen magnum. The aims of these surgical procedures were decompression of the spinal cord and correct alignment of the neural structures. At surgery, normal bone tissue and an atrophic ligamentum flavum were observed. The dural sac was markedly pinched anteroposteriorly by the hypoplastic posterior arch of the atlas.

Postoperative Course. Some improvement in muscle weakness was evident on the day after surgery, and urinary incontinence resolved within 2 weeks. At 2 months after surgery, the patient was able to walk with assistance and upper-extremity strength was almost fully recovered, except for mild residual hyperreflexia and slowing of fine hand movement. Magnetic resonance imaging confirmed decompression of the cervicomедullary junction.

Discussion

to understand craniovertebral junction anomalies, it is necessary to know the embryology and anatomy of this area. The majority of the atlas originates from the rostral half of the first cervical sclerotome, which is the most unique point in the development of the atlas. The chondrification of the posterior arch begins at the pedicles, during the 6th week of embryogenesis and ends in the midline during the 4th month. This cartilaginous arch ossifies by 3 to 4 years of age. Hypoplasia of the atlas could be due to premature fusion of the cartilaginous neurocentral synchondrosis that occurs between the 6th week and the 4th month of gestation. The average normal sagittal canal diameter has been reported to range from 17 to 25 mm at the level of the atlas; in contrast, the cord diameter ranges from 10 to 12 mm. This disparity accounts for the lower incidence of symptomatic developmental canal stenosis at this level. Patients with a sagittal canal diameter of less than 10 mm usually show clinical manifestations; in our patient, the canal diameter was only 7.7 mm. The lower cervical canal diameters in our patient also indicated a mildly narrow canal; the normal range at the C3–7 levels is 14 to 20 mm. Considering the unique developmental process of the atlas, the narrow canal at C3–7 in our patient may be due to the extremely hypoplastic atlas; the canal is expected to be wider at the C-1 level, even in cases of cervical canal stenosis.

Wackenheim described a case of atlas hypoplasia with occipital neuralgia and functional obstruction of the foramen of Magendie; however, in his patient, the stenosis was so mild that it did not cause myelopathy. We found only one report of atlas hypoplasia with myelopathy. Sawada, et al., presented a case of a 38-year-old Japanese man with a 3-year history of progressive tetraparesis. His canal diameter was 7 mm at the level of the atlas and 12.5 to 15.0 mm at the lower cervical levels. His clinical manifestations improved after posterior laminectomy of the atlas. The neurological and neuroradiological presentations were similar to those of our patient: it was an adult man with progressive deterioration and sparing of respiratory function, who benefited from posterior decompression. In these two patients, the clinical manifestations appeared during adulthood in spite of the congenital anomaly. This means that the encasement of neural content by a hypoplastic atlas does not result in serious neurological manifestations until the occurrence of complications.
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during the aging process, such as spondylosis, ligament hypertrophy, venous stasis with cord edema, or injury due to hyperextension. In our case, displacement of the odontoid between the lateral masses of the atlas and thickening of the soft tissue behind the odontoid were observed. These changes may be the sequelae of previous trauma. The compressed spinal cord was atrophic in both cases; however, function was restored by posterior decompression.

When encountering a patient with high cervical myelopathy, the diagnosis of atlas hypoplasia in addition to atlantoaxial subluxation, basilar invagination, and other common abnormalities should be considered.

References


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