Atlas Hypoplasia Manifesting as Myelopathy in a Child
—Case Report—

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Abstract

A 14-year-old Japanese boy presented with myelopathy due to atlas hypoplasia with complete posterior arch. Decompressive laminectomy of the atlas produced good neurological recovery, and follow-up T2-weighted magnetic resonance imaging showed disappearance of spinal cord edema. Congenital atlas stenosis may be symptomatic even in children, with no accompanying cervical spondylotic change. Such cases have previously occurred only in Asian adults. A radiological study of the patient’s brother showed median cleft formation of the posterior arch of atlas, indicative of a wide spectrum of atlas anomalies and a possible genetic relationship between these anomalies.

Key words: atlas, cervical canal stenosis, hypoplasia, posterior arch, median cleft, congenital abnormality

Introduction

Atlas hypoplasia with complete posterior arch is a rare congenital abnormality, exclusively occurring in Asian people, except for one case the details of which are unknown. Clinical manifestation characteristically occurs late in adulthood when cervical spondylotic changes become apparent. We treated a child suffered myelopathy with atlas hypoplasia associated with no degenerative changes. A radiological study of the patient’s brother suggested a possible genetic relationship between atlas hypoplasia and median cleft formation of the posterior arch of the atlas.

Case Report

A 14-year-old boy had a 7-month history of progressive gait disturbance and weakness of the upper extremities. On admission he was found to be short with height of 151 cm below the mean − 2 SD. Neurological examination showed mild quadriplegia (4/5), hyperreflexia, and spasticity in all four limbs. He had no apparent sensory disturbances. He has no family history of Down’s syndrome, achondroplasia, gonadal dysgenesis, or Turner’s syndrome.

Radiography of the cervicovertebral junction showed marked narrowing of the spinal canal at the atlas level, with retrodental space of 10 mm (Fig. 1A). The sagittal diameter of the spinal canal ranged from 12 to 16 mm at the other cervical spine levels (C-2, 16 mm; C-3 and C-4, 12 mm; C-5, 13 mm; C-6, 14 mm; C-7 15 mm). There was no evidence of basilar invagination, occipitalization, or atlantoaxial subluxation. Computed tomography (CT) showed a hypoplastic atlas with an asymmetrical posterior arch, the left side thicker than the right (Fig. 1B). T2-weighted magnetic resonance (MR) imaging disclosed constriction of the dural sac, especially at the atlas level and hyperintense change in the spinal cord (Fig. 1C). Low signal area anterior to spinal cord from C-1 to C-4 may be due to thick posterior longitudinal ligament, engorgement of venous plexus, or bony canal stenosis.

The patient underwent laminectomy of the atlas. The dural sac was markedly constricted by the hypoplastic posterior arch of the atlas. The midline portion of the inner surface of the C-1 lamina had a beak-like protrusion to the spinal canal that was seen on the preoperative CT scan (Fig. 1B). After laminectomy, the dural sac expanded, good pulsation was confirmed, and there was no constriction band on the sac.

Muscle weakness and spasticity in the four limbs gradually resolved after surgery, and had disap-
Fig. 1  A: Radiograph showing marked narrowing of the spinal canal at the atlas level with a relatively narrow spinal canal through C3–5. B: Computed tomography scans showing atlas hypoplasia with complete posterior arch. Note the beak-like protrusion to the spinal canal in the midline portion of the inner surface of the C-1 lamina (upper) and the thicker posterior arch of the atlas on the left than on the right (upper and lower). C: T2-weighted magnetic resonance (MR) image showing constriction of the dural sac at the atlas level and hyperintense change in the spinal cord. D: T2-weighted MR images one year after the C-1 laminectomy showing decompression of the cervicomedullary junction and remarkable decrease in size of the hyperintense lesion in the spinal cord.

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Fig. 2  Radiograph (A) and computed tomography scans (B) showing slit cleft formation of the posterior arch of the atlas in the second brother of the patient.

Symptomatic developmental cervical stenosis with a complete posterior arch at the level of the atlas has been reported in 10 patients. All patients had myelopathic symptoms, except for one of unknown age and sex who had occipital neuralgia. The patients were aged 38, 54, 55, 69, 73, 75, 77, 80, and 81 years. Their histories of myelopathy ranged from 4 months to 11 years, indicating that clinical manifestations characteristically occur somewhat late in adulthood, although atlas hypoplasia is a congenital anomaly. Atlas hypoplasia is generally considered to rarely cause myelopathy unless spondyloitic change in the lower cervical spine occurs, although a sagittal diameter of the C-1 canal of less than 10 mm indicates a high risk of clinical signs and symptoms. Decreased range of motion at the lower cervical levels caused by spondylosis is believed to be important in inducing increased movement of the atlas with subsequent compression of
the spinal cord.\textsuperscript{4,9} Retrodental fibrous granulation is an additional causative factor for myelopathy in elderly patients with atlas hypoplasia.\textsuperscript{4,13} This soft tissue mass appears to be produced by chronic atlantoaxial subluxation or osteoarthrosis of the atlantoaxial joint.\textsuperscript{4,13} All reported cases, including ours, involving atlas stenosis with a complete posterior arch occurred in Asian people, predominantly in men, indicative of ethnic association and sex dominance.\textsuperscript{3,4,6,8–11,13} The cases of brothers with atlas hypoplasia suggested a genetic factor.\textsuperscript{9} Our case shows that myelopathy may be evoked even in a young patient with atlas hypoplasia with complete posterior arch and in the absence of spondylotic change. Dynamic neck movement could cause spinal cord damage. When the atlas hypoplasia is found in a young patient, Down’s syndrome, gonadal dysgenesis, achondroplasia, or Turner’s syndrome must be ruled out, as these are frequently associated with C-1 anomaly.\textsuperscript{5}

Atlas stenosis may be caused by premature fusion of the two neurocentral and one posterior cartilaginous synchondroses during the 6th week to 4th month of gestation.\textsuperscript{2,7} As a result, the three primary ossification centers among these three synchondroses may not grow away from each other.\textsuperscript{1,3,7,9} Continuous growth of the lateral sides to the fused posterior cartilaginous synchondrosis would result in a thick posterior arch of the atlas, as shown in our patient. The median cleft of the atlas found in a brother of our patient with atlas hypoplasia, if not coincidence, suggests that common genetic factors are involved in the two different forms of congenital malformation of the atlas. The frequency of posterior median cleft, which represents approximately 90% of all congenital abnormalities of the posterior arch of the atlas, is 3% to 4% of the population.\textsuperscript{2,9} The underlying embryological failure in this anomaly occurs at the level of local chondrogenesis rather than primary failure of ossification.\textsuperscript{1,9} When the cartilaginous gap, that is itself a posterior synchondrosis, is larger than the normal size (3–4 mm) or fibrous, spina bifida develops.\textsuperscript{7} Posterior median cleft of the atlas is therefore related to failure of normal fusion of the posterior synchondrosis. Stenosis and posterior median cleft of the atlas could be considered as different forms of different stages during the fusion process of posterior synchondrosis. Five cases of Indian male children had bifid posterior atlantal arch with inturning of the ends resulting in cervicomedullary compression.\textsuperscript{2} These cases may indicate the presence of the combination of atlas stenosis and posterior median cleft formation in a wide spectrum of atlas anomalies.

\textbf{References}


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