



Successful Treatment of Basilar Invagination and Platybasia Associated With Cerebellar Atrophy by Decompression Surgery

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Dear Editor,

Craniovertebral junctional abnormalities are rare but underrecognized developmental disorders of the neural axis. The associated conditions include basilar invagination and platybasia, for which there have been only a few related clinical reports,¹ and so the possible pathomechanisms remain unclear. Here we present a case of successful recovery from basilar invagination, platybasia, and cerebellar atrophy after decompressive surgery.

A 41-year-old female patient presented to the clinic with a 2-year history of progressive imbalance. She also complained of voice changes, swallowing difficulties, and dizziness aggravated by upright posture. The patient had no familial loading or developmental delay, and no history of head or neck trauma. A neurological examination revealed dysarthria, dysphagia, gaze-evoked horizontal nystagmus augmented by left gaze, impaired left-sided smooth pursuit with preserved saccades (Supplementary Video in the online-only Data Supplement), and truncal ataxia with veering tendency to the right side. Her motor and sensory functions remained intact. Laboratory evaluations such as inflammation, infection, endocrine, and paraneoplastic antibody tests all produced negative findings. Magnetic resonance imaging (MRI) of the brain and cervical spine showed complete atlanto-occipital assimilation with severe basilar invagination and platybasia, resulting in anterior medullary compression and tonsillar herniation (Fig. 1A and B). Severe cerebellar atrophy was observed, predominantly in the bilateral vermis (Fig. 1D and E). Applying posterior fossa decompression and C1 laminectomy with duroplasty improved the imbalance and bulbar symptoms, including hoarseness and dysphagia, while postsurgery MRI revealed good decompression (Fig. 1C and F).

Previous reports on basilar invagination have focused on surgical outcomes, which vary.^{2,3} Chronic vascular insufficiencies and tonsillar herniation have been suggested as underlying pathomechanisms.¹ However, the treatments applied for anatomical decompression do not always result in good clinical outcomes.³ Therefore, understanding the neuroanatomical basis for the development of a patient's symptoms is critical to achieving an accurate diagnosis and effective treatment. In the present case, magnetic resonance angiography revealed no particular stenosis or occlusion of the cerebral blood vessels. Although the anterior medulla was anatomically compressed, the symptoms and signs of the patient imply that the dorsal medulla, including the nucleus prepositus hypoglossi⁴ and vestibular nuclei, may play primary roles in provoking symptoms. The dorsal part of the medulla abutting the fourth ventricle, known as the area postrema, showed sparse tight junctions in the blood-brain barriers,⁵ which may contribute to a sensitivity to mechanical stress due to the obtuse angulation of platybasia, like in the present case. Since decompressive surgery not only releases local compression but also alleviates crowding among infratentorial structures or ventricu-

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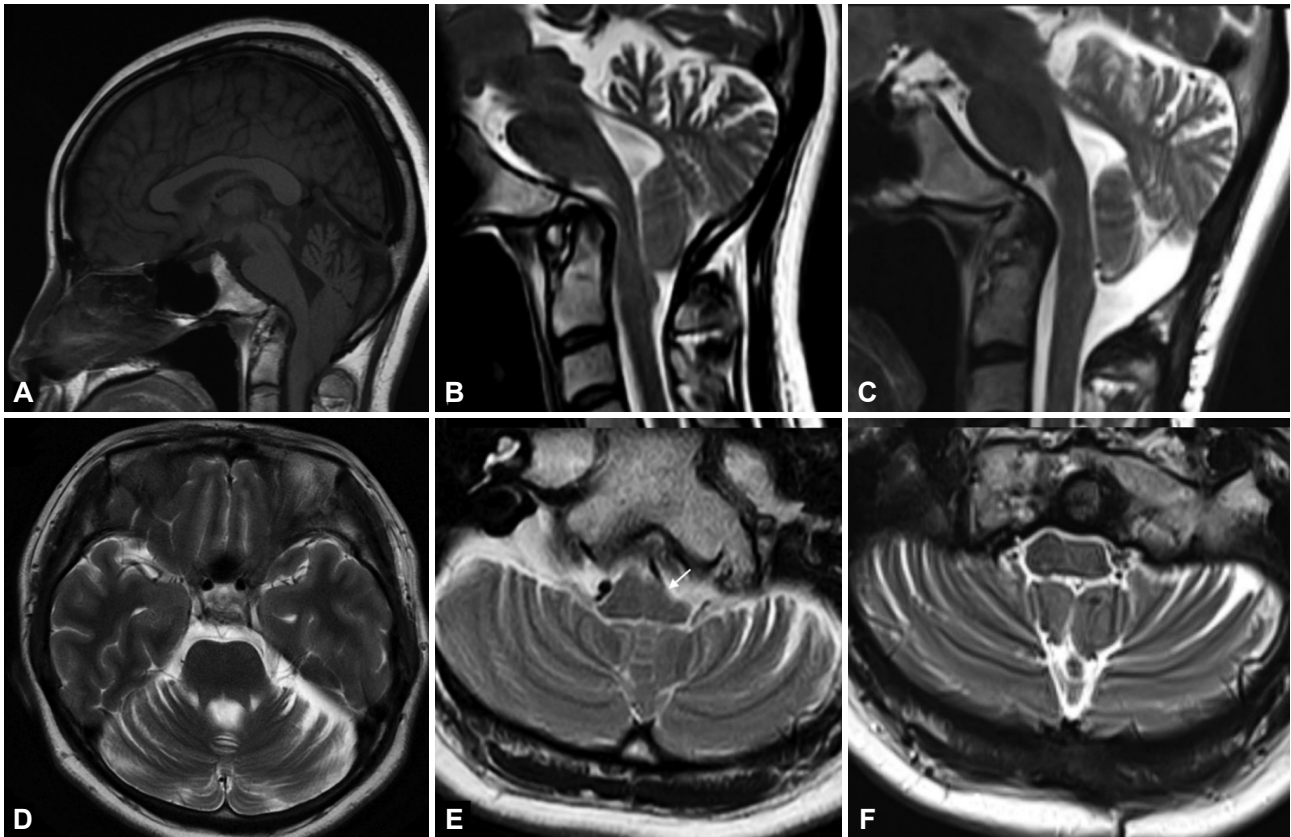


Fig. 1. Radiological findings in the patient. A: Magnetic resonance imaging (MRI) of the brain and cervical spine showing complete atlanto-occipital assimilation with marked basilar invagination, platybasia, and cerebellar atrophy. B: MRI showing severe cerebellar atrophy associated with basilar invagination. C: MRI after surgery revealed good decompression. D: MRI showing severe bilateral cerebellar atrophy, especially in the affected vermis. E: The lower medulla is markedly compressed on the left dorsal surface (white arrow). No signs of brainstem or cerebellar infarction or cerebral cortical atrophy were observed. F: After decompression surgery, the compressed area of the left lower medulla also appeared to have improved.

lar flows, clinical improvement may be achieved through the combinatorial effects of these factors.

Disproportionate atrophy of the cerebellar vermis with preserved tonsillar volume associated with basilar invagination has also been described previously,² like in the present case. Dysfunctioning medullary structures, including the olivary nucleus and nucleus prepositus hypoglossi, may contribute to degeneration of the connecting cerebellar vermis, emphasizing the role of medullary structures and that trans-synaptic neuronal degeneration is likely to be present. Previous animal investigations have shown that cerebellar Purkinje cells and olivary neurons exert mutual trophic effects.⁶ Therefore, in certain circumstances, damage to one may cause loss of the other.

In clinical practice, the onset of progressive ataxia at a young age usually raises the suspicion of developmental or degenerative etiologies, and most treatment options are symptomatic. However, physicians should remember that this could also be caused by rare anatomical problems, like in the present case. In addition, surgical options may be beneficial for patients with basilar invagination and platybasia, including in progres-

sive cases and even when symptoms are localized to the medullary structures. To the best of our knowledge, this is the first case report in Korea of the successful recovery of basilar invagination and platybasia with cerebellar atrophy through decompression, in which medullary structures may have played a role in the development of symptoms in the patient.

Supplementary Video Legend

Video 1. The patient initially presented with gaze-evoked horizontal nystagmus augmented by left gaze, impaired left-sided smooth pursuit with preserved saccades.

Supplementary Materials

The online-only Data Supplement is available with this article at <https://doi.org/10.3988/jcn.2022.18.2.241>.

Ethics Statement

The patient described in this study provided informed consent, and the study design was approved by the appropriate ethics review board.

Availability of Data and Material

The datasets generated or analyzed during the study are available from the corresponding author on reasonable request.

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Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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