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MRI Demonstration of Supratentorial Developmental Abnormalities in Chiari II Malformation: Prognostic Implications in a Treated Myelomeningocele Patient

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Abstract

Chiari II malformation is typically associated with myelomeningocele and hindbrain herniation, but the contribution of supratentorial abnormalities to neurological outcome remains underemphasized. We report the MRI findings of a 10-year-old child with antenatally diagnosed myelomeningocele repaired at birth, presenting motor disability and epilepsy. Brain MRI demonstrated severe Chiari II malformation with marked tonsillar descent, small posterior fossa, and vermian dysplasia, associated with septum pellucidum agenesis, posterior corpus callosum dysgenesis, periventricular nodular heterotopia, and cortical organization abnormalities. Triventricular hydrocephalus with ventriculoperitoneal shunt and extensive closed spinal dysraphism were also identified. This case illustrates that MRI evaluation of Chiari II malformation should extend beyond the posterior fossa because supratentorial developmental abnormalities may substantially contribute to neurological impairment.

Keywords

Chiari II Malformation, Neuronal Migration Disorder, Heterotopia, Spinal Dysraphism, MRI, Myelomeningocele

1. Introduction

Chiari II malformation is a complex hindbrain anomaly almost constantly associated with open spinal dysraphism and hydrocephalus [1]-[4]. It is generally con-

sidered a consequence of abnormal neurulation and cerebrospinal fluid leakage leading to posterior fossa hypoplasia and hindbrain herniation [2] [3].

Beyond the classic hindbrain abnormalities, supratentorial anomalies including corpus callosum dysgenesis and cortical malformations may coexist [5] [6]. These findings suggest that Chiari II malformation may represent a broader neurodevelopmental disorder rather than an isolated neural tube defect [7].

Neuronal migration disorders such as periventricular nodular heterotopia occur later during fetal development and are strongly associated with epilepsy and neurocognitive impairment [8]-[11]. Their identification is therefore essential for prognostic assessment in children with repaired myelomeningocele.

We report a case illustrating an extensive polymalformative spectrum demonstrated by MRI.

2. Case Presentation

A 10-year-old child with antenatally diagnosed myelomeningocele surgically repaired at birth was referred for routine MRI follow-up. The patient presented chronic motor neurological impairment requiring wheelchair use and epilepsy.

The myelomeningocele was located at the lumbosacral level and was surgically repaired during the neonatal period. Hydrocephalus developed early in life and was treated by ventriculoperitoneal shunting. No documented shunt revisions were reported. The patient developed permanent motor impairment of the lower limbs leading to wheelchair dependence. Epileptic seizures appeared during childhood and are currently managed with antiseizure medication. Neurodevelopmental follow-up revealed cognitive difficulties requiring adapted schooling.

2.1. MRI Protocol

MRI was performed on a 1.5-Tesla scanner. The protocol included sagittal and axial T1-weighted sequences, axial and sagittal T2-weighted sequences, and coronal T2-weighted images covering the brain and spinal axis. Tonsillar descent was measured on mid-sagittal T2-weighted images relative to the foramen magnum. Supratentorial abnormalities were assessed according to established neuroradiological criteria for neuronal migration and cortical organization disorders.

2.2. Brain MRI Findings

MRI demonstrated a complex polymalformative pattern organized into posterior fossa, midline, and cortical developmental abnormalities.

Posterior fossa abnormalities (Chiari II malformation)

Severe cerebellar tonsillar herniation measuring 22 mm below the foramen magnum (**Figure 1**) was associated with a small posterior fossa**,** tubular fourth ventricle, and dysplastic vermis and cerebellar hemispheres (**Figure 2(A)**, **Figure 2(B)**).

Midline developmental abnormalities

Agenesis of the septum pellucidum and posterior corpus callosum dysgenesis

(absence of splenium and posterior body) were observed (Figure 2(A)). An inferior anterior falcine deficiency with interdigitating anterior cingulate gyri was also present (Figure 2(C)).

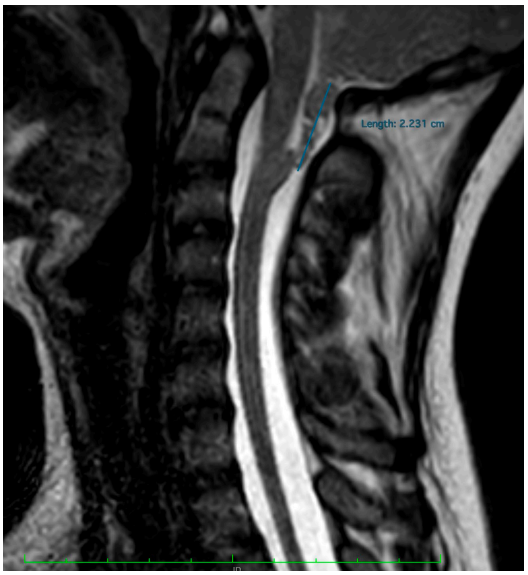

Figure 1. Mid-sagittal T2-weighted MRI demonstrating a small posterior fossa with marked cerebellar tonsillar herniation extending 22 mm below the foramen magnum.

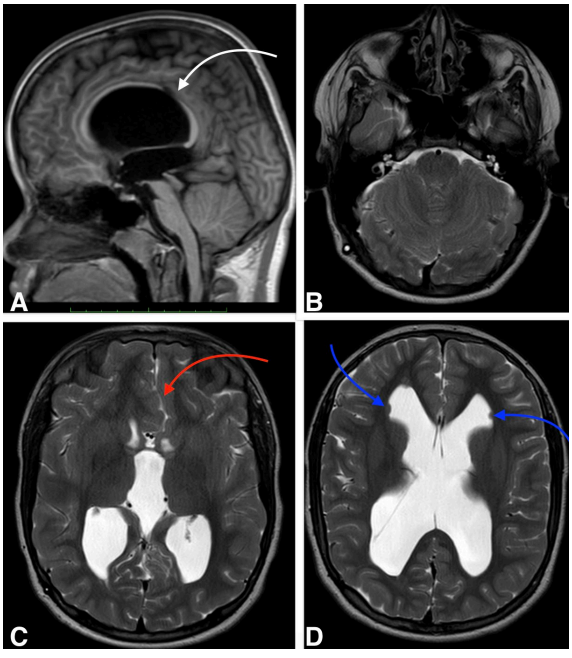

Figure 2. (A) Sagittal T1-weighted image showing posterior corpus callosum dysgenesis with absence of the posterior body (*white arrow*); (B) Axial T2-weighted image demonstrating dysplastic vermis and cerebellar hemispheres; (C) Axial T2-weighted image showing abnormal cortical organization with bilateral stenogyria and interdigitating cingulate gyri (*red arrow*); (D) Axial T2-weighted image demonstrating triventricular hydrocephalus, agenesis of the septum pellucidum and multiple periventricular nodular gray matter heterotopia along the frontal horns (*blue arrows*).

Cortical development abnormalities

Multiple periventricular nodular gray matter heterotopia predominantly along the frontal horns were identified, associated with bilateral stenogyria (Figure 2(D)).

Ventricular system

Triventricular hydrocephalus was present (Figure 2(C), Figure 2(D)). A right posterior parietal ventriculoperitoneal shunt catheter was seen in the right ventricular atrium.

2.3. Spinal MRI Findings

Spinal cord atrophy was present below the cervicothoracic junction without syringomyelia. The conus medullaris was atrophic and terminated at L2 (Figure 3(A), Figure 3(B)).

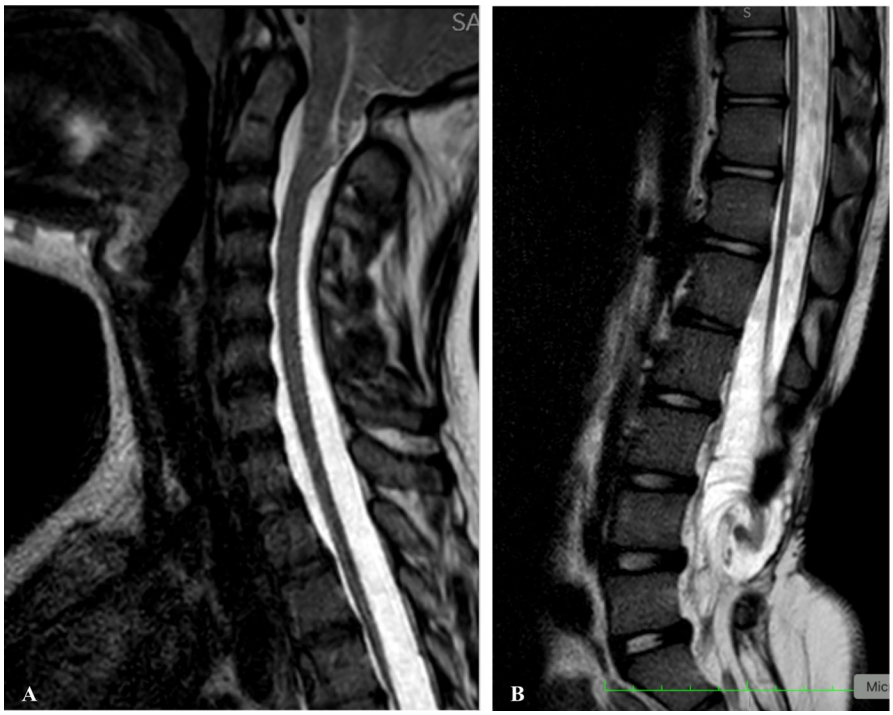

Figure 3. (A) Sagittal T2-weighted image showing spinal cord atrophy below the cervicothoracic junction without syringomyelia; (B) Lumbosacral sagittal image demonstrating extensive posterior spinal dysraphism from L2 to L5 with dural ectasia extending into subcutaneous tissues without cutaneous communication.

An extensive posterior spinal dysraphism extending from L2 to L5 showed dural ectasia protruding into subcutaneous tissues without cutaneous communication (Figure 3(B)).

3. Discussion

Chiari II malformation is traditionally explained by early neurulation failure leading to cerebrospinal fluid leakage and posterior fossa hypoplasia [1]-[4]. However,

the abnormalities observed in this patient extend far beyond the hindbrain and involve multiple stages of central nervous system development.

The hindbrain herniation and associated spinal dysraphism reflect an early defect of neural tube closure. Chiari II malformation is almost invariably associated with myelomeningocele and is considered a consequence of altered embryonic cerebrospinal fluid dynamics [2] [3] [12]. The presence of extensive dysraphism supports a global neurulation disturbance rather than an isolated posterior fossa malformation.

Septum pellucidum agenesis and posterior corpus callosum dysgenesis indicate abnormal midline patterning occurring after neural tube closure. These abnormalities are frequently described in Chiari II and reflect disturbance of commissural formation during early brain organization [5] [6].

Periventricular nodular heterotopia and stenogyria correspond to abnormal neuronal migration occurring between the 8th and 16th gestational weeks [8]-[10].

Periventricular nodular heterotopia was diagnosed based on nodular lesions lining the lateral ventricles that were isointense to cortical gray matter on all MRI sequences. The cortical abnormality characterized by simplified gyral pattern and abnormal sulcation was consistent with stenogyria, which has been previously described in association with Chiari II malformation [13].

These anomalies are strongly associated with epilepsy and neurodevelopmental impairment [9]-[11], explaining the clinical presentation of epilepsy and severe motor disability in this patient.

Hydrocephalus is classically related to CSF circulation impairment in Chiari II. However, long-term neurological outcome correlates poorly with tonsillar descent and more closely with cortical malformations and callosal abnormalities [6] [11]. MRI therefore plays a key role in identifying prognostic factors beyond posterior fossa anatomy [14].

From a clinical perspective, the identification of extensive supratentorial malformations has implications for patient management. These findings support the need for multidisciplinary follow-up including epilepsy management, neuropsychological assessment, and long-term neurological monitoring.

Limitations

One limitation of this report is the absence of detailed longitudinal neuropsychological evaluation, which limits precise correlation between imaging abnormalities and functional outcome. Moreover, conclusions regarding prognosis cannot be generalized from a single case observation.

4. Conclusions

This case illustrates an extensive polymalformative syndrome combining Chiari II malformation, neuronal migration disorders, cortical dysplasia, and closed spinal dysraphism.

This case suggests that supratentorial developmental abnormalities may substantially contribute to neurological impairment in Chiari II malformation, although this observation should be interpreted cautiously given the single-case design.

Comprehensive MRI evaluation is therefore essential for accurate prognostic assessment and clinical management.

Ethical Considerations

Information was collected in accordance with confidentiality requirements and after obtaining the mother's consent.

Authors' Contributions

All authors contributed to the development of this study and declare that they have read and approved this manuscript.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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