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LETTER TO THE EDITOR

Extensive reversible myelopathy secondary to acute quadriventricular noncommunicating hydrocephalus

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Dear Editor:

Hydrocephalus is an active distension of the ventricular system due to various causes. Here, we want to share our experience of a 3.5-year-old girl with acute hydrocephalus and radiologic abnormality of the spinal cord from an underdiagnosed etiology. She was born at 34 weeks of gestation via cesarean section in a primigravida mother. Pregnancy was complicated by intrauterine growth retardation, maternal gestational diabetes, and preeclampsia. There was no family history of any neurodevelopmental diseases. She spent approximately 4 weeks in a neonatal care unit for feeding difficulties and then had repeated admissions in the hospital in early infancy due to failure to gain weight. She was also noted to have facial dysmorphisms, microcephaly, hypotonia, and global developmental delays. A comprehensive workup was completed including brain magnetic resonance imaging (MRI), extensive metabolic workup, and chromosome microarray testing. Her MRI brain at the age of 6 months showed mild bilateral periventricular leukomalacia, mild diffuse white matter volume loss, and mild prominence of the lateral ventricles. Microarray testing revealed a large interstitial deletion in chromosome 7 (starting at q31.2). During the follow-up over the next 3 years, she was diagnosed with atrial septal defect, dysphagia and gastroesophageal reflux requiring gastrostomy tube placement, and severe global developmental delays. She was nonverbal but communicated with gesture and pointing. She also had delayed gross and fine motor milestones with only ability to cruise and grasp objects with an immature grasp at the age of 3 years.

At the age of 3.5 years, she presented with a 2-week history of progressive irritability and headaches with no definitive precipitating factors such as trauma or high-grade fever. She held her right side of the head and neck intermittently with the facial expression of pain and cried in discomfort. She also had episodes of vomiting. Her neurological exam was non-focal and unaltered from her baseline. She had the global appendicular and axial hypotonia but no change in her ability to move the upper and lower extremities against gravity spontaneously and in response to touch. Brain MRI with and without contrast demonstrated acute hydrocephalus with minimal periventricular hyperintensity and enlargement of the 4th ventricle (Fig. 1a, b). Initially, no definitive etiologic diagnosis was possible for this acute hydrocephalus. There was an additional finding of T2 and FLAIR hyperintensity at cervical cord without enhancement which prompted whole-spine MRI that revealed diffuse edema from cervical (C2) to thoracic (T4) segments (Fig. 2a).

Due to diffuse cord expansion and effacement of the CSF space, obstructive hydrocephalus was thought to be secondary to a cord pathology such as inflammatory or neoplastic involvement of the cord. Subsequently, a ventriculoperitoneal (VP) shunt was placed. Cerebrospinal fluid (CSF) obtained from the ventricles during the shunt placement and repeat lumbar puncture 7 days after the procedure showed no evidence of inflammation or neoplastic involvement. Serum aquaporin-4 receptor antibody and myelin oligodendrocyte protein antibody were both negative. Follow-up non-contrast head CT right after the VP shunt

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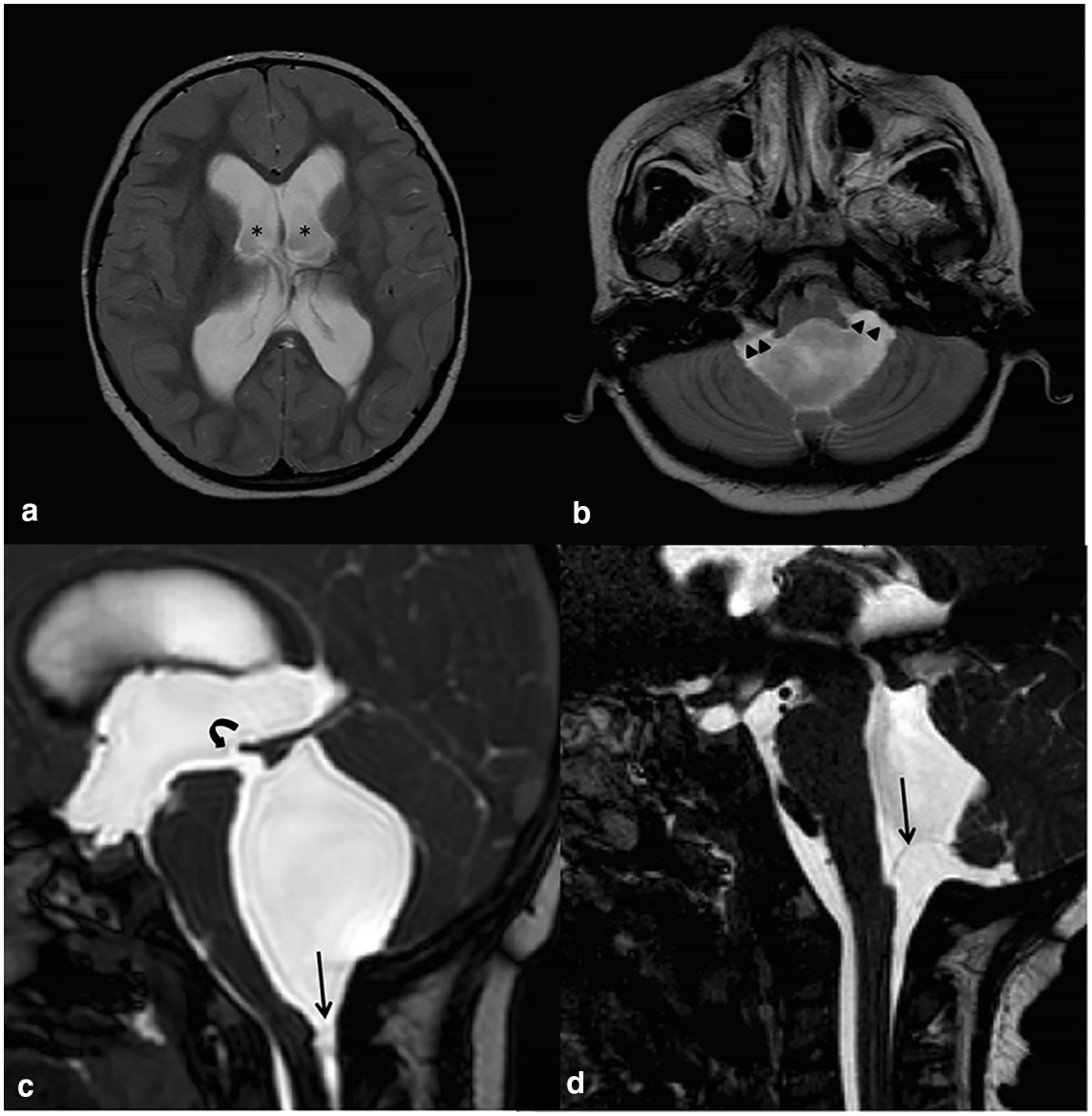

Fig. 1 Axial T2-weighted image (**a**) at the level of the lateral ventricles shows acute hydrocephalus with minimal periventricular hyperintensity suggestive of CSF seepage. Axial T2-weighted image (**b**) at the level of the fourth ventricle shows widened bilateral foramen Luschka with bulging covering membranes. Sagittal high-resolution T2-weighted image (**c**) shows a similar thin membrane occluding the foramen Magendie (arrow). Notice that the cerebral aqueduct is widely open (curved arrow). Sagittal high-resolution T2-weighted image (**d**) following placement of a shunt catheter show fourth ventricular decompression and the occluding membrane covering the foramen Magendie flipping back into the fourth ventricle (arrow)

placement showed decompression of the lateral ventricles, 3rd and 4th ventricles. Reevaluation of the first brain MRI confirmed the diagnosis of the foramen of Magendie obstruction causing quadriventricular hydrocephalus. The repeat spine MRI 10 days after the procedure demonstrated complete resolution of the spinal cord edema (Fig. 2b). The lesions in the spinal cord causing progressive hydrocephalus have been reported in cases with spinal cord tumor, penetrating spinal cord injury, and cervical cord compression. In the present case, the resolution of the spinal cord edema following VP shunt placement supports the inverse etiology where the acute hydrocephalus resulted in spinal cord abnormalities.

The congenital membrane obstruction of foramen Lushka and Magendie noted on the brain MRI contributed to the development of obstructive hydrocephalus. This is a rare and underdiagnosed etiology of noncommunicating quadriventricular hydrocephalus [1]. In children, this is particularly associated with Chiari malformation or basilar impression. This is the first report of the foramen of Magendie obstruction associated with

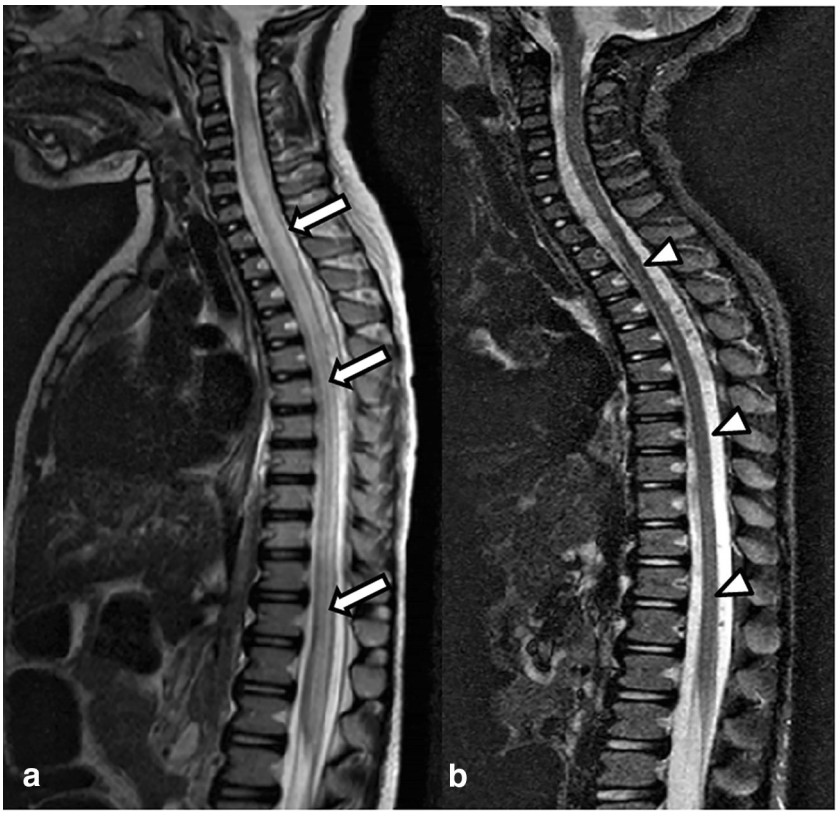

Fig. 2 Sagittal T2-weighted image (**a**) from the initial MRI study of the spine show acute onset central holocord hyperintensity suggestive of edema (arrows). The follow-up T2-weighted sequence (**b**) of the spine following shunt and decompression of the ventricles shows complete resolution of the central holocord edema (arrowheads)

chromosome 7 deletion. MRI flow study and direct visualization of the obstructive membrane are helpful for prompt diagnosis. Preoperative radiologic diagnosis is important, as that might be useful in providing alternative surgical approaches such as surgical exploration and incision of the thickened membrane, rather than using a ventriculoperitoneal shunt with associated long-term risks such as shunt failure and infection.

Saroretti et al. reported two adult cases with transient edema of the cervical spinal cord secondary to obstruction of the CSF pathways [2]. One case developed cervical spinal cord edema secondary to post-meningitic adhesive obstruction of the outlet foramina of the fourth ventricle and the other case with infratentorial mass. It has been hypothesized that nontraumatic obstruction of the CSF pathway secondary to Chari malformation, tumor, and arachnoiditis may produce a spinal cord parenchymal T2 prolongation, known as pre-syrinx [3]. However, acute development of myelopathy secondary to the foramen of Magendie obstruction has not been previously reported, to our knowledge. It is also important to note that this important radiologic finding can be missed easily as it can be clinically silent as noted in this patient. However, untreated patients might develop syringomyelia with disability associated with the pathology.

In the setting of altered CSF flow, as with the foramen of Magendie obstruction, fluid in the subarachnoid space is subjected to increased pressure, especially with systolic pulsation. If the central canal is not patent, rather than the accumulation of fluid within the central canal, diffuse edema of the spinal cord parenchyma develops. However, as seen in our case, the lesions are reversible if prompt decompression therapy is applied. Humphries et al. reported a case of myelopathy secondary to VP shunt failure which was completely resolved by correction of shut placement [4].

We would like to emphasize that the foramen of Magendie obstruction causing quadriventricular communicating hydrocephalus should be specially looked for in the case of prominent 4th ventricular dilatation, especially as the obstructive nature of this rare entity is difficult to demonstrate, even by MRI images, because of the complex morphology of the fourth ventricle. We would also like to impress the possibility of reversible myelopathy in this rare entity.

Compliance with ethical standards

Conflict of interest The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participant included in the study.

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