Epidural blood patch reversing acquired Chiari malformation due to chronic CSF leak


The authors describe the case of a 19-year-old woman with Gorham-Stout disease (GSD), an intractable disorder characterized by massive osteolysis caused by abnormal lymphangiogenesis in bone, presenting with orthostatic headache due to intracranial hypotension (5 cm H2O). Orthostatic headache was associated with a CSF leak from the thigh after pathological fractures of the femur and pelvis. The chronic CSF leak led to acquired Chiari malformation (CM). After an epidural blood patch (EBP) the CSF leak resolved, the CM disappeared, and her neurological status improved.

In the paper, the authors affirm that the association between the CSF leak and CM can be explained as follows. GSD led to the development of the chronic low-grade CSF leak. The CSF leak led to brain sag, including downward displacement of the brainstem and cerebellar tonsils, and acquired CM developed. The CSF leak led to brain sag, including downward displacement of the brainstem and cerebellar tonsils, and acquired CM developed. The authors conclude that cases of GSD presenting with a CSF leak and CM have been previously reported, but reversible CM has never been reported. In the previous cases, patients underwent suboccipital decompression and duraplasty; however, the authors propose sealing the CSF leak rather than suboccipital decompression for acquired CM resulting from a CSF leak.

We would like to add that acquired CM often associates with spontaneous CSF leak and subsequent spontaneous intracranial hypotension (SIH) syndrome. SIH has also been reported in childhood, but patient ages range from 2 to 86 years, occurrence peaks between 35 and 42 years of age, and the disorder is most common in women. SIH typically results from spontaneous CSF leak at the spinal level, especially at the thoracic spine or cervicothoracic junction. The exact cause of the spontaneous CSF leak often remains undetermined. Treatment ranges from conservative management (such as bed rest, over-hydration, and caffeine) to invasive procedures (such as the autologous EBP).4

In our series of more than 400 patients with SIH, observed from 1992 to 2018, about 300 were treated with a lumbar EBP using autologous blood, with good outcome.1–3 The acquired CM was found in about 30% of cases. About 90 patients with acquired CM from SIH were treated using a lumbar EBP with good outcome, as occurs in SIH without acquired CM.

Considering our experience, a lumbar EPB may also exert an immediate effect on orthostatic headache in chronic SIH (from 3 months to 4.5 years duration), as it does in acute SIH cases.2 Furthermore, an EBP may also ameliorate chronic brain sagging, with reversibility of acquired CM and normalization of CSF circulation flow at the foramen magnum level.2

We agree with the suggestions of the authors. In fact, although the treatment for congenital CM involves posterior fossa decompression, the mainstay treatment for SIH involves an EBP and, consequently, it should be the main treatment of acquired CM from spontaneous or secondary CSF leak.

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References

Disclosures
The authors report no conflict of interest.

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Response
We thank Drs. Ferrante and Pontrelli for expressing their interest in our case report of secondary CM due to unusual intracranial pressure abnormalities in GSD. The treatment of idiopathic CM has already been established, i.e., foramen magnum decompression. In our case, we provided evidence of dynamic improvements in tonsillar herniation and syringomyelia after the arrest of the CSF leak without foramen magnum decompression. Awareness of this disease needs to be promoted to differentiate secondary CM from idiopathic CM.

Neuroimaging findings differ between patients with secondary CM due to a CSF leak and those with idiopathic CM. As shown in the T2-weighted brain MR image of our case (Fig. 1), we identified effacement of the third ventricle (arrow), diencephalic-mesencephalic deformities (asterisk), flattening of the pons (double arrowheads), and tonsillar herniation (double asterisks), showing this patient had secondary CM due to a CSF leak. Because the treatment of secondary CM differs from that of idiopathic CM, the clinical manifestations and neuroimaging findings of patients with CM need to be carefully assessed.

Although an EBP is the first-line treatment for secondary CM, surgical dural repair is indicated for patients with a CSF leak for whom the spinal level of the dual abnormality has been identified: a ventral dural tear or radicular meningeal diverticulum.

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We would like to express some criticisms on the rationale behind the choice of the surgical procedure. Vascular problems sustained in the perinatal period lead to ischemic lesions in one hemisphere or quadrant (anterior or posterior). This infantile hemiplegia seizure syndrome is a well-recognized cause of childhood epilepsy. The patients in the Scavarda and colleague series had ischemic events of the middle cerebral artery (MCA), leading to

FIG. 1. Sagittal T2-weighted brain MR image of the present case.