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Case study

Chiari-like displacement due to spontaneous intracranial hypotension in an adolescent: Successful treatment by epidural blood patch

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ABSTRACT

Background: Spontaneous intracranial hypotension is a rarely diagnosed cause of headache, especially in children and adolescents. It is due to cerebrospinal fluid (CSF) leakage via spinal fistulae occurring without major trauma.
Case presentation: An adolescent patient presented with a 3-month history of strictly postural headache. Cranial magnetic resonance imaging (MRI) showed pronounced Chiari-like prolapse of the cerebellar tonsils, narrow ventricles and enlarged cerebral veins. On spinal MRI, myelographic sequences revealed a large collection of CSF around the first sacral roots. CT myelography proved extensive spinal CSF leakage. Hence, we applied epidural patches at multiple levels. Afterwards, symptoms and radiologic findings, including Chiari-like displacement, completely resolved.
Conclusion: A Chiari-like descent of the cerebellar tonsils alone does not secure the diagnosis of a Chiari I malformation. Especially if other findings indicate spinal CSF leakage, a systematic work-up should be initiated. In most cases, interventional techniques seal the leak successfully, resulting in a favorable outcome.

1. Introduction

Spontaneous intracranial hypotension (SIH) is a rarely diagnosed cause of headache.[1,2] It is due to spinal cerebrospinal fluid (CSF) leaks which occur without a major trauma. A predisposing dural fragility may thus be suspected, and several studies have indicated an association with connective tissue disorders.[1,3] Abnormalities in fibrillin-1 synthesis and

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deposition have been linked to the pathogenesis of SIH, but a relevant involvement of other extracellular matrix components seems likely.[4] While SIH is increasingly recognized in the adult population, only single case reports and one larger case series[5] have been published on children and adolescents.

A distinguishing feature is that headache is postural in the vast majority of cases.[1,2] On cranial MRI, sagging of the brain and enhancement of the pachymeninges are the most frequent findings in the pediatric age group.[5] CSF leaks or a predisposition to these can be revealed by CT or MR myelography.[1,2] In equivocal cases, lumbar puncture can be helpful if a low opening pressure is found.[6] Conservative management is initially recommended, but only seldom sufficient.[1,2] Interventional techniques, such as epidural blood patching[7] or percutaneous injections of fibrin glue,[8] are in contrast remarkably effective. More invasive surgical approaches remain reserved for refractory cases.[2]

We report on an adolescent male with strictly postural headache and a Chiari-like prolapse of the cerebellar tonsils due to extensive spinal CSF leakage. Following epidural patches at multiple levels, symptoms resolved and radiologic findings were reversible.

2. Case study

A 15-year-old boy was admitted to our hospital after a 3-month history of postural headache. Pain started about one minute after changing from horizontal to vertical position and completely resolved again in horizontal position. Headaches were felt predominantly in the frontal and occipital area. Pain intensity was usually 5–6 of 10 on visual analogue scale and the boy hardly attended school for several weeks. Over-the-counter analgetics (paracetamol, ibuprofen), physical therapy and oral magnesium supplementation yielded no significant relief. Apart from mild morning nausea, no concomitant symptoms were reported. In retrospect, the family recalled that two days before headache onset, the patient had complained of a sudden lower back pain during a football training session. The boy's past medical history was otherwise unremarkable, as well as his family history.

On examination, no neurological abnormalities were found. The patient's stature was tall and slim, but without characteristic signs of Marfan syndrome. Consultation of an ophthalmologist, otolaryngologist and orthopaedic surgeon provided unremarkable findings. Laboratory tests were within normal limits except for a slightly elevated ANA titer.

Initial cranial magnetic resonance imaging (MRI) was unremarkable. However, six weeks later a follow-up study showed cerebellar tonsils prolapsing 11 mm into the foramen magnum, narrow ventricles and enlarged cerebral veins (Fig. 1A). On spinal MRI, a narrow spinal canal was noticed (Fig. 1B). Three-dimensional constructive interference in steady state (3D-CISS) sequence showed a large CSF collection around the roots of the first sacral nerves (Fig. 1C).

Anesthesia was consulted for sedation. For myelography, a lumbar puncture was performed and 15 mL of Solutrast™ 250 M (Bracco Imaging Deutschland GmbH, Konstanz, Germany) were infused intrathecally. Next, the patient was laid down and turned from prone to supine and then to side position. After 30 min, CT was performed in prone position. Extensive CSF leakage was confirmed at lumbar nerve root 4

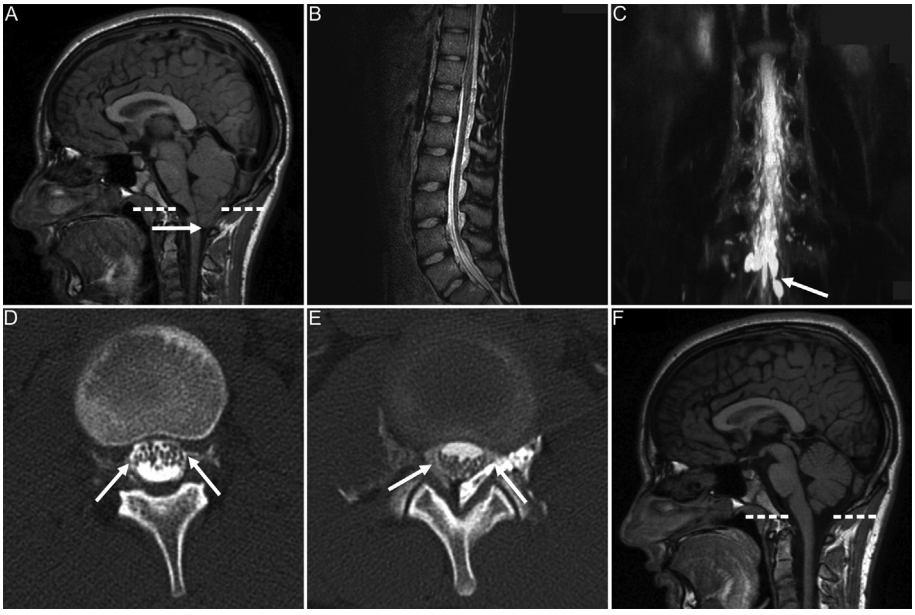

Fig. 1 – (A) Sagittal T1-weighted MRI 6 months after the onset of headaches, showing 11 mm prolapse of the cerebellar tonsils (arrow) into the foramen magnum (dashed line), narrow ventricles and enlarged cerebral veins. (B) Sagittal T2-weighted MRI, showing a narrow spinal canal. (C) Coronal 3D-CISS sequence, showing a CSF collection around the first sacral nerve roots, especially on the left side (arrow). (D) Axial CT myelography, showing extensive CSF leakage bilaterally (arrows). (E) Re-CT after application of epidural blood patch, showing that the patch (arrows) covers the whole circumference of the dural sac. (F) Sagittal T1-weighted MRI 2 months after the intervention, showing that findings normalized. Note in particular re-ascent of the cerebellar tonsils above the foramen magnum (dashed line).