Chiari malformation type I presenting with cognitive impairment and parkinsonism: temporary reversal following epidural blood patch

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Objectives

Chiari malformation type I (CMI), characterized by caudal herniation of the cerebellar tonsils through the foramen magnum, may present with headache, gait disturbances, brainstem dysfunction, and, more rarely, cognitive deficits. In adults, diagnosis is frequently delayed due to symptomatic overlap with conditions such as spontaneous intracranial hypotension (SIH).

We describe a case of CMI in an adult initially misdiagnosed as SIH, featuring progressive cognitive and neurological symptoms with transient improvement following epidural blood patches (EBPs).

Materials

A 51-year-old man without neurological history presented with orthostatic headaches, psychomotor slowing, dysarthria, ataxic gait, dysphagia, sialorrhea, and parkinsonian features. during years he underwent extensive clinical, radiological, and neuropsychological evaluation, including four EBP procedures.

Methods

Investigations included brain and spinal MRI (with and without contrast), MRI myelography, cine phase-contrast MRI, CT myelography, and CSF analysis. Neurophysiological studies comprised EEG, evoked potentials, EMG, and PET-CT. Cognitive function was assessed before and after EBP interventions

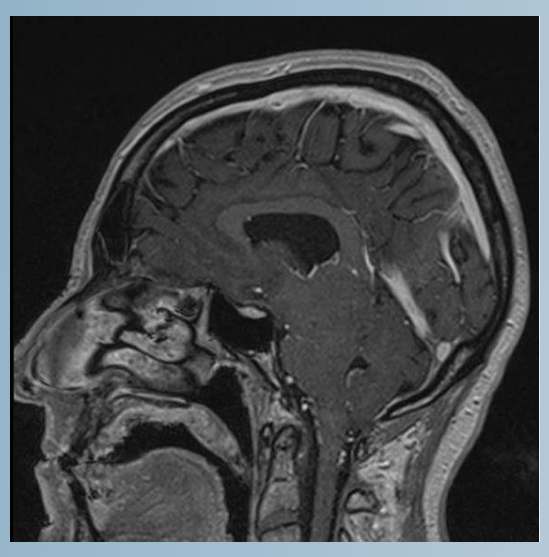

Fig. 1: Sagittal T1 gad+ sequence MRI showing a small posterior fossa results in crowding of hindbrain structures, producing downward herniation of cerebellar tonsil of 7 mm

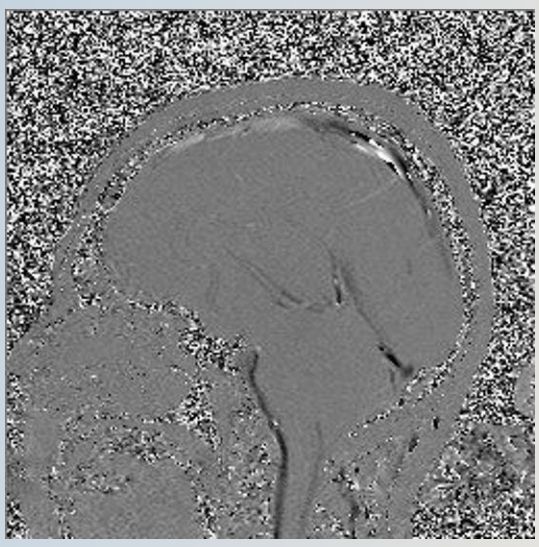

Fig. 2: Phase-contrast Cine-MRI study shows a disorganized CSF pulsation, with reduced flow through the posterior part of foramen magnum.

Results

MRI revealed 7 mm cerebellar tonsillar descent without contrast enhancement. CSF pressure measurement showed a normal pressure (14 cm H_2O). Cine MRI demonstrated CSF flow obstruction at the foramen magnum. EBP treatment led to transient but reproducible improvement in headaches, motor function, and cognition, including resolution of behavioral disinhibition. Neuropsychological testing identified multidomain deficits (executive function, memory, visuospatial skills, apraxia, and emotional regulation), which temporarily improved post-EBP. However, symptoms consistently recurred within 10 days of each EBP. The patient declined posterior fossa decompression and continued to experience symptom fluctuations at long-term follow-up.

Video 1: Evolution of the patient's clinical picture throughout the follow-up. Evident, although transient, clinical improvement of parkinsonian signs, gait and dysarthria occurred after epidural blood patch (EBP) treatment. Video-assessment 1: before EBP; Video-assessment 2: one day after EBP; Video-assessment 3: one week after EBP; Video-assessment 4: ten years after EBP (homemade).

Conclusion

CMI should be considered in adult patients presenting with atypical neurological or cognitive symptoms, even in the absence of classic headache patterns. MRI and cine-MRI are critical for evaluating CSF flow dynamics. The reproducible yet transient benefit from EBP may offer diagnostic insight and help identify candidates for posterior fossa decompression. Further research is warranted to explore the predictive value of EBP response in surgical decision-making for CMI.

Riferimenti

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