
Spontaneous Resolution of a Chiari I Malformation: MR Demonstration

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Summary: The MR findings in a case of a Chiari type I malformation that resolved spontaneously over a 4-year period are presented. Differential growth of the skull and spine might have accounted for the resolution of this anomaly.

Index terms: Chiari malformations; Spine, abnormalities and anomalies

It is generally accepted that downward displacement of the cerebellar tonsils by less than 2 mm can be considered normal (1, 2). Tonsillar herniation between 3 and 5 mm is indeterminate, and herniation of more than 5 mm is compatible with a Chiari type I malformation. A Chiari I malformation more often involves both tonsils, but a unilateral tonsillar ectopia occasionally is encountered (3). It is uncertain whether the latter is a true anomaly or just an anatomic curiosity (3). A Chiari I malformation is seen most often in female subjects, and the degree of tonsillar herniation correlates directly with the severity of symptoms (3). We present a case of a girl with a Chiari I malformation that underwent spontaneous resolution.

Case Report

A 9-year-old girl underwent magnetic resonance (MR) imaging of the brain for partial complex seizures that had been present for the last 5 years. Although no focal lesions were found, the cerebellar tonsils were displaced inferiorly and were triangular, a finding compatible with a Chiari type I malformation (Figs 1A and B). No prior lumbar puncture had been performed, and the remainder of the brain showed no edema or signs of increased intracranial pressure. Four years later, the patient underwent a second brain MR examination, which revealed a focal lesion in the right hippocampus. The cerebellar tonsils were still somewhat pointed, but only minimally displaced inferiorly (Fig 2A and B).

Discussion

Our case suggests the possibility of “out-growing” a Chiari I malformation, a phenomenon we were not aware of before this patient. The diagnosis of primary tonsillar ectopia can be made when displacement below the foramen magnum exceeds 6 mm in the first decade of life (2). More than 5 mm of downward displacement during the second and third decades of life should also be considered abnormal (2). In addition to inferior displacement, “beaking” of the tonsils strongly supports the diagnosis of Chiari I malformation. In our patient, the first study was definitively compatible with a Chiari I malformation (Fig 1A and B). When the inferior displacement of the tonsils is between 5 and 10 mm, most patients are asymptomatic (3). Conversely, patients with more than 10 mm of tonsillar herniation may have signs of decreased sensation, pain, weakness in the extremities, nystagmus, cranial-nerve palsies, and ataxia, among other symptoms (3). As many as 40% of patients with Chiari I malformations harbor a syrinx; in these cases, symptoms are generally referable to the syrinx rather than to the tonsillar ectopia (3). Cerebellar tonsils herniated by 2 to 3 mm can be considered borderline for normality (3). Transient tonsillar herniation can occur after cerebrospinal fluid removal procedures (such as lumbar puncture or shunting) or chronic cerebrospinal fluid leakage, which in itself may be cryptic. These patients have postural headaches because of traction on the pain-sensitive dura. Although benign intracranial hypotension can produce radiographic findings similar to Chiari I malformation, other MR features such as effacement of basilar cisterns, “slitlike” ventricles, and dural enhancement are

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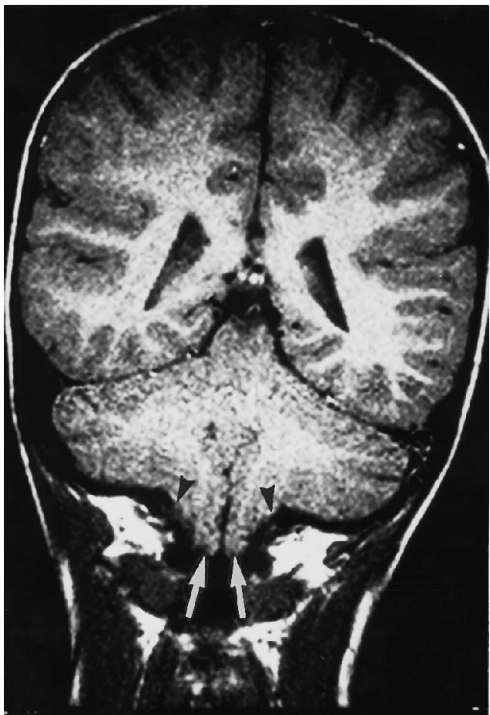
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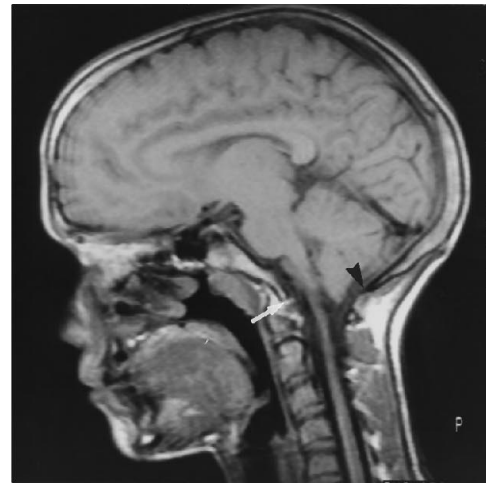
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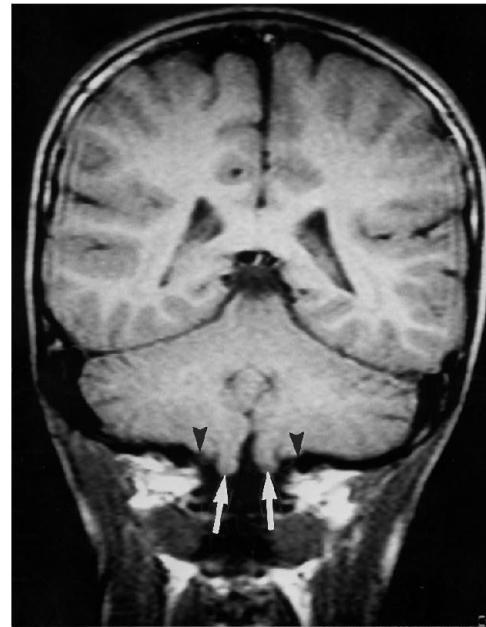
B

Fig 1. A, Initial midsagittal T1-weighted MR image (600/15/2 [repetition time/echo time/excitations]) shows marked downward displacement of the cerebellar tonsils (*large arrow*) with respect to the anterior (*small arrow*) and posterior (*arrowhead*) borders of the foramen magnum. The tonsils are beaked.

B, Coronal T1-weighted MR image (600/15/2) shows bilateral, symmetric cerebellar tonsillar herniation (*arrows*). *Arrowheads* indicate borders of foramen magnum.



A



B

Fig 2. A, Slightly tilted midsagittal T1-weighted MR image (600/15/2) 4 years after initial study. Note the position (well within the limits of normal) of the left cerebellar tonsil with respect to the anterior (*arrow*) and posterior (*arrowhead*) borders of the foramen magnum.

B, Coronal T1-weighted image (600/15/2) shows the normal position of both cerebellar tonsils (*arrows*). *Arrowheads* indicate borders of foramen magnum.

almost always present (4, 5). Our patient had no history of prior cerebrospinal fluid procedures or of postural headaches, thus making the diagnosis of benign intracranial hypotension unlikely. Intracranial masses and cerebral edema are relatively common causes of secondary cerebellar tonsillar herniation. It is conceivable that cerebral edema resulting from seizure activity can produce downward displacement of the cerebellar tonsils. In our patient, the initial MR study was performed at a time when the seizures were medically controlled, and as assessed by imaging, the remainder of the brain was normal. Atrophy of the cerebellar tonsils leading to their elevation and return to a normal position was also unlikely in our case, because the rest of the cerebellum showed no signs of volume loss.

The present case suggests that, occasionally, a Chiari I malformation may spontaneously resolve, especially during periods of rapid corporeal growth. Differential growth between the bone structures (skull and spine) and the central nervous system might have been responsi-

ble for the findings described here. Mikulis et al (2) suggested that in newborns, the volume of the posterior fossa is relatively small; therefore, the cerebellar tonsils are "conveniently" low in position. As corporeal growth accelerates, the skull expands to fully accommodate the cerebellum, and tonsils become "normal" in position. Neuronal cell loss (atrophy) might account for the high position of the cerebellar tonsils seen in older people (2).

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