LETTER TO THE EDITOR

Sagging and swelling of the midbrain suggest spontaneous intracranial hypotension rather than a malformation

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Sir, In a recent Brain review article, Barkovich et al. (2009) proposed a new classification for midbrain–hindbrain malformations that takes into account a vast amount of neuroimaging data, as well as results of research in developmental biology and molecular genetics. Based on these findings, the authors also envisage the existence of some malformation that is still not completely understood and for which the pathogenetic mechanism is inferred from biological and genetic studies in animal models. This is the case in a malformation secondary to early anteroposterior transformation at the diencephalic–mesencephalic junction, resulting in shortening and thickening of the midbrain with midline mesencephalic cleft (labelled in the article: Group 1, A.1.c. Gain of diencephalon and loss of midbrain). This malformation is shown in Fig. 4.

The magnetic resonance images presented in the figure, however, are identical to those observed in a subgroup of the most severe cases of spontaneous intracranial hypotension, reported in the literature and examined with diffusion studies by our group (Savoiardo et al., 2007).

Spontaneous intracranial hypotension, usually due to leakage of cerebrospinal fluid (CSF) along a spinal root, is clinically characterized by orthostatic headache and, on magnetic resonance imaging, thickening of the dura with post-contrast enhancement, subdural collections, sagging of the brain and dilatation of the venous compartment (with dural sinuses and pituitary gland dilatation in the head and engorgement of the spinal epidural plexus in the spinal canal) (Chiapparini et al., 2002; Mokri, 2005; Schievink, 2006). These features are explained by the Monro–Kellie doctrine: in a ‘closed’ compartment, such as the cranial cavity and the space of the spinal canal, where nervous tissue, blood and CSF are contained, the loss of one component (e.g. CSF) must be compensated by an increase of the others to maintain unaltered the total volume and pressure of the content (Mokri, 2001). The most easily available component is venous blood because veins expand more easily than arteries (Fishman and Dillon, 1993); more rarely swelling of the brain may contribute to the compensation (Savoiardo et al., 2007).

The decreased amount of CSF in the cranial and spinal subarachnoid spaces also causes the sagging of the brain, manifested by different degrees of tonsillar herniation and, more frequently, by downward displacement of the diencephalon and mesencephalon at the tentorial incisura. The line of buoyancy is lowered. In clinically severe cases, when the patients become obtunded or lapse into coma, lowering the position of the head or lumbar intrathecal injection of normal saline solution (the ‘uncorking procedure’ of Binder et al., 2002) may immediately reverse the situation, thus proving that the loss of volume of CSF is the ‘primum movens’.

When the cases of spontaneous intracranial hypotension are typical in their clinical and magnetic resonance presentation, there are no problems of interpretation. However, when the orthostatic headache or the dural thickening with post-contrast enhancement, or both, are absent, the images become puzzling and may be misinterpreted. In our series of >150 cases of spontaneous intracranial hypotension, we observed a severe downward displacement of the diencephalon and mesencephalon without dural thickening or effusions in 15. The caudal displacement obviously also involves the third ventricle, which is very thin and appears as a cleft in an unusually low position (see Figs 2, 3, 5 and electronic Fig. 1 in Savoiardo et al., 2007). In coronal sections, the red nuclei that normally are just above the level of the free margins of the tentorium are several millimetres below the tentorial notch. We interpreted the enlargement of the midbrain as a deformity resulting from diencephalic–mesencephalic caudal displacement associated with vasogenic oedema due to venous stagnation. In coronal sections, even the lowered pons appears...
deformed, with a superior concavity on the midline. The corpus callosum may descend, and the splenium may be partially hooked below the confluence of the vein of Galen into the straight sinus. Decrease of the angle formed by vein of Galen and straight sinus causes a functional stenosis that may hamper the deep venous drainage, causing oedema from venous stagnation mainly in the diencephalon and mesencephalon (Savoiardo et al., 2007).

The evidence that in these cases the deformity of the diencephalic–mesencephalic junction is not due to a malformation but the result of spontaneous intracranial hypotension is given by its reversibility that, however, may be slow on the magnetic resonance controls (and slower than the clinical recovery); this is probably due to the scarce elasticity of the brain and the long period of time that the deformity had been present. Acute cases may reverse more rapidly. We would also like to point out that a few of our cases with long history of the disease were referred to us as having a midbrain malformation, sometimes associated with Chiari malformation, or a diencephalic–mesencephalic glioma, before being proved to have spontaneous intracranial hypotension.

So far we have not seen a case with the kind of deformity illustrated in Fig. 4 of the Barkovich et al. (2009) paper that could be attributed to a malformation. Probably such cases exist as envisaged by these authors. We would only like to point out that a treatable condition, i.e. spontaneous intracranial hypotension, might be responsible for similar or identical findings.

References


