Spontaneous intracranial hypotension with deep brain swelling

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Spontaneous intracranial hypotension (SIH) is caused by leakage of CSF, and characterized on MRI by brain sagging, dilatation of veins and dural sinuses, subdural fluid collections and post-contrast enhancement of the thickened dura. A few cases may present a very severe brain sagging through the tentorial notch and swelling of the diencephalic-mesencephalic structures, with absent or scarce subdural collections and post-contrast enhancement. These patients may have surprisingly few neurological signs or may become drowsy and even lapse into coma due to central herniation. We retrospectively examined the diffusion studies obtained in five patients with these MRI findings, in seven patients with SIH without brain swellings and in ten controls.

Mean diffusivity was increased in SIH patients with brain swelling in areas draining into the deep venous system, collected by the vein of Galen (vG) and straight sinus (SS). In the hypothesis that central herniation might be responsible for venous stagnation because of impaired flow of the vG into the SS, the vG/SS angle was measured. The angle formed by the vG entering the SS was not altered in patients without brain swelling (group E, 67.8° ± 10.3°, mean ± SD, range 49°–80°) when compared to controls (group C, 73.3° ± 12.3°, mean ± SD, range 56°–95°). It was, however, grossly decreased in patients with brain swelling (group D, 40.7° ± 12.8°, mean ± SD, range 22°–61°), P < 0.001 for comparison with groups E and C. As suggested by previous studies, downward stretching of the vG and narrowing of the vG/SS angle may cause a functional stenosis at the vG–SS junction. We suggest that in the application of the Monro–Kellie doctrine to SIH, the brain volume should not be considered as always invariable.

Keywords: spontaneous intracranial hypotension; diffusion imaging; vasogenic oedema; venous stagnation; Monro–Kellie doctrine

Abbreviations: ADC = apparent diffusion coefficient; DWI = diffusion-weighted imaging; ROI = region of interest; SIH = spontaneous intracranial hypotension; SS = straight sinus; vG = vein of Galen


Introduction

Leakage of CSF is always considered as the pathogenetic factor of spontaneous intracranial hypotension (SIH), even in those cases in which the site of leakage remains occult (Mokri, 2003, 2005; Schievink, 2006). Loss of CSF volume explains the usual complaint of orthostatic headache, which is relieved by lying down, and the characteristic features seen on MRI, i.e. thickening of the dura mater enhancing after administration of contrast medium, subdural fluid collections, sagging of the brain, engorgement of the venous structures with dilatation of the intracranial dural sinuses and of the spinal peridural plexuses and enlargement of the pituitary gland (Rando and Fishman, 1992; Hochman et al., 1992; Renowden et al., 1995; Dillon and Fishman, 1998; Shimazu et al., 1998; Rabin et al., 1998; Chiapparini et al., 2002; Schievink, 2006). These features are explained by the Monro–Kellie doctrine: in a closed compartment, such as the intracranial and spinal canal spaces which contain nervous tissue, blood and CSF, the loss of volume of one component, CSF in our case, must be compensated by a corresponding increase in the volume of the other ones. Since the CNS is generally considered to be invariable, compensation should only occur through increase of blood volume, essentially of venous blood, because veins are more easily expandable than arteries (Fishman and Dillon, 1993; Mokri, 2001; Savoiardo et al., 2006).
A few reports described SIH patients who presented an ominous situation, manifested on MRI by very severe transtentorial brain sagging and by a swollen aspect of the upper brainstem, sometimes leading to parkinsonian features (Pakiam et al., 1999), frontotemporal dementia (Hong et al., 2002) or obtundation and coma probably due to compression of the diencephalic structures on the dorsum sellae (Pleasure et al., 1998; Beck et al., 1998; Binder et al., 2002; Evans and Mokri, 2002; Whiteley et al., 2003; Kashmere et al., 2004; Sayer et al., 2006). Although most of these cases had large subdural haematomas, a disproportion between subtle subdural fluid collections and a very severe brain sagging is occasionally observed (Pleasure et al., 1998; Beck et al., 1998; Pakiam et al., 1999; Hong et al., 2002). In these patients, diagnosis of SIH is confirmed by demonstration of the point of leakage, or by response to the epidural blood patch.

In a review of our series of 86 cases of SIH we identified 11 patients with these peculiar features, subsequently referred to as brain swelling, for which we suggest a particular pathogenetic mechanism. We define brain swelling as an obvious enlargement of diencephalo–mesencephalic structures with obliteration of the surrounding cisterns. In a few of them, large subdural haematomas coexisted with severe sagging, a swollen aspect of the upper brainstem and diencephalon and diencephalic compression on the dorsum sellae with flattening of the pons against the clivus. In most of these patients, however, subdural fluid collections were essentially absent and post-contrast enhancement of the thickened dura was scarce. We hypothesized that the swollen aspect of the diencephalic and mesencephalic structures might be due to vasogenic oedema, caused by venous stagnation resulting from impaired venous drainage.

Mean diffusivity of water is known to be a sensitive marker of vasogenic oedema, accumulation of extra-cellular fluid determining an increase in the diffusion coefficient (Kuroiwa et al., 1999). Therefore, in order to evaluate our hypothesis, mean diffusivity values from patients with SIH with brain swelling were compared with those from SIH patients without brain swelling and from controls. Additionally, in the hypothesis that narrowing of the angle formed by the vein of Galen (vG) and the straight sinus (SS) might be the cause of a functional stenosis impairing venous drainage, this angle was measured in patients with and without brain swelling, and in control subjects.

**Subjects and methods**

All the patients included in this report had been admitted to our Institute between 1993 and 2006 and were diagnosed as having SIH. Due to the retrospective nature of the present study, diffusion-weighted imaging (DWI) was not available for all patients. It was available only for five patients with brain swelling.

**Clinical data**

Diagnosis of SIH fulfilled the diagnostic criteria of the Headache Classification Subcommittee of the International Headache Society (2004), although blood patch or other invasive treatments were not always performed.

Clinical data, initial diagnosis and outcome for the five patients with brain swelling who had diffusion imaging are summarized in Table 2. Case histories of these patients are provided as Supplementary material.

**Investigations**

For all patients, brain MRI included T1- and T2-weighted sequences on the three orthogonal planes, fluid attenuated inversion recovery (FLAIR) and post-contrast T1-weighted images. Spinal MRI included pre- and post-contrast sagittal T1-weighted sections, sagittal T2-weighted sections, sequences with myelographic effect and coronal and axial sections as deemed necessary.

Images from all patients with brain swelling were re-evaluated retrospectively. Controls were randomly chosen on the basis of age and availability of DWI and sagittal sections among healthy subjects that had been previously imaged. All participants were scanned after providing written informed consent. The study complied with institutional guidelines and regulations.

Diffusivity values were measured in three groups: five patients with brain swelling without subdural haematomas (group A, age 54.5 ± 7.4 years), seven patients without brain swelling (group B, age 38.5 ± 14.5 years) and 10 volunteers (group C, age 44.6 ± 12.6) for whom CNS pathology had been excluded (Table 1).

Subjects in these groups were scanned with a 1.5 Tesla unit using a diffusion-weighted single-shot twice-refocused spin-echo echo-planar sequence, at b = 1000 s/mm², with a 128 × 128 matrix, FoV 230 × 230 mm, thickness 3.5 mm, interslice gap 0.3 mm, TR = 3800 ms, TE = 72 ms, NEX = 6; bulk mean diffusivity was computed from the average of three orthogonal gradient directions.

Mean diffusivity (i.e. the apparent diffusion coefficient, ADC) was measured by means of planar regions of interest (ROI) manually drawn, jointly, by two senior neuroradiologists, blinded to patient identity and morphological imaging. These were positioned in the central pons, in the central midbrain, bilaterally.

<table>
<thead>
<tr>
<th>Table 1 Synopsis of the patient groups</th>
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<tr>
<td>Diffusivity measurements</td>
</tr>
<tr>
<td>A 5 pts. with BS, without subdural haematomas</td>
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<tr>
<td>B 7 pts. without BS</td>
</tr>
<tr>
<td>C 10 normal controls</td>
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</table>

vG/SS angle = angle between vein of Galen and straight sinus; BS = brain swelling.
in tegmental areas of the midbrain, in the thalami, in the lentiform nuclei and in deep frontal and parietal white matter (Fig. 1). The area of these ROI ranged between 71 and 84 mm², and sides were considered separately in the statistical analysis.

The angle between the vG and the SS was measured on sagittal midline T2- (when available) or T1-weighted sections. Because of the curvature of the vG, the direction of its final 8–10 mm was considered. The angle measurements were performed jointly by two senior neuroradiologists, blinded to patient identity. The angle was measured for the whole group of 11 patients with brain swelling (group D), which included the five patients of group A, three patients with brain swelling, without subdural collections, who did not have diffusion studies and three patients with brain swelling and subdural haematomas. The age of this group was 51.7 ± 7.4 years (mean ± SD). The angle was also measured in 12 consecutive patients without brain swelling (group E, age 44.3 ± 8.4 years). Patients in group E had the usual MRI correlates of SIH, with post-contrast dural enhancement, less-severe brain sagging, and without the swollen aspect of the deep structures. The same controls chosen for DWI (group C) were considered (Table 1).

In order to exclude confounding effects of age, we tested the study hypotheses using ANCOVA, with three linear contrasts: healthy controls versus patients without brain swelling, healthy controls versus patients with brain swelling and patients with brain swelling versus patients without brain swelling. Tukey post hoc contrast tests were employed.

Results

MRI

In all eight patients with brain swelling without subdural haematomas the following striking features were present: (i) severe brain sagging with transtentorial descent of the diencephalon and third ventricle; (ii) minimal or absent post-contrast enhancement of the dura, and dural thickening or subdural fluid collections; (iii) the whole brain, particularly the diencephalon and the midbrain, and to a milder degree the deep periventricular white matter, appeared swollen; (iv) on T2-weighted images, the signal intensity in the central regions of the brain and in the midbrain was slightly increased (Figs 2 and 3, and electronic Fig. 1). An extremely thin third ventricle was visible in axial sections at levels much lower than usual, even caudal to the dorsum sellae.

The iter to the aqueduct (i.e. the entrance to the aqueduct from the third ventricle), whose lowest normal position is within 2 mm below the incisural line connecting the tuberculum sellae with the confluence of the vG into the SS (Reich et al., 1993; Pannullo et al., 1993), was displaced downwards by up to 22 mm (Table 2). The midbrain was deformed, elongated in its sagittal diameter and compressed by the temporal lobes, which presented some degree of uncal herniation (Fig. 3A and electronic Fig. 11). On the coronal sections, however, the midbrain appeared enlarged below the tentorial incisura as if it had been squeezed through the tentorial notch like toothpaste. The pons, caudally displaced, presented a curved shape, concave upwards (Fig. 3). The midbrain tectum was markedly displaced backwards, mainly in its upper part, with compression of the culmen of the upper vermis. The pons was flattened against the clivus. Descent of the tonsils through the foramen magnum was also present, but it was much less marked than the sagging of the brain through the tentorial incisura.

Additional imaging findings for these patients are summarized in Table 2.

Diffusion measurements

The ANCOVA did not reveal any effect of age on diffusivity. Values of mean diffusivity measured in the three groups are given in Table 3, in which the F-values and P-values of the group comparisons are also reported. The overall F-value of the ANCOVA was significant for all regions with the exception of the pons and of frontal white matter. When compared to controls, diffusivity in patients without brain swelling (group B) was not significantly higher in any region. When compared to controls, diffusivity in patients with brain swelling (group A) was found to be higher in the midbrain (P < 0.008), bilaterally in the basal ganglia (P = 0.01), bilaterally in the thalamus (P < 0.03) and bilaterally in periventricular white matter (P < 0.05). Additionally, diffusivity was significantly higher in group A when compared to group B in the central midbrain (P = 0.04). Patients with brain swelling always had the highest values of diffusivity, while patients without brain swelling were characterized by intermediate values.

Angle measurements

As expected, the ANCOVA did not reveal any effect of age. The angle formed by the vG entering the SS was not altered in patients without brain swelling (group E, 67.8° ± 10.3°, mean ± SD, range 49°–80°) when compared to controls (group C, 73.3° ± 12.3°, mean ± SD, range 56°–95°). It was grossly decreased in patients with brain swelling (group D, 40.7° ± 12.8°, mean ± SD, range 22°–61°), P < 0.001 for comparison with groups E and C. The scatter plot of angle measurements is shown in Figure 4. An example of angle measurement is shown in Figure 5.

Discussion

MRI findings similar to those observed in our patients with brain swelling have been occasionally reported (Pleasure et al., 1998; Beck et al., 1998; Pakiam et al., 1999; Hong et al., 2002; Binder et al., 2002; Evans and Mokri, 2002; Whiteley et al., 2003; Kashmere et al., 2004; Sayer et al., 2006), without specific mention of swelling. The deformity of the diencephalon and midbrain was only attributed to the severe brain sagging. Although subtle changes in signal intensity are usually visible, hyperintensities in T2-weighted images have almost never been mentioned, and, when
Table 2 Clinical and radiological data for the patients with SIH and BS in group A

<table>
<thead>
<tr>
<th>Case/ Age (years)</th>
<th>Sex</th>
<th>Age at onset</th>
<th>Clinical findings</th>
<th>Radiological findings</th>
<th>Previous diagnoses</th>
<th>Outcome</th>
</tr>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Usual SIH symptoms</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Other symptoms</td>
<td>Signs</td>
<td>Brain MRU</td>
<td>Brain MRI</td>
</tr>
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<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1/60F 57</td>
<td>F</td>
<td>57</td>
<td>OH, dizziness, hearing changes, neck pain</td>
<td>Weakness, difficulty in swallowing liquids</td>
<td>Frontal signs, ataxia, ideomotor slowing</td>
<td>–</td>
</tr>
<tr>
<td>2/56F 55</td>
<td>F</td>
<td>55</td>
<td>OH, dizziness, neck pain, nausea and vomiting</td>
<td>Weakness, drowsiness</td>
<td>Ideomotor slowing</td>
<td>–</td>
</tr>
<tr>
<td>3/60F 58</td>
<td>F</td>
<td>58</td>
<td>OH, diplopia, dizziness</td>
<td>Weakness, emotionally unstable</td>
<td>Frontal signs, ataxia, Romberg +, ideomotor slowing</td>
<td>–</td>
</tr>
</tbody>
</table>

Continued
recently described in the midbrain, they were not commented upon (Mokri et al., 2006). To our knowledge, there are no reports of diffusion studies in SIH patients.

Although most of the severe cases reported in the literature had impaired consciousness or even lapsed into coma, attributed to dysfunction of the midbrain reticular formation caused by severe sagging and deformity of the diencephalon and midbrain, some of our cases presented surprisingly few neurological signs. Therefore, there is a wide spectrum of neurological impairment with similar degrees of brain sagging. Hence, in these patients MRI does not appear to be a good predictor of clinical state. Rapidity of evolution of sagging is certainly a crucial factor. As occurs in patients with obstructive hydrocephalus, in whom signs of midbrain dysfunction are more frequently observed at the time of shunt malfunction than at first diagnosis, rapidity of onset of pressure changes seems to be more important than the degree of anatomical distortion (Cinalli et al., 1999). The relatively good neurological status of some SIH patients with severe brain sagging may be related to very slow evolution which might confer a remarkable tolerance to high degrees of central herniation. In our series, only one patient with brain swelling and without subdural haematoma (not studied with DWI) had fluctuating severe impairment of consciousness. During these fluctuations, this patient had three MRI studies (Figure 5A) which failed to demonstrate any recognizable difference during the periods of alertness and obtundation. Although our data do not enable to draw conclusions, it is possible that minimal changes in intracranial pressure and in brain sagging may greatly affect the functions of diencephalic and mesencephalic structures, perhaps altering the local perfusion.

We are convinced that the peculiar correlates of SIH with brain swelling, namely (i) swelling of the deep cerebral structures leading to mass effect and striking brain sagging at the level of the tentorial incisura, (ii) scarce evidence of dural venous engorgement and subdural fluid collection and (iii) slightly increased signal intensity on T2-weighted images in the same deep regions can only be explained by a mild degree of diffuse vasogenic oedema. The finding of elevated diffusivity and the morphological changes both provide support for this hypothesis. Indeed, mild diffuse vasogenic oedema, extending to the white matter of the cerebral hemispheres, may be a determining factor for the scarcity of dural thickening and subdural fluid collections observed in eight cases.

One must also consider the hypothesis that enlargement of intraparenchymal venules and capillaries due to venous stagnation could contribute to swelling of the deep brain structures and to increased ADC values. The relative entity of this effect compared to vasogenic oedema cannot be determined on the basis of our findings alone.

Two hypotheses for the causative factors of increased diffusivity have to be considered. One is that it is caused by engorgement of the venous system compensating for the
Fig. 1 Positioning of ROI on apparent diffusion coefficient (ADC) maps: central pons, central midbrain, bilateral tegmental midbrain, lentiform nuclei, thalami, bilateral frontal and parietal white matter.

Fig. 2 Case 5. T1- and T2-weighted midline sagittal sections (A and B) demonstrate severe brain sagging at the level of the tentorial incisura with deep brain swelling causing complete obliteration of the perisellar cisterns, downward displacement and closure of the interpeduncular cistern (arrowhead in A), diencephalic compression on the dorsum sellae, inferior and posterior displacement of midbrain tectum and posterior compression of the cerebellar culmen. The swollen diencephalon and midbrain (asterisk in B) appear slightly hyperintense in T2-weighted image. Position of the iter can only be inferred from the position of the superior colliculi. Note the low position of the internal cerebral vein (arrowhead) and the downward stretching of the vein of Galen (vG) around the splenium of the corpus callosum and compare these findings with the normal aspect in a control (C).
loss of CSF volume, accompanied by slowing of venous flow in the dural sinuses (Canhao et al., 2006) which may cause venous stagnation in the brain tissue and hence oedema. The second one is that oedema is caused by impaired drainage of the deep venous system, due to the sagging of the GC entering the SS. The two hypotheses are obviously not mutually exclusive and when functional oedema occurs, oedema develops probably because of increase of a more modest fluid accumulation already caused by venous engorgement compensating for the CSF loss.

Table 3 Diffusivity values (in mm² s⁻¹) measured in groups A (patients with SIH with BS), B (patients with SIH without BS) and C (controls), along with ANCOVA $F$-values and $P$-values from post hoc comparisons

<table>
<thead>
<tr>
<th>Group</th>
<th>Pons</th>
<th>Central</th>
<th>Tegmental</th>
<th>Tegmental</th>
<th>Basal</th>
<th>Basal</th>
<th>Thalamus, L</th>
<th>Thalamus, R</th>
<th>Frontal</th>
<th>Frontal</th>
<th>Parietal</th>
<th>Parietal</th>
</tr>
</thead>
<tbody>
<tr>
<td>A (with BS)</td>
<td>0.88 ± 0.09</td>
<td>0.89 ± 0.07</td>
<td>0.92 ± 0.06</td>
<td>0.92 ± 0.07</td>
<td>0.82 ± 0.01</td>
<td>0.81 ± 0.01</td>
<td>0.82 ± 0.09</td>
<td>0.83 ± 0.08</td>
<td>0.77 ± 0.04</td>
<td>0.79 ± 0.03</td>
<td>0.85 ± 0.06</td>
<td>0.85 ± 0.06</td>
</tr>
<tr>
<td>B (w/o BS)</td>
<td>0.81 ± 0.09</td>
<td>0.77 ± 0.08</td>
<td>0.86 ± 0.07</td>
<td>0.83 ± 0.07</td>
<td>0.79 ± 0.03</td>
<td>0.77 ± 0.05</td>
<td>0.79 ± 0.06</td>
<td>0.77 ± 0.06</td>
<td>0.73 ± 0.03</td>
<td>0.75 ± 0.04</td>
<td>0.81 ± 0.04</td>
<td>0.81 ± 0.06</td>
</tr>
<tr>
<td>C (controls)</td>
<td>0.81 ± 0.09</td>
<td>0.75 ± 0.06</td>
<td>0.78 ± 0.07</td>
<td>0.76 ± 0.07</td>
<td>0.71 ± 0.06</td>
<td>0.70 ± 0.06</td>
<td>0.71 ± 0.05</td>
<td>0.73 ± 0.03</td>
<td>0.77 ± 0.06</td>
<td>0.76 ± 0.05</td>
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A Versus C $F$ = 3.5, $P$ = 0.007* $F$ = 3.5, $P$ = 0.006* $F$ = 4, $P$ = 0.002* $F$ = 3.5, $P$ = 0.01* $F$ = 3.3, $P$ = 0.01* $F$ = 3.3, $P$ = 0.01* $F$ = 2.5, $P$ = 0.05 $F$ = 1.8, $P$ = 0.2 $F$ = 2.7, $P$ = 0.04* $F$ = 3.2, $P$ = 0.01* $F$ = 1.9, $P$ = 0.2 $F$ = 2.7, $P$ = 0.05 $F$ = 1.2, $P$ = 0.4 $F$ = 1.3, $P$ = 0.4 $F$ = 2.1, $P$ = 0.1 $F$ = 1.9, $P$ = 0.2 $F$ = 2.2, $P$ = 0.1 $F$ = 1.7, $P$ = 0.2 $F$ = 2.7, $P$ = 0.05 $F$ = 2.0, $P$ = 0.1 $F$ = 1.2, $P$ = 0.4 $F$ = 1.3, $P$ = 0.4 $F$ = 1.7, $P$ = 0.2 $F$ = 1.6, $P$ = 0.3

*A diffusivity values measured in ROIs of Fig. 1 (mean ± SD); L = left; R = right. Asterisks (*) indicate statistically significant differences.
loss. The first hypothesis is supported by the fact that patients with SIH without brain swelling (group B) were characterized by intermediate diffusivity values in all regions, which, however, were not significantly higher than those found in controls. The second one finds support in the distribution of increased ADC values, in the stretching of the vG, and in the reduction of the vG/SS angle, which, taken together, suggest that oedema is due to impairment of deep venous drainage. In fact, as represented in Fig. 6, the venous territories corresponding to the areas of increased ADC values (midbrain, thalami, basal ganglia and periventricular white matter) are drained by the basal veins of Rosenthal (which receive the inferior striate veins), by perimesencephalic veins, thalamic veins, subependymal veins of the lateral ventricles and internal cerebral veins that ultimately collect their blood into the vG and SS. Veins of the posterior fossa, in particular the precentral cerebellar vein and the superior vermian vein, are also tributaries of the vG. However, blood from veins draining the cerebellum find an alternative route, together with veins of the pons and lower brainstem, into the anterolateral (petrosal) and the inferior draining systems of the posterior fossa.

A distribution of swelling analogous to that seen in our cases was observed by Van Roost et al. (2003) as a rare and potentially fatal complication after uneventful brain surgery, probably related to acute intracranial hypotension induced by excessive suction drainage. It has been referred to as ‘pseudohypoxic brain swelling’. This complication, in which the brain sagging and the midbrain deformation are very similar to those seen in our SIH cases, may be completely reversed by the Trendelenburg position (Samadani et al., 2003; Kelley and Johnson, 2004; Komotar et al., 2005, 2006).

We found a support to our hypothesis of venous drainage impairment in the investigation published by Stolz et al. (2002), who analysed with ultrasound the

![Fig. 4 Scatterplot of angle measurements (in degrees) obtained in patients with SIH with BS, in patients with SIH with BS and with concomitant subdural haematomas (H), in patients with SIH without BS, and in controls.](image)

![Fig. 5 Measurements of vG/SS angle obtained in a patient for whom diffusion studies were not available, who presented with severe obtundation lapsing into coma when in sitting position, and awakening when lying down. The angle measured on T1-weighted post-contrast sagittal section (A) is 23°. Transient improvement after targeted blood patch was followed by complete clinical recovery after operation on right T11 root sleeve cyst. In spite of normal clinical conditions, the brainstem has not returned to a completely normal shape 18 months after operation (B). Although the angle is still rather narrow (39°), the interpeduncular cistern has largely reopened, the stretching of the vG has diminished, and the vG itself appears thicker.](image)
Intracranial venous haemodynamics in patients with compression of the mesencephalon due to tentorial brain herniation. Changes in flow velocities of the basal veins of Rosenthal, vG and SS were found and a functional stenosis at the transition between the vG and the SS was considered crucial in determining poor outcome (Stolz et al., 2002).

In accordance with these observations, we had noticed that severe transtentorial brain sagging causes stretching of the vG, which, together with narrowing of the vG/SS angle, is, in our opinion, the putative cause of functional stenosis. Due to the retrospective nature of our study, we were unable to obtain a quantitative indicator of venous narrowing. However, the downward stretched vG that accompanies a reduced vG/SS angle generally appears thinner than it is after resolution of SIH (Fig. 5). Impaired venous drainage may be the final result in all cases of downward central herniation, irrespective of the primary cause.

In a few SIH patients with brain swelling, either with or without subdural haematomas, the vG/SS angle increased considerably after evacuation of the haematoma or treatment of SIH, accompanied by regression of brain swelling (Fig. 5). The relative contribution of the factors potentially affecting intracranial pressure in SIH cannot be elucidated on the basis of our limited number of patients. DWI studies in patients with SIH with brain swelling and subdural haematomas should be performed to verify whether the ADC values are increased also in these patients. Although we still do not have sufficient data in this regard, clinical improvement after treatment seemed more rapid in patients with than in patients without subdural haematoma.

A diagnosis of SIH can be established with confidence, even in patients with brain swelling, when its characteristic features, that is subdural collections and post-contrast dural enhancement, are present. Conversely, diagnosis is difficult when these features are absent. It is important to note that SIH cases with swelling and severe brain sagging not associated with subdural haematomas or obvious post-contrast dural enhancement were often misdiagnosed as tumours or malformations of the posterior fossa structures. When properly diagnosed and treated, these patients showed prompt clinical improvement, although the sagging sometimes diminished very slowly and only partially, indicating that a long-lasting deformity of the brain may not be fully reversible due to loss of compliance of the nervous tissue (Fig. 5). Cases with brain swelling are fortunately rare, and the relatively high incidence (about 13%) in our series can be explained by the fact that our Institute is a referral centre.

The main limitation of our study is the relatively small number of SIH patients with brain swelling studied with DWI. Our interpretation of the pathogenic mechanism of brain swelling needs to be confirmed by further observations. In particular, the primary factor giving rise to this cascade of events in a small percentage of cases should be investigated. There might be predisposing anatomical variations, such as a high position of the apex of the tentorial notch where the vG enters the SS, or a particular shape and position of the splenium. In several cases including some of those reported in the literature (Pleasure et al., 1998; Beck et al., 1998; Whiteley et al., 2003; Kashmere et al., 2004; Sayer et al., 2006), the splenium appears stumpy, and, when displaced downwards, seems to become incarcerated below the tentorium (electronic Figure 2). Under both conditions, the lower level of buoyancy of the brain due to SIH could stretch the vG more severely than in ‘classical’ cases, leading to a reduction of the vG/SS angle.

On the basis of our data, we propose that, in the application of the Monro–Kellie doctrine to SIH, the volume of the brain should not be considered as always invariable. The total volume of brain, blood and CSF obviously remains unchanged. However, small increases in brain volume occurring in a few cases may have the effect of compensating for the loss of volume of CSF.

Conclusions

This study highlights that swelling of the deep brain structures can occur in SIH. The finding of increased diffusivity in patients with brain swelling suggests that vasogenic oedema is present. As the vG/SS angle was found to be reduced in these patients, venous stagnation caused by functional stenosis at the confluence of the vG into the SS is a plausible explanation.
Supplementary material
Supplementary material are available at Brain Online.

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