

CLINICAL CORRESPONDENCE

Delayed cerebral venous thrombosis in a patient with spontaneous intracranial hypotension

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Spontaneous intracranial hypotension (IH) may indicate cerebrospinal fluid (CSF) leakage, in the absence of a known dural puncture or tear. Intracranial vascular complications of spontaneous IH include subdural haematoma (SDH), subdural effusion (SDE) and, rarely, cerebral venous thrombosis (CVT) (1–3) and dural arteriovenous fistula (AVF) (3). In this report, we describe a patient with spontaneous IH who unusually developed all of these intracranial vascular complications. It is noteworthy that CVT occurred as late as 4 months after onset of IH.

Case report

A 36-year-old man suffered from a sudden pain in his neck and upper back after snorkel diving in June 2005. Holocranial headache with nausea and vomiting developed 2 days later. The headache was characterized by prominent postural variations, experienced on standing or sitting, whereas it resolved rapidly on recumbency. He visited a hospital for help and brain gadolinium-enhanced magnetic resonance (MR) T1-weighted images showed diffuse dural enhancement (Fig. 1A). The patient denied any previous dural puncture, surgical intervention or trauma. Spontaneous IH was diagnosed based on the characteristic clinical and imaging presentation. His headache resolved gradually after bed rest, hydration and use of analgesics. He could sustain an upright position for >2 h within 10 days.

Unfortunately, a severe headache with different features developed 1 month later. It was located in the right frontal region, persisted without postural variation and was aggravated by coughing or sneezing. Repeated brain MR imaging revealed a SDH on the right cerebral hemisphere and a SDE on

the left counterpart (Fig. 1B). The patient declined the suggestion of surgical intervention and his headache subsided with conservative management in the following 2 weeks.

In October, 4 months after onset of IH, he had sudden weakness and clonic jerking on the left side of his face and limbs without loss of consciousness. He was admitted to hospital, where brain MR imaging showed an acute intracerebral haemorrhage in the right posterior frontal cortex (Fig. 1C). Meanwhile, the previous abnormal meningeal enhancement, SDH and SDE had all resolved. He was treated with phenytoin for the partial motor seizures.

In November he developed recurrent convulsive seizures with loss of consciousness. Repeated brain computed tomography showed brain oedema around the resolved right frontal lobe haemorrhage. The seizures were successfully controlled by intravenous infusion of phenytoin. He recovered consciousness within 1 day and neurological examination revealed a mild left-side weakness. He was discharged in December and was treated with sodium valproate.

The patient was first seen by the authors in January 2006. His physical examination was normal and neurological examination demonstrated weakness and hyperactive tendon reflexes of his left limbs. To investigate the cause of intracerebral haemorrhage, cerebral digital-subtraction angiography was performed. An AVF between the meningeal branches of the superficial temporal arteries and dural veins was demonstrated in the parasagittal region of the right parietal lobe (Fig. 1D). Retrograde venous flow was also revealed at the periphery of a region lacking cortical venous drainage in the right frontoparietal cortex (Fig. 1E),

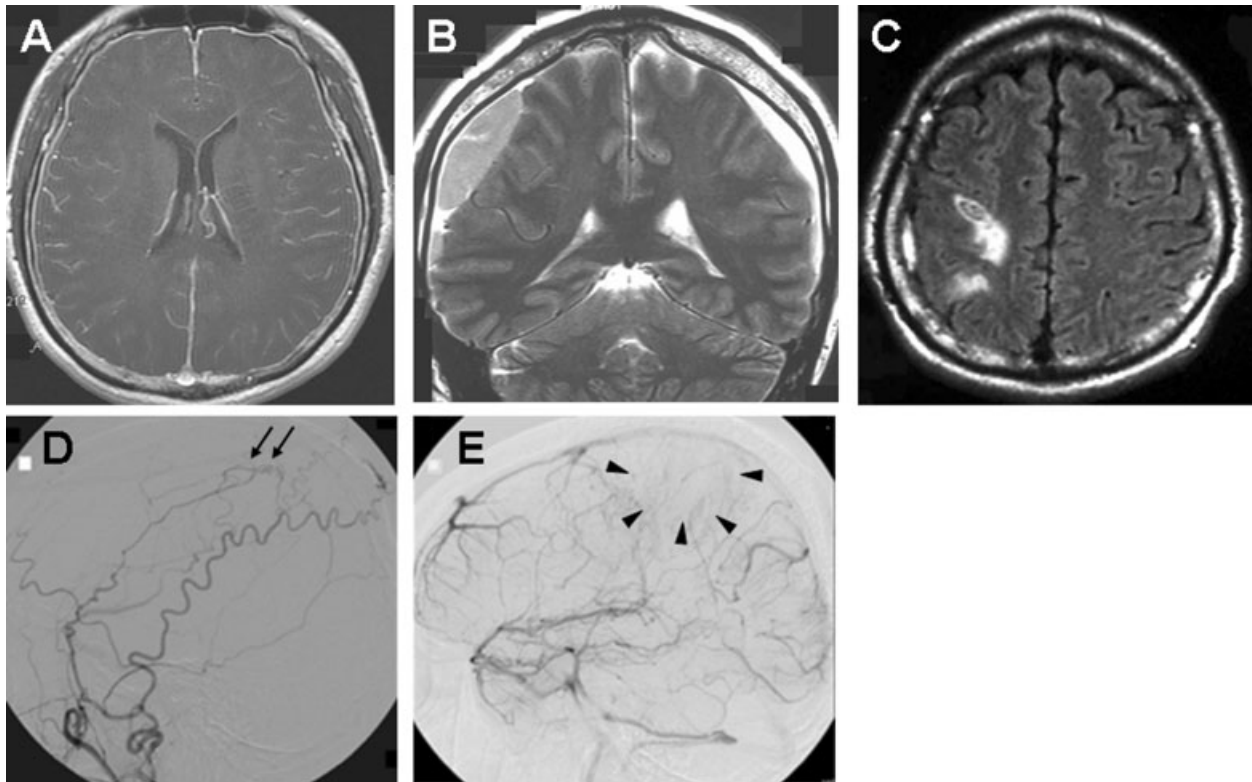


Figure 1 (A) Gadolinium-enhanced axial T1-weighted magnetic resonance (MR) image showing diffuse meningeal enhancement, a typical finding for intracranial hypotension (IH). (B) Repeated MR imaging study for a newly developed headache 1 month after onset of IH. Coronal T1-weighted MR image revealing subdural haematoma with a mass effect on the right side and a subdural effusion on the left side. (C) T1-weighted MR image 4 months after onset of IH showing an acute cerebral haemorrhage in the right posterior frontal cortex. (D) External carotid arteriogram showing a dural arteriovenous fistula (arrows) fed by the meningeal branches of the right superficial temporal artery. (E) Internal carotid arteriogram revealing a region lacking cortical venous filling at the right frontoparietal cortex (arrowheads).

suggesting occlusion of regional cortical veins. The main venous sinuses were patent for blood flow. Tests for autoantibodies (antinuclear antibody, anti-dsDNA antibody, extractable nuclear antigen antibodies, antiphospholipid antibodies and rheumatoid factor) and procoagulation disorders (complete blood counts, prothrombin time, activated partial thrombin time, protein C, protein S and antithrombin III) were all negative.

Discussion

IH can be confirmed by demonstrating an opening pressure of <60 mmH₂O on performing a lumbar puncture. Although CSF pressure was not measured in our patient, the clinical feature of orthostatic headache, in combination with diffuse meningeal enhancement on MR images and the absence of previous dural dissection and puncture, strongly supports the diagnosis of spontaneous IH.

In IH, reduced buoyant force for brain support and altered cerebral venous flow dynamics underlie the development of vascular complications. Depleted CSF volume leads to downward displacement of the brain and traction of dural veins. In addition, to compensate for the decreased intracranial volume, cerebral veins dilate prominently so as to fill with more blood (4). Both conditions may cause blood extravasation from the dural veins, resulting in SDH and SDE. On the other hand, increased blood volume and stagnant flow in engorged cerebral veins may predispose to CVT. Dural AVF is a well-established complication of CVT. Occlusion of a large cerebral vein may create a high resistance, forcing arterial flow to seek new pathways. Recanalization of thrombosed veins by sprouting of meningeal arteries into the venous walls may also promote development of dural AVF (5). Additionally, in the face of venous dilation in IH, progression of the vascular shunting could be facilitated.

To our knowledge, the patient described here is unique in the occurrence of SDH, SDE, CVT and dural AVF following IH. Interestingly, a latency of up to 4 months between the onset of IH and CVT was observed, in contrast to 1–4 weeks in previously reported cases (1–3). In the absence of a procoagulation state, delayed development of CVT in the patient suggests that abnormal venous flow dynamics may persist even when the clinical and imaging features of IH have apparently resolved. However, as this is a report of a single case, we cannot exclude the possibility that CVT and subdural AVF were unrelated to IH in this patient. Recognition of other similar cases may help to elucidate the pathogenic mechanisms and temporal sequence in the development of IH-related multiple intracranial vascular complications.

References

- 1 Berroir S, Grabli D, Héran F, Bakouche P, Bousser MG. Cerebral sinus venous thrombosis in two patients with spontaneous intracranial hypotension. *Cerebrovasc Dis* 2004; 17:9–12.
- 2 Sopelana D, Marcos A, Arroyo R, Gutierrez E, Cuenca R, Vazquez AV et al. May intracranial hypotension be a cause of venous sinus thrombosis? *Eur Neurol* 2004; 51:113–5.
- 3 Flemming KD, Link MJ. Spontaneous CSF leak complicated by venous thrombosis and dural arteriovenous fistula. *Cephalalgia* 2005; 25:751–3.
- 4 Roll JD, Larson TC III, Soriano MM. Cerebral angiographic findings of spontaneous intracranial hypotension. *Am J Neuroradiol* 2003; 24:707–8.
- 5 Mas JL, Meder JF, Meary E. Dural sinus thrombosis: long-term follow-up by magnetic resonance imaging. *Cerebrovasc Dis* 1992; 3:137–44.