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Short communication

Stroke after spontaneous intracranial hypotension: Not a single mechanism. Case report and review of literature

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ABSTRACT

Spontaneous intracranial hypotension (SIH) is a rare syndrome, typically manifests as orthostatic headache. Sometimes considered as benign illness, neurological complications are well described, in particular subdural hematoma and cerebral venous sinus thrombosis. Brain infarction as complication of SIH is rarely reported. The main mechanism supported in the literature is the stretching of arteries due to the sagging of the brain. We report a case of SIH followed with brain infarction, with a distinct presentation from previous literature, suggesting a different mechanism. A 35 year-old had severe orthostatic headache, responsible for prolonged bed rest. One month later, he had acute left hemiparesis secondary to stroke and right posterior cerebral artery occlusion. Stroke MRI showed arguments for intracranial hypotension (thickened meninges). He was successfully treated with intravenous rtPA thrombolysis. Headache were resolved after an epidural blood patch. A patent foramen ovale was detected. Clinical features of this description were compared with previous literature. This case suggest a different mechanism for cerebral infarction after intracranial hypotension. In case of prolonged lying down due to intracranial hypotension, the presence of patent foramen ovale could be a risk factor for embolic stroke.

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1. Introduction

Subdural hematoma, cerebral venous sinus thrombosis and associated venous infarction are the most common neurological complications of intracranial hypotension. Posterior reversible encephalopathy syndrome (PRES), reversible cerebral vasoconstriction syndrome (RCVS), and arterial cerebral infarctions are more rarely reported in literature. We report herein the case of a spontaneous intracranial hypotension (SIH) followed with acute ischemic stroke and intracranial artery occlusion. We discuss pathophysiological mechanisms in regard to previous literature.

2. Case report

A 35 year-old male without history of primary headache, reported daily headaches, beginning in the following days after an accidental fall. Headache were diffuse, increased by anteflexion of the head, and relieved by lying flat. They were severe and

particularly disabling. He spent a large part of the day lying down. Headache were firstly confused with sinus headache and no brain imagery was done. One month later, he reported sudden left hemiparesis and visual blurring. Brain magnetic resonance imaging (MRI) showed recent ischemic lesions in the right thalamus and right hippocampus, associated with right posterior cerebral artery (PCA) occlusion (Fig. 1). He was successfully treated with intravenous rtPA thrombolysis. A CT angiography of supra-aortic arteries was normal. A patent foramen ovale (3 mm) and atrial septal aneurysm (10 × 12 mm) were detected with transthoracic and transesophageal echocardiography. No deep vein thrombosis was detected with doppler ultrasound. A holter electrocardiogram was normal. Stroke MRI (without contrast) showed thickened meninges in Flair sequence (Fig. 1). A lumbar puncture was performed to rule out meningitis or meningeal hemorrhage, but cerebrospinal fluid (CSF) pressure was not measured. Headaches features didn't changed after lumbar puncture. The CSF cellularity was normal. Two months later, orthostatic headaches persisted, with a lower intensity. A new brain MRI showed diffuse pachymeningeal enhancement, cerebellar tonsillar descent, and right thalamic and hippocampal ischemic lesions (Fig. 1). Spine MRI

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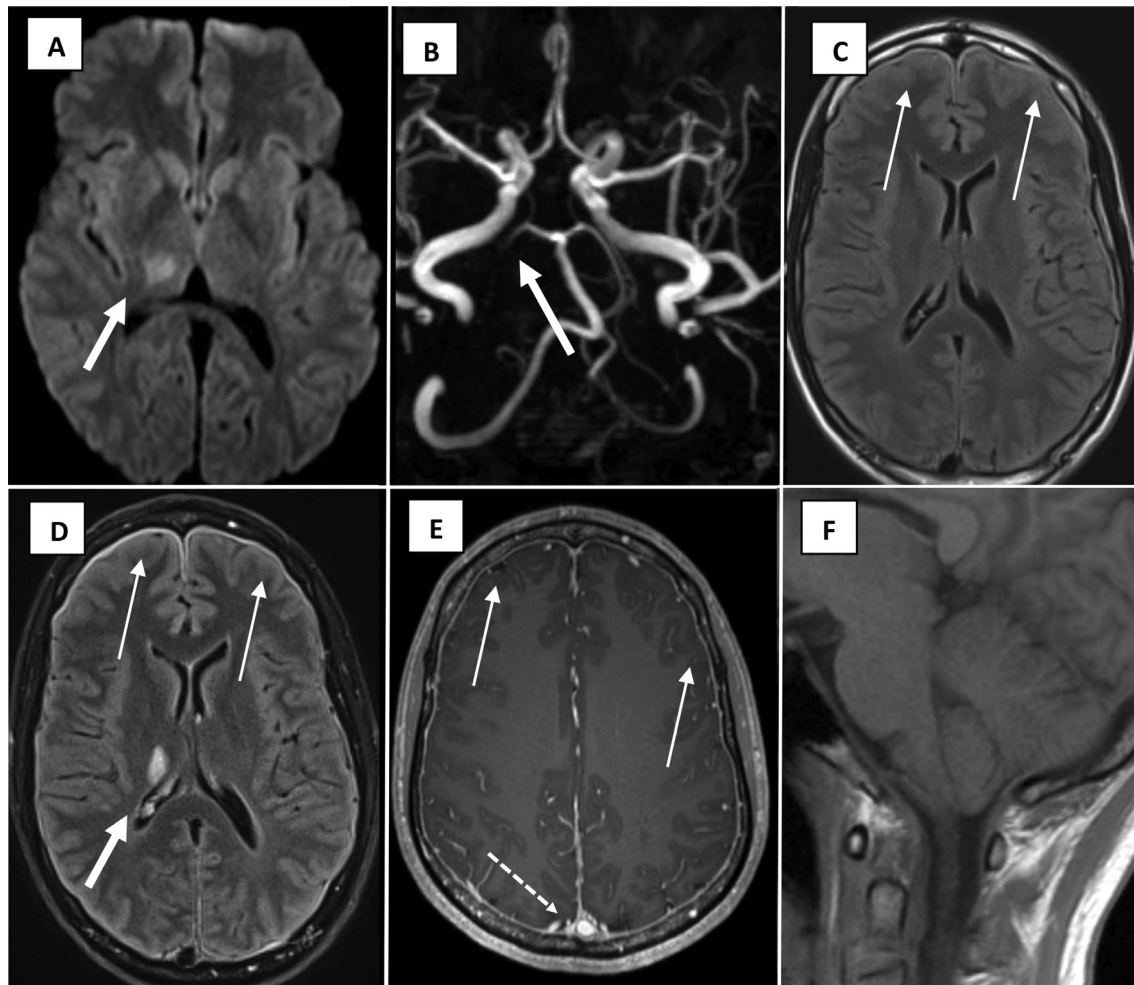


Fig. 1. Stroke MRI without contrast (A, B, C), and brain MRI with contrast, 2 months after stroke (D, E, F). A: Diffusion-weighted MRI lesion indicating a thalamic infarction (wide arrow). B: 3D time of flight MR-angiography showing right posterior cerebral artery occlusion (wide arrow). C: FLAIR sequence: bilateral thickening of the meninges (thin arrows). D: Flair sequence: Right thalamic ischemic lesion (wide arrow) and diffuse pachymeningeal hyperintensity (thin arrows). E: T1 sequence: Venous distension sign (dotted arrow) and diffuse pachymeningeal enhancement (thin arrows). F: T1 sagittal sequence: Mild cerebellar tonsillar descent.

was normal. A blood patch was performed, with complete resolution of headache. The patent foramen ovale was closed.

3. Discussion

This observation highlights the diagnostic difficulties of spontaneous intracranial hypotension. This disease is rare, misdiagnoses are common and associated with radiological complications [1].

According to the last classification of International Classification of Headache Disorders, the diagnosis of intracranial hypotension is confirmed by low cerebrospinal fluid (CSF) pressure (<60 mm CSF) at lumbar puncture or evidence of CSF leakage on brain or spine imaging. Although stroke MRI showed thickened meninges, the typical aspect of intracranial hypotension with dedicated sequences (brain sagging and pachymeningeal enhancement) has been showed 2 months after stroke. Orthostatic headaches secondary to lumbar puncture could be considered. However, the presence of orthostatic headache before stroke, the absence of worsening headache after lumbar puncture, the thickened meninges in stroke MRI supports the diagnosis of spontaneous intracranial hypotension prior to stroke.

Six cases of arterial cerebral infarcts associated with spontaneous intracranial hypotension are reported in literature [2–6]. Key points of each case are summarized in Table 1. The most serious issues are the two cases described by Schievink (fatal outcome or quadriplegia) [6]. Ischemic lesions are typically localized in brainstem and/or posterior cerebral artery. In one case, infarct localization was only defined with computed tomography scan [6]. Pachymeningeal enhancement and bilateral subdural collections were showed in all cases. Evidence of brain sagging or transtentorial herniation was found in all cases. The absence of arterial occlusion was specified in 2 cases. In other cases, no arterial occlusion was reported. As described in our observation, the delay between headache beginning and stroke, and the delay before SIH diagnosis are often several weeks.

The main pathophysiological hypothesis in cases of stroke secondary to SIH is the stretching and occlusion of arteries (in particular perforating cerebral arteries) due to the sagging brain structures. In these cases, intracranial hypotension is typically severe, associated with subdural collections. In 2 cases, brain sagging was associated with Duret haemorrhage. This usually reflects the rapid descent of brain structures and transtentorial herniation.

Table 1
Review of literature.

Author	Age/sex	Brain infarction area (CT scan or MRI)	Other radiologic abnormalities	Delay between SIH and stroke
Whiteley 2003	62/M	brainstem (MRI)	Dural enhancement, bilateral subdural collections, transtentorial herniation	5 weeks
Lai 2007	NS	bilateral PCA territories (MRI)	Duret haemorrhage, subdural hematoma, transtentorial herniation	NS
Chi 2007*	43/M	PCA territory : bilateral occipital and mesial temporal lobes right thalamus, midbrain, upper pons (MRI)	diffuse pachymeningeal enhancement, bilateral subdural haematomas, brain sagging, midbrain and upper pons haemorrhage (Duret haemorrhage)	2 months
Schievink 2013 (case 1)	45/M	PCA territory: bilateral temporal and occipital lobes (CT scan)	pachymeningeal enhancement, bilateral subdural hematoma, brain sagging	1 month
Schievink 2013 (case 2)*	42/M	multiple infarcts in the pons and midbrain (MRI)	pachymeningeal enhancement, bilateral subdural hematomas, brain sagging	10 days
Matosevic 2016**	54/M	recurrent brainstem infarctions (MRI)	pachymeningeal enhancement, bilateral subdural hematoma, brain sagging	Chronic headache

PCA: posterior cerebral artery. NS: Not specified.

* In 2 cases, the absence of arterial occlusion is specified in manuscript.

** In 1 case, ischemic lesions was shown in absence of previous subdural evacuation or decompressive craniotomy.

Other etiologies of subdural hematoma are rarely associated with deep cerebral infarcts or transient ischemic stroke [7,8]. These collections may play a role in cerebral infarction mechanism, by elevation of intracranial pressure, reduction of cerebral perfusion pressure, and cerebral blood flow.

In all cases but one, brain infarction was found in the following days after subdural evacuation or decompressive craniotomy. As reported by Kelley et al., brain sagging could be precipitated by craniotomy, particularly in the case of previous intracranial hypotension [9]. The exposure to atmospheric pressure allows the entrance of air in the subarachnoid space. The loss of CSF buoyancy may result in caudal herniation of the brain.

Such clinical and radiological differences are present in our case, suggesting a different ischemic mechanism than previously described. There was no subdural hematoma. Brain sagging was firstly absent in sagittal MRI sequence. Intracranial occlusion has never been reported in cases of stroke related to intracranial hypotension. In our young patient without other stroke risk factors, a patent foramen ovale was found. This anomaly can be detected in 15% to 25% of the adult population and has been associated with increased risk for ischemic stroke [10]. An embryonic defect in the interatrial septum can be the conduit for a paradoxical embolism from the deep veins of the legs to the brain. The presence of deep vein thrombosis usually supports the implication of patent foramen ovale in cryptogenic stroke. No thrombosis was detected in this case. However, prolonged bed rest is a risk factor for venous thromboembolism. We suggest that association between spontaneous intracranial hypotension and stroke could be explained by thromboembolism in this observation.

4. Conclusion

Stroke is a rare complication of spontaneous intracranial hypotension. The clinical and radiological presentation is typically severe, associating subdural collections and brain sagging. The first pathophysiological hypothesis for arterial cerebral infarction is the stretching of arteries due to the sagging of the brain. We report the first case of stroke complicating spontaneous intracranial hypotension, with definite intracranial artery occlusion. Radiological features are different from other cases, suggesting a different mechanism. In case of prolonged lying down, the presence of patent foramen ovale could be a risk factor for embolic stroke in SIH.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.jocn.2020.01.019>.

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