

CASE REPORTS

Cerebral venous sinus thrombosis caused by spontaneous intracranial hypotension

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Abstract

The association of spontaneous intracranial hypotension with cerebral venous sinus thrombosis is rare. We report here a case of extensive cerebral venous sinus thrombosis involving three sinuses following spontaneous intracranial hypotension. The patient presented no other thrombotic risk factors except for spontaneous intracranial hypotension. This case adds to the evidence that spontaneous intracranial hypotension is a risk factor for cerebral venous sinus thrombosis.

INTRODUCTION

The association of spontaneous intracranial hypotension (SIH) with cerebral venous sinus thrombosis (CVST) is rare. We report here a patient with SIH who subsequently developed CVST. The patient had no other thrombotic risk factors. We believe that SIH should be considered as a risk factor of CVST.

CASE REPORT

A 40-year-old man was admitted to the Qilu Hospital of Shandong University for acute onset of severe fronto-occipital headache without any obvious causes. The headache worsened in the upright position in 10 seconds and was completely relieved within 20 seconds after lying down. The headache was severe with visual analogue score of nearly 9. He did not have fever, nausea, vomiting or tinnitus. The patient was otherwise well with no past medical history of significance. General and neurological examination was normal. Investigations including blood cell count, ESR, coagulation function were all normal. Serological investigations including anti-phospholipid antibodies (anticardiolipin antibody, lupus anticoagulant, anti- β_2 -glycoprotein I antibody), and rheumatoid antibodies were also normal. MR venous angiography showed normal flow in the venous sinuses, but contrast MRI revealed diffuse dural thickening and enhancement, which were consistent with intracranial hypotension

(Figure 1). Lumbar puncture revealed a low initial CSF pressure of 5 cmH₂O. The white cell count was 38/mm³ (lymphocytes 94%), protein 2mg/ml, elevated IgG and IgA level, normal glucose and chloride. He was diagnosed to have SIH, and was treated with hydration and prednisone for two weeks. His headache was much improved 3 weeks after commencement of treatment.

The patient developed new symptoms one month after the initial presentation, with numbness, slurred speech and a feeling of swelling at the top of the head in the upright position lasting for more than half an hour. MRI repeat showed thrombosis in the superior sagittal sinus, left lateral sinus and sigmoid sinus (Figure 2). The second lumbar puncture revealed CSF pressure of 9 cmH₂O, white cell counts of 10/mm³ (predominantly lymphocytes), protein level of 1.7mg/ml, and normal glucose and chloride level. The patient was diagnosed to have CVST caused by SIH. He was treated with low-molecular-weight heparins for 7 days followed by oral anticoagulant (warfarin) for 10 months. The clinical symptoms and signs disappeared after 2 months of anticoagulant therapy. Follow-up brain MR venous angiography at 1, 3, 8 months after warfarin therapy showed that the clotted sinuses improved gradually with near complete recanalization of the dural sinuses by 8 months (Figure 3). One month after stopping oral warfarin therapy, the patient had his blood checked for protein C, protein S and antithrombin. The results were all within the normal range.

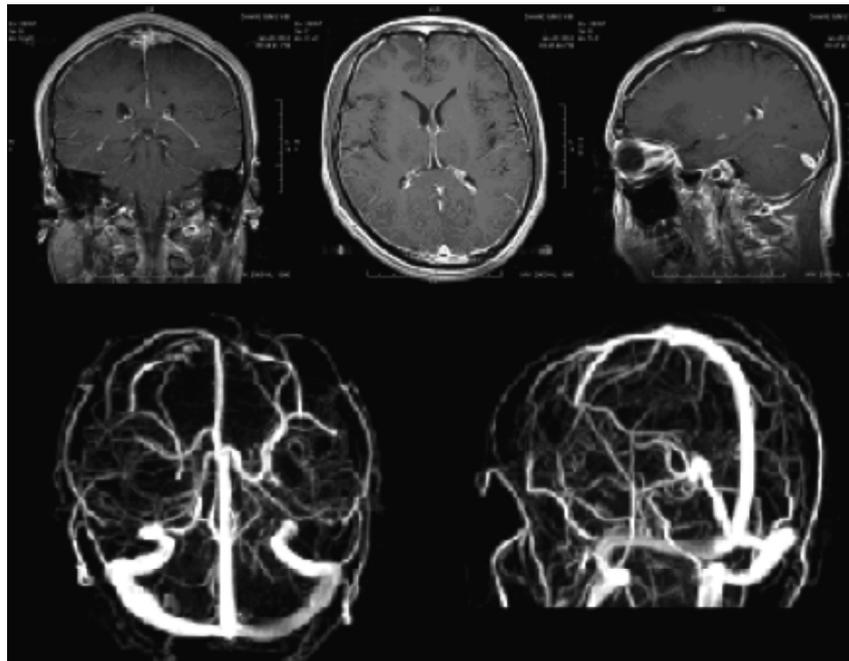


Figure 1. Contrast-enhanced T1 weighted MRI images at presentation revealed diffuse dural thickening and enhancement consistent with intracranial hypotension during the initial presentation. MR venous angiography showed normal flow in the venous sinuses.

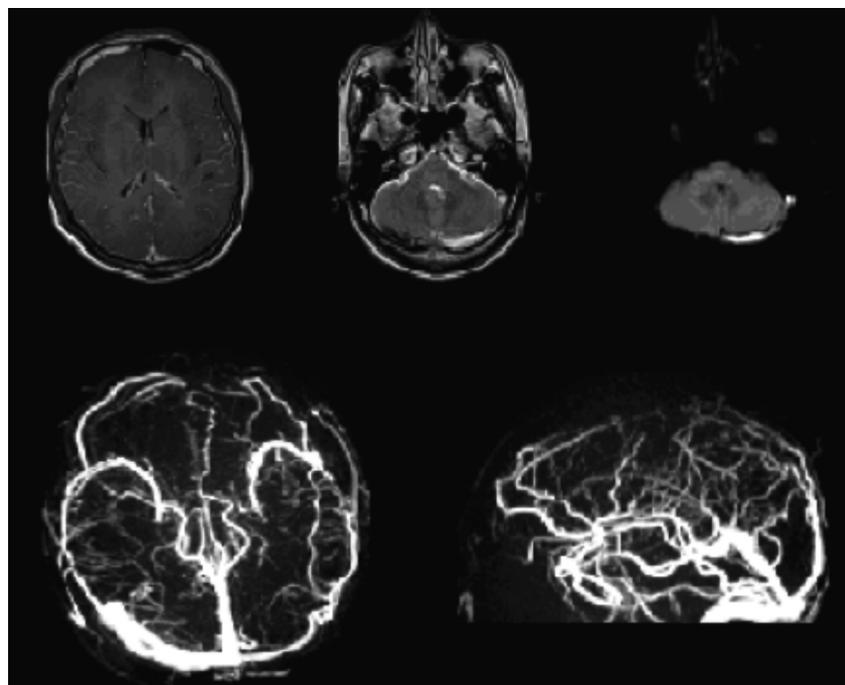


Figure 2. Repeat MRI and MR venous angiography one month after initial presentation. Upper panel showing filling defect in the superior sagittal sinus in contrast-enhanced T1WI, increased signal intensity of left transverse sinus in T2WI and DWI suggestive of cerebral venous sinus thrombosis which was confirmed by MR venous angiography in the lower panel.

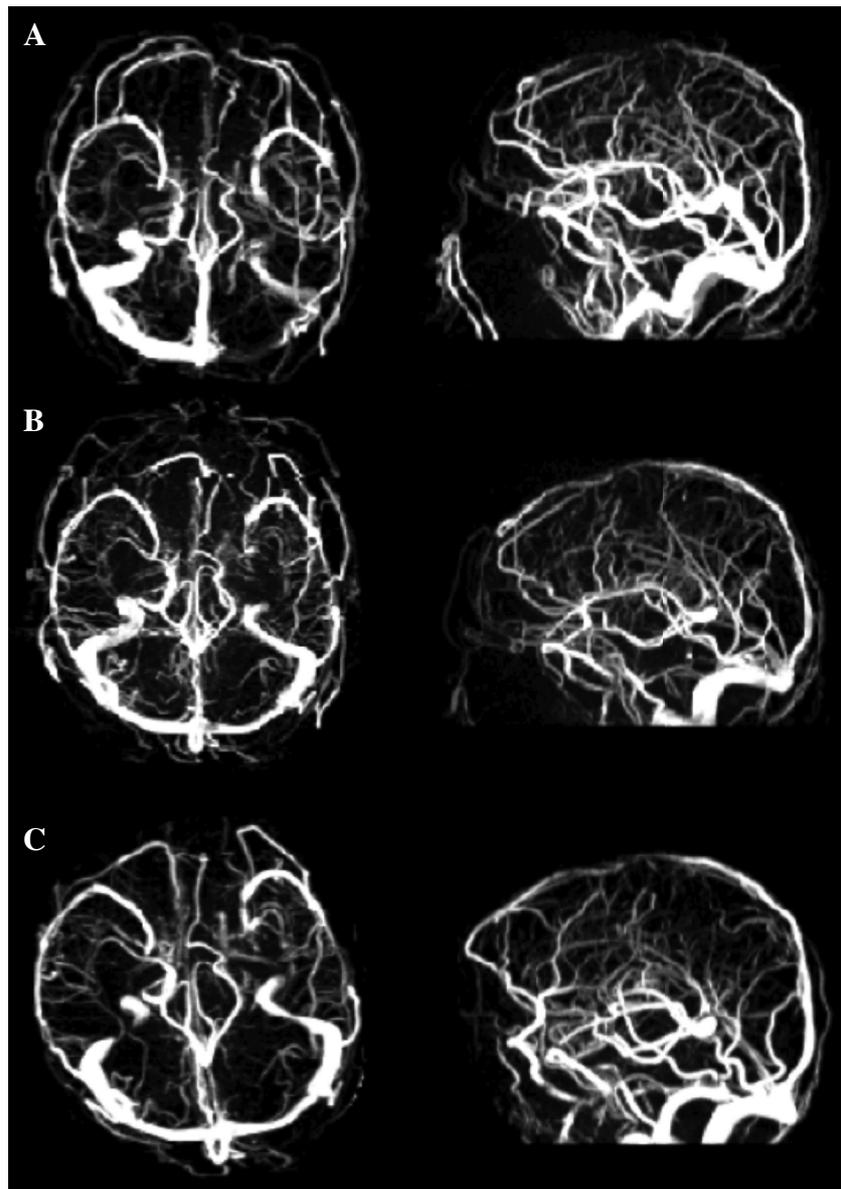


Figure 3. Follow up MR venous angiography after warfarin treatment. A: One month of warfarin therapy: filling defect in left transverse sinus and the superior sagittal sinus; B: Three months of warfarin therapy: Recanalization of left transverse sinus and superior sagittal sinus thrombosis; C: Eight months of warfarin therapy: Near complete recanalization of the dural sinuses indicating gradual improvement of the CVST.

DISCUSSION

The majority of patients with SIH presents with orthostatic headache. Intracranial hypotension may also produce traction on cranial nerves. MRI is the non-invasive gold standard for diagnosis of SIH.¹ From the onset, our patient's headache were orthostatic and could be completely relieved by lying down, which suggested that the headache

was due to SIH. The initial MRI was also consistent with SIH, and the MR venous angiography then was normal. Our patient subsequently developed new neurological symptoms and signs. The repeat MRI and MR venous angiography confirmed the diagnosis of CVST. Our patients had no any acquired risk factors for CVST, such as trauma, malignancy, or any evidence for a genetic thrombophilia on extensive laboratory testing.

The SIH that precede the onset of CVST suggests that SIH is a causative factor for the development of CVST in our patient.

CVST has occasionally been observed in patients with SIH.² To our knowledge, less than 10 cases of SIH with CVST have been reported in the medical literature.^{1,3-8} It has been shown⁶ that the areas of dural sinus sections were about 70% larger in patients with SIH than in normal. The enlargement of the dural sinuses was well recognized by comparing the MR images obtained during the onset of SIH and after its disappearance, either spontaneous or following epidural blood patch or other therapeutic interventions.^{3,7}

This venous dilatation compensates for the fall in CSF volume and pressure. But the venous dilatation also leads to a decrease of blood flow velocity. A 47% decrease of this flow was measured in the straight sinus after a lumbar puncture.⁹ The decrease of CSF absorption into the cerebral venous sinuses in CVST may also result in an increase of blood viscosity in the venous compartments.¹⁰ SIH is associated with sagging of the brain tissue due to the loss of buoyancy. This may cause traction on cerebral veins and sinuses and results in a mechanical distortion of the vessel wall. The endogenous blood coagulation pathway is then activated.² All these factors may contribute to SIH developing CVST. We thus agree with Schievink and Maya², who feel that although CVST is found in only about 2% of patients with SIH, SIH should be included in the risk factor for the development of CVST.^{11,12}

DISCLOSURE

Conflicts of interest: None

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