Spontaneous intracranial hypotension complicated by cerebral venous thrombosis: a case report

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1 INTRODUCTION

Cerebral Venous Thrombosis (CVT) has been rarely reported as a complication of Spontaneous Intracranial Hypotension (SIH). Venous sinus dilation causing blood flow slowing combined with dural sinus anatomical distortion due to brain sagging were two proposed mechanisms by which SIH could precipitate CVT. However, a few case studies have reported a clear temporal association between the two conditions, with SIH preceding the occurrence of CVT.

We report a patient with SIH who presented with postural headache with an initially normal venous CT angiography and had an MRI performed one week later confirming SIH and revealing the development of CVT.

2 CASE REPORT

A 47 years-old female patient with medical history of episodic migraine and use of oral contraceptive experienced a sudden onset of a severe bilateral headache associated with neck pain, tinnitus, dizziness, and ear fullness. The pain progressed over the following hours and became daily persistent, being relieved by lying supine and restarting a few minutes after standing. One week after symptoms had started, she had a head CT with venous angiography performed, which returned normal.

Her headache persisted over the following week and she evolved with a focal motor seizure. An MRI angiography was then performed and revealed pachymeningeal diffuse enhancement, subdural effusions, decreased mamillopontine distance, and enlarged venous sinuses with thrombosis of the superior sagittal sinus and cortical veins.

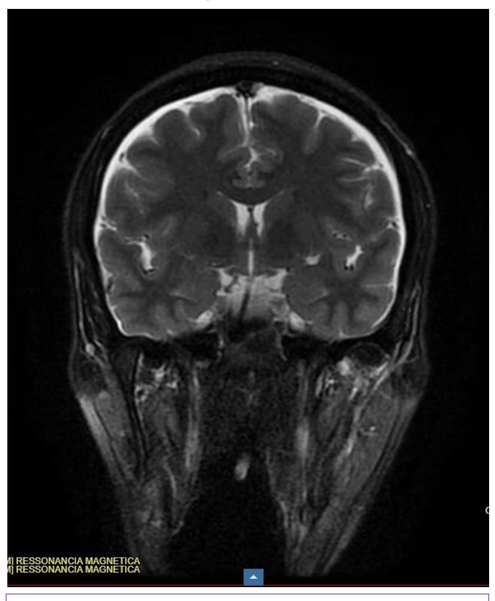

Figure 1: subdural fluid collections

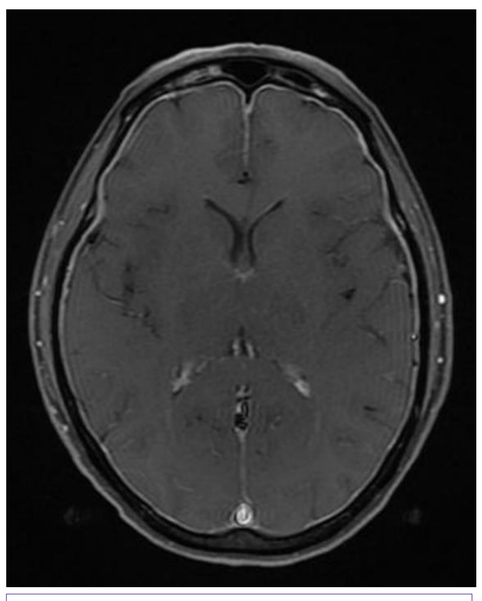

Figure 2: pachymeningeal diffuse enhancement

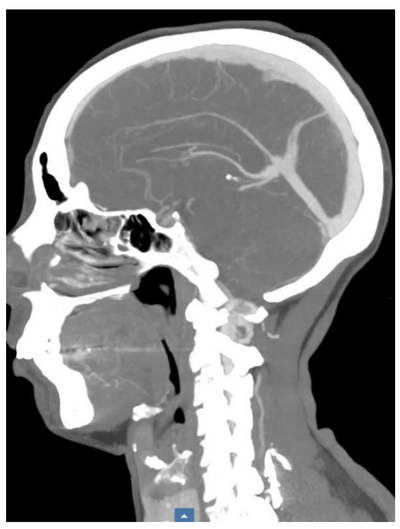

Figure 3: Superior sagittal sinus showed **no** signs of thrombosis in the first week of symptoms

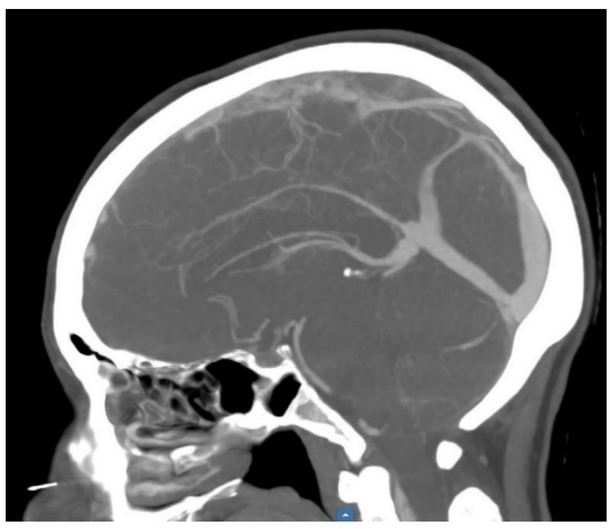

Figure 4: One week after, CT showed an extensive filling defect involving the superior sagittal sinus, consistent with acute cerebral venous thrombosis.

An extensive investigation for genetic and acquired thrombophilia was performed, including tests for Factor V Leiden, prothrombin mutation, anticardiolipin, lupus anticoagulant, and Beta2-glycoprotein 1 antibodies, all of which returned negative results. Additionally, the homocysteine and antithrombin III levels were found to' be within the normal range. In the search for occult neoplasia, both abdominal and pelvic CT scans, as well as mammography, were conducted, revealing no significant changes.

She underwent a spine MRI, which revealed an epidural fluid collection in the mid-thoracic segment. To investigate the presence of a cerebrospinal fluid (CSF) fistula, dynamic CT myelography was performed, revealing a dural contrast leak at the T12-L1 spinal level.

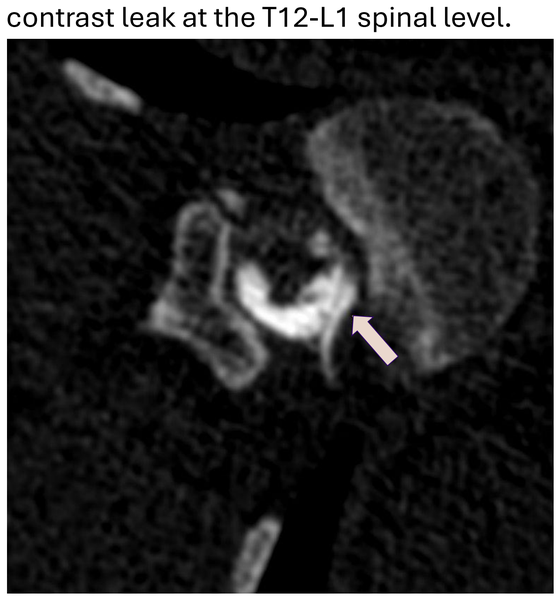

Figure 5: Dynamic CT Myelotomography showing a contrast leak at the T12-L1 spinal level. The arrow indicates the site from which the contrast began to flow.

To treat the CSF leak, a guided epidural blood patch was performed, followed by the initiation of enoxaparin for the treatment of cerebral venous thrombosis. The patient experienced complete relief from the headache after a few days.

3 DISCUSSION

The presence of CVT has been observed in 2% of patients with SIH [1]. The high rates of co-occurrence of these two rare conditions make it unlikely that they are associated by chance. However, there is still controversy over their causal relationship, akin to the old 'chicken and the egg' problem: CVT could increase CSF pressure, contributing to fistula formation. On the other hand, SIH could lead to CVT through its effects on Virchow's triad.

Currently, the most widely accepted understanding is that SIH probably contributes to the genesis of CVT. At least 7 case reports have suggested that SIH occurred before CVT [1,3], as documented through neuroimaging in our case, and none have shown the opposite sequence of events. In addition, at least three pathophysiological mechanisms have been proposed to explain how SIH could lead to CVT, based on Virchow's triad which suggests venous stasis, hypercoagulability, and endothelial injury as the main determinants of venous thrombosis:

1. According to the Monroe-Kelly doctrine, the reduction in CSF volume through leakage, as observed in SIH, leads to compensatory dilation of the venous compartment. Indeed, engorgement of the dural venous sinuses is reported in 75% of patients with SIH [4]. Venous sinus dilation increases the sinuses' cross-sectional area, resulting in a decrease in venous flow speed. In fact, transcranial Doppler has demonstrated a 25% decrease in blood peak velocity in the transverse sinus after a spinal tap [5], supporting this hypothesis. This venous stasis could contribute to CVT.

2. CSF leakage could also reduce CSF absorption in the arachnoid granulations located in the venous sinuses. This would make the blood flowing through the dural sinus more concentrated, leading to hypercoagulability and CVT [6,3].

3. The sagging of the brain observed in patients with SIH could lead to anatomical distortion of the venous sinuses, further increasing venous stasis. This concept is supported by the higher rates of involvement of the superior sagittal sinus in patients with SIH than reported in general cohorts of CVT [3].

4 CONCLUSION

CVT has recently been reported as a rare complication of SIH. Despite the presence of putative mechanisms relating the two conditions, there is still controversy over their causal relationship. By showing a temporal progression from SIH to CVT in this case, we reinforce the concept that SIH could lead to CVT.

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