

Spontaneous intracranial hypotension in Hashimoto's thyroiditis

A case report

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Abstract

Rationale: Autoimmune thyroid diseases (ATDs) seldom affect intracranial pressure. Here, we describe a case of spontaneous intracranial hypotension (SIH) related to Hashimoto's thyroiditis (HT), which has never been previously published.

Patient concerns: A 54-year-old woman was admitted to our hospital due to sudden-onset postural headache, neck pain stiffness, visual blurring, nausea and vomiting. The headache was aggravated when the patient sat or stood up yet rapidly resolved within 1 minute after recumbency.

Diagnosis: The patient had a grade 1a diffusely palpable thyroid gland and the laboratory report revealed elevated titers of anti-thyroid antibodies, a diagnosis of HT was established. Gadolinium-enhanced magnetic resonance imaging exhibited dilatation of the venous sinuses, obvious diffuse pachymeningeal enhancement, and narrowing of the ventricular system, combined with the lumbar puncture pressure was 60 mmH₂O in the lateral recumbent position, and a diagnosis of SIH was established.

Interventions: The patient was placed on strict bed rest and hydrated for 15 days.

Outcomes: The headache was gradually relieved, a follow-up at 1 month after discharge revealed that the patient could stand and function normally.

Lessons: SIH can be related to ATD.

Abbreviations: ATD = autoimmune thyroid disease, CSF = cerebrospinal fluid, HE = Hashimoto's encephalopathy, HT = Hashimoto's thyroiditis, MRI = magnetic resonance imaging, SIH = spontaneous intracranial hypotension, T3 = triiodothyronine, T4 = thyroxine, TSH = thyroid-stimulating hormone.

Keywords: autoimmune thyroid disease, Hashimoto's thyroiditis, intracranial hypotension

1. Introduction

An intractable headache is a typical feature of spontaneous intracranial hypotension (SIH), and cerebrospinal fluid (CSF) leakage is the most common cause of SIH. Most cases involve thoracic or cervical localization of the CSF leak, and until now, the specific etiology of spontaneous CSF leaks has been unknown.^[1] Orthostatic headache is a classical symptom of SIH and occurs within seconds to minutes after adopting an erect

position and is relieved with a recumbent position within several seconds or minutes. In addition, there are still some atypical symptoms, including nausea, vomiting, defects in the visual field, blurred vision, neck stiffness, neck pain, diplopia, tinnitus, and numbness of the face.^[2]

Hashimoto's thyroiditis (HT) is considered an autoimmune thyroiditis, which is characterized by the production of autoantibodies and immune cells by the thyroid gland.^[3] Hashimoto's encephalopathy (HE) is a type of encephalopathy related to HT, and the clinical presentations of "encephalopathy related autoimmune thyroid disease (ATD)" may involve stroke-like events, epilepsy, deteriorating cognition, myoclonus, nystagmus, and neuropsychiatric symptoms.^[4] Although rare, an autoimmune thyroid disorder can alter intracranial pressure. SIH in a patient with Graves' disease was reported by Cusan et al in 2016.^[5] Additionally, a patient who had reversible benign intracranial hypertension with autoimmune hyperthyroidism was reported by Merckenschlager et al.^[6] Herein, we describe a case of SIH related to HT to further strengthen the association between ATD and SIH.

2. Case presentation

A 54-year-old woman was admitted to our department because of sudden-onset headache, nausea, vomiting, neck pain stiffness, and visual blurring. The headache was severe if the patient sat or stood up but was ameliorated in less than 1 minute if the patient

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Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.

We declare that we have no conflict of interest.

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laid down. The patient had a previous history of nasosinusitis and had undergone an operation 2 years previously. The patient denied having any headaches since then. She denied any recent colds or fevers, tinnitus, and photophobia and had no history of lumbar puncture, trauma of the spinal column, surgery, strenuous exercise, sneezing, or straining. In addition, the patient had no relevant family history or medication history.

The patient's temperature (36.7 degrees Celsius), blood pressure (110/70 mmHg) and pulse (72/min) were all normal. The thyroid gland was determined to be grade 1a and was diffusely palpable. In addition, the cardiovascular, respiratory, abdominal, and nervous system examinations were all within normal limits. The lumbar puncture pressure was 60 mmH₂O in the lateral recumbent position. The laboratory test results were as follows. The CSF sample was normal, triiodothyronine (T3) 1.26 nmol/L (normal 0.88–2.44 nmol/L), thyroxine (T4) 82.17 nmol/L (normal 62.68–150.8 nmol/L), serum free T3 4.01 pmol/L (normal 2.63–5.7 pmol/L), serum free T4 13.46 pmol/L (normal 9.01–19.05 pmol/L), thyroid-stimulating hormone (TSH) 1.19 mIU/L (normal 0.35–4.94 mIU/L), TSH receptor antibody 0.48 IU/L (normal ≤1.75 IU/L), anti-thyroid microsomal antibody 275.93 IU/mL (normal ≤5.61 IU/mL) and anti-thyroglobulin antibody 13.35 IU/mL (normal ≤4.11 IU/mL). The patient was a stationary alpha Mediterranean gene carrier (−α^{3.7}/αα), and her hemoglobin was 115 g/L (normal 115–150 g/L). The erythrocyte sedimentation rate was 11 mm/first hour (normal ≤38.4 mm), C-reactive protein 1.03 mg/L (normal 0.068–8.2 mg/L), anti-cyclic citrulline peptide antibody 5.35 RU/mL (normal ≤5 RU/mL), antinuclear antibodies, antiphospholipid antibodies, anti-neutrophil cytoplasmic antibodies, and rheumatoid factor were all within normal limits.

A thyroid color ultrasound showed thyroid parenchymal echo thickening and an increased blood flow signal. Gadolinium-enhanced magnetic resonance imaging (MRI) displayed diffuse enhancement of the pachymeningeal area, a decrease in ventricular volume, and engorgement of the venous sinus, which are findings consistent with SIH (Fig. 1). Treatment of the patient included strict bed rest and hydration for 15 days. The headache was gradually relieved. A follow-up at 1 month after discharge revealed that the patient could stand and function normally.

3. Discussion

Intracranial hypotension, especially SIH, is well known to result from a CSF leak, leading to a decrease in CSF volume, which in turn causes common clinical presentations and sometimes rare clinical manifestations. The former symptoms include orthostatic headache, pain in the neck and interscapular region, numbness of the face, nausea, vomiting, visual blurring, tinnitus, changes in the sense of taste, and limb paresthesias, and rare symptoms include poor memory, parkinsonism, chorea, cerebellar ataxia, change in personality, and galactorrhea.^[7] Orthostatic headache is a type of headache that is aggravated by standing or sitting up and disappears within a few minutes when the patient lays down. The headache experienced by our patient was typical for SIH, in addition to the accompanying symptoms of neck stiffness pain, nausea, vomiting and blurred vision.^[2] The mechanisms of the clinical manifestations of SIH involve sinking of the brain and dilatation of the intracranial venous structures, which gives rise to traction or distortion of the pain-sensitive structures of the brain and further potentiates the orthostatic headache. Compared with men, women are more vulnerable to SIH, and the

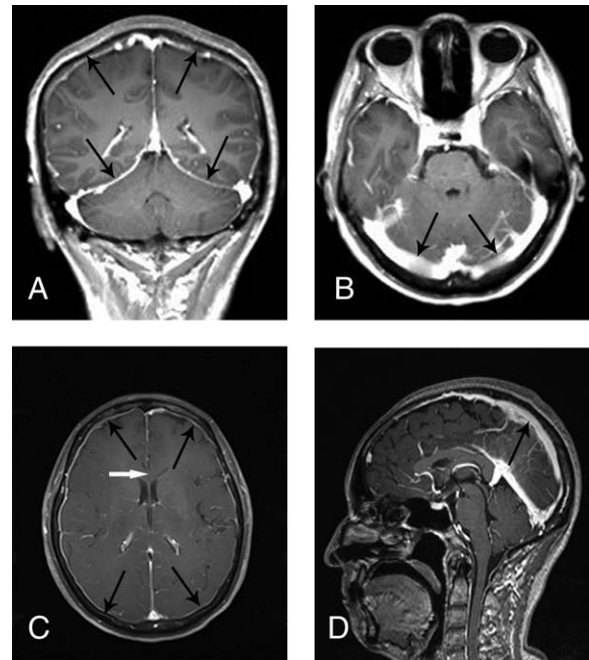


Figure 1. Coronal gadolinium-enhanced T1-weighted MR image showing hypertension of diffuse pachymeningeal enhancement (A, black arrow), axial gadolinium-enhanced T1-weighted MR images demonstrating diffuse pachymeningeal enhancement (B, C, black arrow) and decrease of ventricular volume (C, white arrow), sagittal gadolinium-enhanced T1-weighted MR image revealing engorgement of superior sagittal sinus (D, black arrow). MRI = magnetic resonance imaging.

incidence and prevalence of SIH has been reported to be approximately 2 to 5:100,000, with most patients diagnosed in the 3rd to 5th decade of their life.^[8]

MRI has played a considerable role in the diagnosis of SIH and involves several nonspecific characteristic imaging features. Those imaging features include enhancement of the meninges, engorgement of cerebral veins, pituitary hyperemia, sagging of the brain, subdural fluid collection, and a decrease in the size of the ventricles.^[2] The MRI manifestations of our patient included diffuse meningeal enhancement, decreased ventricular volume, and dilatation of the venous sinuses, which are findings consistent with SIH.

The 3rd edition of the diagnostic criteria for SIH was developed by the International Headache Society in 2013^[9] (Table 1), and our patient met the criteria. As we know, most of the etiologies of spontaneous spinal CSF leak are undetermined, but the underlying fragility of the spinal meninges is generally suspected.^[8] Luckily, in some cases, the CSF leak stops spontaneously with time, and patients become asymptomatic.

Table 1

Diagnostic criteria for headache attributed to SIH according to the ICHD 3rd edition.

- A. Any headache fulfilling criterion C
- B. Low CSF pressure (<60mmCSF) and/or evidence of CSF leakage on imaging
- C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or has led to its discovery
- D. Not better accounted for by another ICHD-3 diagnosis.

CSF=cerebrospinal fluid, ICHD=international classification of headache disorders.

Currently, there are recommended therapeutic modalities for managing SIH including strict bed rest, a generous intake of liquid, caffeine, analgesia, theophylline, abdominal binding, epidural injections (including fibrin glue or homologous blood epidural blood patches) and rarely surgery.^[7] The therapeutic modalities adopted in our case were strict bed rest plus a generous intake of liquid for 15 days, and the patient improved gradually with time.

To our knowledge, HE is a form of ATD and is related to Hashimoto's disease and Graves' disease to a lesser degree. Patients with HE may display euthyroidism (as in our patient), subclinical or clinical hypothyroidism, or rarely hyperthyroidism. The primary laboratory features of HT and HE are the presence of anti-thyroid antibodies, such as anti-thyrotropin antibody, anti-thyroid microsomal antibody and anti-thyroglobulin antibody (particularly the latter 2), which is in agreement with this patient's presentation. Nevertheless, the pathogenesis of HE is still unclear. Whether the existence of anti-thyroid antibodies is only an autoimmune epiphenomenon in the setting of HE or they represent real pathogenic factors in HE is still unknown.^[10]

The clinical features of HE do not distinguish between patients with HT or Graves' disease, which usually consists of epilepsy, mood disturbances, cognitive impairment, changes in overt behavior, stroke-like episodes, myoclonus, and psychosis, as well as less common symptoms, such as nystagmus, dystaxia, headache, palatal tremor, and cerebellar signs.^[11] However, the clinical manifestations of our patient did not include any of these features. Until now, only a few cases in the literature have reported an association between an alteration of intracranial pressure and ATD. Coutinho et al^[12] presented a case of intracranial hypertension with thyroid disturbance, and Cansu et al^[5] reported a case of SIH in GD. These case reports imply a connection between an alteration of intracranial pressure and ATD, but the pathogenesis is still unclear. No articles have described a relationship between SIH and HT. Herein; we are the first to report such an association. The possible mechanism still needs to be determined. In conclusion, the possibility of SIH

should not be ignored in patients with encephalopathy related to ATD. More research is needed to confirm the possible association between these 2 conditions.

Author contributions

Conceptualization: Hui Liang, Jun Shi Hu, Xiong Shi Huang.

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