CASE IMAGE

Supine headache and papilledema: A case and review of cerebral venous sinus thrombosis

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Key clinical message

Cerebral venous sinus thrombosis (CVST) should be on the differential for intracranial hypertension, and the preferred diagnostic tests are CT venogram or MR venography.

Abstract

Cerebral venous sinus thrombosis (CVST) is a rare cause of stroke and is on the differential for intracranial hypertension. Non-contrast head CT is often normal. CT venogram or MR venography are the preferred diagnostic tests, as was required in our patient. We review the presentation, diagnosis, and management of CVST.

KEYWORDS

cerebral venous sinus thrombosis (CVST), magnetic resonance venogram (MRV)

1 INTRODUCTION

A 42-year-old woman presented with headache and papilledema. Magnetic resonance imaging (MRI) brain suggested intracranial hypertension, and MR venography showed a cerebral venous sinus thrombosis (CVST). CVST is uncommon and often missed on non-contrast head CT or MRI brain, thus requires MR venography or CT venogram for diagnosis.

2 | CASE PRESENTATION

A 42-year-old woman presented with 3 years of severe headaches. The headaches worsened when supine or performing valsalva, and were associated with intermittent visual disturbances. Ophthalmologic evaluation identified bilateral papilledema concerning for intracranial hypertension.

Non-contrast MRI brain showed prominent cerebrospinal (CSF) within the optic nerve sheaths, a partially empty sella, and prominent Meckel caves, suggestive of intracranial hypertension. MR venography subsequently revealed a band-like partial filling defect within the left sigmoid sinus down to the jugular bulb, consistent with a chronic left sigmoid sinus thrombosis. (Figures 1 and 2). Our patient was diagnosed with CVST and started therapeutic low-molecular weight heparin. Ultimately, no underlying cause (i.e., see full list below) of CVST was identified.

3 | DISCUSSION

CVST is an uncommon cause of stroke and intracranial hypertension, with an annual incidence of 1–2 per 100,000 person-years. Risk factors include pregnancy/postpartum, oral contraceptives, malignancy, chemotherapy,

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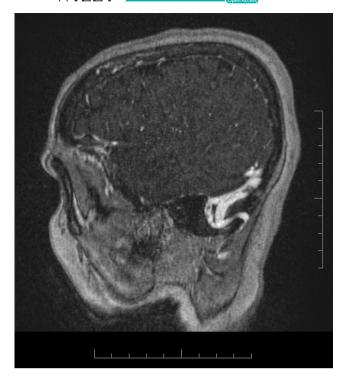


FIGURE 1 MR venography showing left sigmoid sinus chronic thrombosis.



FIGURE 2 Annotated MR venography showing left sigmoid sinus chronic thrombosis.

obesity, genetic thrombophilia, and more recently associated with COVID-19 infection. Thirteen percent of cases have no identifiable cause and are classified as cryptogenic.¹

Patients often present with one of three syndromes: isolated intracranial hypertension (headache, papilledema, visual problems); focal neurologic deficits or seizure; or encephalopathy. Headache is the most common syndrome and is present in 90% of cases. Headache characteristics can be variable, including gradually worsening migraines with or without aura, sudden onset and severe, local or diffuse.

The diagnosis requires urgent neuroimaging. Non-contrast head CT may demonstrate hyperattenuation in the thrombosed sinus, hemorrhagic or non-hemorrhagic edematous lesions adjacent to the thrombosed sinus, or associated intracranial hypertension. However, non-contrast CT is insensitive and can be normal in 30% of cases. The preferred diagnostic tests are CT venogram or MR venography. CT venogram classically shows a filling defect in the venous sinus, called the "empty delta sign" when involving the superior sagittal sinus, with a 95% sensitivity for CVST. MR venography shows absent flow in the involved cerebral venous sinuses or a filling defect.

Therapeutic antiocoagulation is the primary treatment. Associated intracerebral or subarachnoid hemorrhage is not necessarily a contraindication to anticoagulation.

Endovascular treatment (EVT) has been attempted for patients with neurologic worsening despite anticoagulation. However, EVT failed to show improved functional outcomes in a randomized trial (the TO-ACT trial) comparing mechanical thrombectomy and/or local intrasinus thrombolytic administration to standard care.²

Anticoagulation continues for 3–12 months to prevent recurrent thrombosis. Warfarin and direct acting oral anticoagulation (DOAC) have similar outcomes for recurrent thrombosis, death, and recanalization, but DOACs have lower rates of bleeding.³

4 | CONCLUSION

CVST should be considered on the differential diagnosis for intracranial hypertension. A normal non-contrast head CT or MRI brain does not rule out a CVST. The preferred diagnostic tests are CT venogram or MR venography, and acute treatment is therapeutic anticoagulation.

AUTHOR CONTRIBUTIONS

Sachin V. Pasricha: Conceptualization; data curation; formal analysis; investigation; methodology; project administration; resources; software; visualization; writing – original draft; writing – review and editing. **Rajesh Bhayana:** Conceptualization; data curation; formal analysis; investigation; methodology; project administration; supervision; visualization; writing – original draft; writing – review and editing. **Peter E. Wu:** Conceptualization;

data curation; formal analysis; investigation; methodology; project administration; resources; software; supervision; validation; visualization; writing – original draft; writing – review and editing.

FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

The three authors listed above have no conflicts of interest to disclose.

DATA AVAILABILITY STATEMENT

Data pertaining to this patient case, beyond that contained in this publication, is not available publicly due to how this could breach patient consent/confidentiality/anonymity.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

ORCID

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