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American Journal of Emergency Medicine xxx (2016) xxx-xxx

Contents lists available at ScienceDirect



American Journal of Emergency Medicine

The American Journal of Emergency Medicine

journal homepage: www.elsevier.com/locate/ajem

Case Reports

Central venous sinus thrombosis presenting with generalized tonic-clonic seizures in a patient with no known risk factors

Abstract

Central venous sinus thrombosis (CVST) is an uncommon etiology of stroke that predominantly affects women. Nonspecific symptoms include headache, seizure, focal neurologic findings, encephalopathy, and elevated intracranial pressure. We present a case of a previously healthy woman with a new onset seizure, whose only other complaint was weakness despite a normal neurological examination. Her head computed tomography (CT) showed an intraparenchymal hemorrhage of the left frontal lobe and signs of infarction of the right frontal lobe. Magnetic resonance imaging (MRI)/magnetic resonance venography (MRV) revealed a thrombosis in the superior sagittal and right transverse sinus. Further testing revealed a previously undiagnosed malignancy and a previously undiagnosed clotting disorder, both of which contributed to her hypercoagulability. Her case highlights the importance of keeping a high index of suspicion for CVST as examination and CT may be insufficient to make the diagnosis.

The CVST is uncommon primarily affecting women 16 to 60 years of age with peak incidence occurring between 21 and 40 years old. Its varied presentation can make the diagnosis difficult so the clinician must keep a high-level suspicion. Its presentation ranges from headache, intracranial hypertension, seizure, focal neurologic deficits, to encephalopathy [1,2].

The pathogenesis of CVST has 2 main mechanisms:

- 1) Obstruction of venous structures causes increased capillary pressure, which leads to a disruption of the blood-brain barrier and cause vasogenic edema and hemorrhage [3,4].
- 2) Obstruction of venous structures can also cause increased venous pressures and obstruction to the flow of cerebrospinal fluid and result in elevated intracranial pressure [3,4].

Risk factors include prothombotic conditions, exogenous hormone use, pregnancy and puerperium, malignancy, infection, and head trauma.

A 38-year-old woman presented to the emergency department (ED) 30 minutes after a generalized tonic-clonic seizure. She arrived fully alert, and only complained of mild right upper extremity weakness. Review of systems revealed headaches that began 4 months ago. The patient had no history of seizures, coagulopathy, deep vein thrombosis or pulmonary embolism, malignancy, use of oral contraceptives or hormone medications, or head trauma.

The patient's neurological examination was unremarkable and no weakness to the right upper extremity was appreciated. The remainder of her physical examination was normal. ED workup consisted of complete blood count, basic metabolic panel, hepatic panel, lipase, drug screen, urine human chorionic gonadotropin, urinalysis, electrocardiogram, and noncontrast head CT. The head CT showed intraparenchymal hemorrhage in the left frontal lobe with probable infarction in the right frontal lobe.

The patient was given Dilantin for seizure prophylaxis and was admitted to the neurological intensive care unit. While in the intensive care unit, she had a CT angiography of the head and neck which revealed superior mediastinal lymphadenopathy concerning for malignancy. The MRI, MRV, and MRA showed partial thrombosis of the superior sagittal and right transverse sinus with associated hemorrhagic venous infarction of the right frontal region in conjunction with left frontal parietal subcortical hematoma. The patient had a thrombophilia workup, which revealed 2 copies of 4G variant of the plasminogen activator inhibitor-1 gene. Because of the lymphadenopathy seen on CT, she also had a malignancy workup which revealed metastatic papillary thyroid carcinoma.

A patient with suspected CVST should be screened for risk factors (prothombotic conditions, exogenous hormone use, pregnancy, malignancy, and head trauma) when in the ED. Initial laboratory evaluation should include complete blood cell, chemistry panel, and prothrombin time/partial thromboplastin time. A normal D-dimer result does not rule out CVST. In the ED, CTs are readily available and a noncontrast head CT is a good initial test to look for a multitude of causes for the patient's presenting symptoms. The CT may reveal a cord sign, dense triangle sign, or empty delta sign (Fig. 1), which are direct signs of CVST [1,3,5]. However, the noncontrast CT may only be abnormal in 30% of patients with CVST [1]. A normal CT does not rule out CVST [4].

Further imaging of the cerebral venous system is recommended by the American Heart Association and American Stroke Association in patients with lobar intracerebral hemorrhage of otherwise unclear origin or with cerebral infarction that crosses typical arterial boundaries, as well as in patients with symptoms of idiopathic intracranial hypertension. It is reasonable to exclude CVST in patients with headache associated with atypical features [4].

The best imaging modalities to evaluate for CVST are CT venography and MRV. The MRI with MRV is the preferred imaging modality as it has the greatest sensitivity (Fig. 2). On CT venography or MRV, one should see an absence of flow and intraluminal venous thrombus to confirm CVST [4].

Treatment varies according to patient presentation and underlying etiology. Patients should be admitted to a stroke unit for further neurological workup. If the patient presented with seizures, seizure prophylaxis with antiepileptic medications should be started. If increased intracranial pressure is present, it is reasonable to start acetazolamide. If there are no contraindications, anticoagulation should be started with unfractionated heparin or low-molecular-weight heparin and continued for 3 to 12 months. Currently, steroids are not recommended. Neurological worsening occurs in 23% of patients and one-third of

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Please cite this article as: Khatib P, et al, Central venous sinus thrombosis presenting with generalized tonic-clonic seizures in a patient with no known risk factors, Am J Emerg Med (2016), http://dx.doi.org/10.1016/j.ajem.2016.10.026

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Fig. 1. Computed tomography scan showing an empty delta sign [5] Empty delta sign of dural venous sinus thrombosis within the superior sagittal sinus on contrast enhanced CT.

these patients will have a new parenchymal lesion on repeat imaging [3]. If neurological deterioration or coma occurs, more invasive measures such as endovascular therapy or decompressive hemicraniectomy can be undertaken.

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http://dx.doi.org/10.1016/j.ajem.2016.10.026

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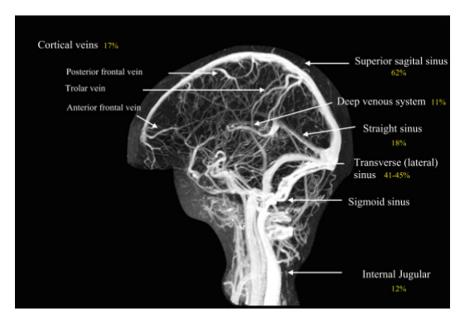


Fig. 2. Magnetic resonance venogram showing the cerebral venous system and most frequent (%) location of cerebral venous and sinus thrombosis [4].

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