Cerebral Venous Thrombosis **Twitter/Instagram:** @emboardbombs

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Introduction

Cerebral vein and dural sinus thrombosis is an uncommon stroke type but is very difficult to diagnose. Only with recent availability of MRI have we been able to diagnose this condition more. The average incidence is 0.22 to 1.57 per 100,000, with a female to male ratio of 3:1.[1,2] Let's review the risk factors, presentation, diagnosis, and management of this uncommon but elusive condition.

Pathophysiology

Incompletely understood, but it's proposed that thrombosis of the cerebral veins or dural sinus blocks blood drainage in the brain, causing stroke. There could be occlusion of the dural sinus, resulting in poor CSF absorption and increased intracranial pressure.[3]

Risk factors

Patients with cerebral vein thrombosis (CVT) are much younger than their average stroke counterparts (~30s vs 60s).[2]

Any prothrombotic condition (genetic or acquired) is a risk factor. Virtually the same risk factors as other venous thromboembolism.

- Think about the usual: malignancy, infection, pregnancy & <6 weeks postpartum, oral contraceptives or hormone replacement therapy, prothrombotic conditions.

Females are at a much higher risk than males. This is likely due to increased risks from pregnancy/postpartum and oral contraceptive use. Oral contraceptives, malignancy, pregnancy/postpartum is the *#1 most common risk factor overall*. It is imperative you have this on your differential in your postpartum HA patients.

In more than 85% of patients with CVT, at least one of these risk factors above is present.
Infections are *rarely* the cause of CVT (about <10% of cases).[2]

Presentation

This is a difficult diagnosis to suspect. It's not helpful that the presentation is extremely variable. It can range from acute to chronic symptoms.[4]

Most common presenting symptom: headache (90% of patients).[2] The headache has no predictable qualities and usually starts gradually. Along with the headache, look for these: papilledema, visual problems, vomiting. Something should be off here. It's not a "typical" tension headache.

Less commonly, there might be seizures, focal deficits, encephalopathy.
Seizures, although uncommon overall in initial presentations, are more frequent in CVT than *any other cerebrovascular disorder*.[5]
As for focal deficits, motor weakness is the most observed deficit.

Cerebral edema, venous infarction, and hemorrhagic infarction are associated with more severe conditions.
Younger patients are more likely to present with acute, less severe symptoms (like headache, vomiting, etc.), versus older adults who have an insidious onset of more abstract symptoms like altered mental status.

Each isolated thrombosis of a particular sinus may present with different symptoms, but there is no need to memorize this. You are still going to work them up the same.

When to think of CVT? nyone with the above risk factors + new onset headache or headache different from usual pattern, symptoms/signs of intracranial hypertension, seizures, encephalopathy, abstract focal neurologic signs/symptoms that do not fit a classic stroke pattern. ***Further, it is critical*** to think of this diagnosis in patients who have presented multiple times to healthcare settings with a headache within a short period of time.

Diagnosis

No lab studies can rule in or rule out CVT. Dimers are not sensitive enough to rule it out.
Lumbar punctures do not rule out CVT either, while they are typically safe to perform and important in ruling out meningitis or encephalitis as a cause of CVT. LP findings are nonspecific and may include lymphocyte pleocytosis, elevated RBC count, and elevated protein.

30-40% can present with intracranial hemorrhage. These hemorrhages are usually focal.[6]

CT imaging is not effective in ruling out CTV. CT imaging is normal in up to 30% of patients, and the findings are often nonspecific.[4] CT can rarely show the "classic" signs of CVT:

- dense triangle sign: hyperdensity with a triangular or round shape in the posterior part of superior sagittal sinus.
- empty delta sign: triangular pattern of contrast enhancement surrounding a central region lacking contrast enhancement in superior sagittal sinus.
- cord sign: linear hyperdensity over cerebral cortex from a thrombosed vein.

These signs are notoriously unreliable and have poor sensitivity.

More commonly, there are "indirect" signs on CT that are associated with CVT, like intracranial hemorrhages, hemorrhagic infarcts, and hypodense lesions caused by edema or venous infarction.

MRI with MRV is the most reliable method of diagnosing a thrombus. An occluded dural sinus or vein is the most sensitive finding.

What if you don't have MRI/MRV available at our facility or during the hours your patient presents?. CT venography might improve accuracy upwards to 90-100%.[7] CT venography has been declared at least equivalent to MRI/MRV, and it is faster and more readily available.

Conventional angiography is reserved for cases where CT venography or MR venography are inconclusive.

Management
The key management pearls revolve around treating the underlying cause and initiating antithrombotic therapy. l These patients will need to be admitted to the hospital. Anyone with neurologic deficits or altered mental status will need ICU level of care for close neurological assessments.

IV heparin drip or subcutaneous low molecular weight (LMW) heparin are both appropriate to start with.
Importantly, the presence of a hemorrhagic venous infarction, intracranial hemorrhage, or isolated subarachnoid hemorrhage are not contraindications. No patients who received heparin in studies developed further bleeding.[8-10]
The consensus from multiple randomized trials is that LMW heparin is more effective and safer than IV unfractionated heparin.[11,12]

For patients who develop further neurologic symptoms despite heparin therapy, endovascular thrombolysis or thrombectomy are traditionally discussed, but studies lack sufficient evidence- this is outside the scope of our review.

Other issues besides anticoagulation include managing increased intracranial pressure (see separate handout on our website here), and seizures.

Observational and retrospective studies (that's all we have available) support seizure prophylaxis in those who have both seizures at presentation and a supratentorial lesion on scan. You should use valproate or levetiracetam.[13,14]

After the acute phase, inpatient specialists will discuss long term anticoagulation with warfarin or a direct oral anticoagulant for up to 12 months.
CVT can result in disability and death, but most cases result in recovery. Recurrence risk is 2-4%.

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