



EM CASES SUMMARY

Episode 181 CVT, IIH, GCA, Peripartum Headache

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Differential diagnosis of acute headache in the peripartum period

While migraine is common in the peripartum period, do not assume that all new onset headaches in this period are caused by migraine.

Life threatening causes of headache to consider in the paripartum patient

1. **Pre-eclampsia/eclampsia/HELLP Syndrome** – highest risk is in first 48hrs however can occur up to 6 weeks after delivery, BP can be only mildly elevated, have a low threshold to check for proteinuria ([deep dive into pre-eclampsia and preterm labour on CritCases](#))
2. **PRES – Posterior Reversible Encephalopathy Syndrome**

- A misnomer as it may include the parietal lobes and frontal in addition to the occipital lobes and it is not always reversible (mortality rate 15%),
 - Presents with headache, seizures, altered mental status and visual loss
 - Diagnosed on MRI (white matter vasogenic edema)
 - Overlap with Pre-eclampsia/eclampsia as it is associated with acute hypertension
3. **Cerebral venous thrombosis** – see below
 4. **Pituitary apoplexy** – rare abrupt severe headache (similar to SAH) with loss of visual field or diplopia, nausea, neck stiffness, altered LOA, some have underlying pituitary adenoma, incited by enlargement of pituitary in peripartum period leading to infarction/hemorrhage, may lead to adrenal insufficiency, diagnosed on MRI (may see hemorrhage on plain CT)
 5. **Cervical artery dissection** – see [Part 1 of this podcast series](#)

Non-life threatening causes of headache to consider in the peripartum patient

1. **Migraine** is the most common peripartum cause of headache that presents to the ED, but do not assume that a new acute headache in peripartum period is migraine – consider it a diagnosis of exclusion
2. **Post-dural puncture headache** – usually within 5 days of epidural, bilateral frontal or an occipital, postural (worse in the upright position, improved in supine position), may have nausea, dizziness, neck pain/stiffness, visual changes,

tinnitus, hearing loss, upper extremity radicular symptoms, treated with caffeine, pregabalin +/- epidural patch

A useful way to think about the differential diagnosis of vascular headaches

One way to think about the differential diagnosis of non-traumatic headaches that originate from blood vessels as outlined by Dr. Baskind includes:

- Clots (arterial and venous) – ischemic stroke, cerebral venous thrombosis
- Tears – cervical artery dissections
- Rupture – SAH (aneurysms, AVMs)
- Inflammation (giant cell arteritis)
- Vasodilation (migraine)

Cerebral venous thrombosis (CVT): A very challenging diagnosis

CVT (or Cerebral venous sinus thrombosis – CVST) can be thought of as 'a DVT of the brain', with similar risk factors. However, in addition to venous obstruction, upstream raised intracranial pressure and decreased cerebral perfusion pressure may lead to brain ischemia and subsequent hemorrhage which are often devastating. COVID infection and vaccination with Astra-Zeneca vaccine have been implicated as risk factors (See [EM Quick Hits 28 on VIPIT – Vaccine-Induced Thrombotic Thrombocytopenia](#)) as well as head and neck infections, although these causes are exceedingly rare. The

challenge with COVID patients and patients with head/neck infections is that most present with headache as one of their symptoms, so unless they present with an obvious neurologic deficit it is difficult to know which of these patients require a workup for CVT. For post Astra-Zeneca COVID vaccine headache, one can safely rule out VIPIT if the platelet count is normal.

CVT is a very difficult diagnosis to make in the ED because the clinical presentation is highly variable and non-specific, a reflection of the various syndromes/locations of the venous obstruction (superior sagittal sinus and transverse sinus being the most common) and pathophysiological changes over time. As a result, the median time to diagnosis between initial presentation and diagnosis is 7 days and return visits to the ED for the same headache should be considered a risk factor. An additional challenge is that plain CT has only a 41% to 73% sensitivity for the diagnosis of CVT, with CT venogram being the diagnostic test of choice in the ED and MRI venogram being the gold standard. An elevated opening pressure on LP is consistent with CVT. Headache caused by cerebral venous thrombosis has no specific characteristics: it is most often diffuse, progressive and severe, but can be unilateral and sudden (even thunderclap), or mild, and sometimes migraine-like. It can mimic migraine, cluster headache, SAH (CVT can be a cause of SAH), headache attributed to low cerebral spinal fluid and primary thunderclap headache.

The many faces of cerebral venous thrombosis

CVT can be divided into 4 syndromes (in order of most to least common):

1. Isolated elevated intracranial hypertension – most common
2. Focal syndrome

3. Diffuse encephalopathy
4. Cavernous sinus syndrome – rare

Headache can be the only manifestation of CVT but, in over 90% of cases, it is associated with focal signs (neurological deficits or seizures) and/or signs of intracranial hypertension, subacute encephalopathy or cavernous sinus syndrome.

Pain can originate from tension on the vein itself or from raised ICP which causes diffuse headache. It may be positional (worse in supine position) and may be aggravated by Valsalva reflecting raised ICP. Additional clinical features may include a variety of neurologic deficits that may not fit a typical anatomical distribution, reflecting the complex pathophysiology (hemiparesis, hemisensory loss, hemianopsia, aphasia), altered level of awareness and seizures. Cavernous sinus thrombosis, a rare complication of head and neck infections, may cause chemosis, proptosis and oculomotor palsy. A key clinical feature of advanced CVT is *papilledema and loss of venous pulsations*, which is often difficult to identify in the ED depending on the fundoscopic skills of the clinician. PoCUS may aid in identifying papilledema by measuring optic nerve sheath diameter, however this too depends on the skill of the clinician and its accuracy is far from perfect. [PoCUS Cases 3 IIH and Ocular POCuS](#).

Pearl: A key feature of advanced CVT is *papilledema*. *Unexplained headache plus papilledema/loss of venous pulsations should be considered CVT until proven otherwise.*

Features that make the diagnosis of CVT *less* likely include purely unilateral pain, scintillating scotoma, recurrent/episodic headache.

D-dimer may have limited utility in low risk patients for cerebral venous thrombosis

D-dimer is not sensitive enough to rule out CVT alone but can be considered in a Bayesian model taking pretest probability into account according to our experts. D-dimer could be part of a rule out strategy when pretest probability is low, concern with radiation exposure is high and there is limited access to MR venogram. The caveat for use of D-dimer is that the literature is not as mature as for PE/DVT, as CVT is way less prevalent. Metanalyses suggest it has somewhat comparable performance compared to use in PE/DVT and can be used in low-risk patients, but there are no good quality prospective RCTs to support this. The clinical application is that D-dimer should not be used in moderate or high-risk pre-test probability patients ie those with risk factors, neurologic deficits, significant clinical suspicion. Shared decision making becomes paramount in these situations.

When to consider CT venogram in the ED to rule out cerebral venous thrombosis

While the pre-test probability needs to be weighed carefully in each patient and there are no validated clinical decision tools to help decide which patients with headache require a CT venogram in the ED, some suggested indications include:

Unexplained headache plus:

- Presence of CVT risk factors
- Sign(s) or symptom(s) of raised ICP (eg. papilledema)
- New focal neurologic sign(s)
- Altered LOC
- Seizure(s)

The mainstay of treatment of CVT is heparin, usually LMWH

The mainstay of treatment of CVT is anticoagulation with heparin, usually LMWH. Hemorrhagic transformation is *not* a contraindication to anticoagulation according to European guidelines.

For critically ill patients who may require decompressive craniotomy or ventricular drain, unfractionated heparin may be preferable. Second line treatments include endovascular therapy, although there is no good evidence in the literature to support its use. Patients are generally admitted to hospital for treatment, MRI to uncover any ischemic transformation and further workup of the underlying cause of the CVT (eg cancer workup).

Idiopathic Intracranial Hypertension (IIH) shares some features with CVT but carries less morbidity/mortality

IIH used to be called "*Pseudotumor Cerebri*" because one of its features is raised intracranial pressure which mimicked the raised ICP seen in some patients with brain tumors before the advent of advanced brain imaging.

In IIH CSF pressure is elevated typically leading to insidious gradual onset diffuse headache over weeks that is aggravated by Valsalva and lying supine, and may progress to visual obscurations and pulsatile tinnitus. Rarely IIH can lead to visual loss due to compression of the optic nerve (hence the change of name from "*Benign Intracranial Hypertension*" to IIH). Like CVT, a key clinical feature is papilledema +/- loss of venous pulsations.

Diagnostic criteria – Idiopathic Intracranial Hypertension (IIH)

1. New headache, or a significant worsening of a pre-existing headache, fulfilling criterion 3
2. Both of the following:
 - Idiopathic intracranial hypertension (IIH) has been diagnosed
 - Cerebrospinal fluid (CSF) pressure exceeds 250 mm CSF (or 280 mm CSF in obese children)
3. Either or both of the following:
 - Headache has developed or significantly worsened in temporal relation to the IIH, or led to its discovery
 - Headache is accompanied by either or both of the following:
 - a) Pulsatile tinnitus
 - b) Papilledema
4. Not better accounted for by another diagnosis

IIH shares some features with CVT – risk factors include young females taking exogenous estrogen, headache aggravated by Valsalva and papilledema with raised ICP, and any patient in whom you are considering the diagnosis of IIH in should also be considered for a work-up for CVT.

Pearl: *key clinical clues of IIH include insidious diffuse headache over days to weeks that is worse on lying supine/on waking in the morning/Valsalva, leading to pulsatile tinnitus and papilledema*
Plain CT is usually unremarkable, and the diagnosis is supported by elevated lumbar puncture opening pressure >250 mm CSF.

Giant cell arteritis (GCA) is a common cause of new onset headache in older patients that is under-recognized in the ED

Presentation and diagnosis of Giant Cell Arteritis – it's not only headache presentations in which GCA should be considered

Giant Cell Arteritis (or Temporal Arteritis) GCA is a common cause of new onset headache in older people with a lifetime risk of 1/100 in women and 1/200 in men, and should be considered in all patients > 50 years of age with a new type of headache, and strong consideration of obtaining a screening ESR and CRP in the ED should be entertained in these patients.

The classic presentation is that of an older person with gradual onset headache over weeks/month, jaw claudication, constitutional symptoms, low grade fever, +/- Polymyalgia Rheumatica (PMR) symptoms (symmetrical aching and stiffness about the shoulders, hip girdle, neck, and torso, worst on arising in the morning) that then progresses to visual loss; however, this classic presentation is seldom seen in clinical practice; in fact, none of the 27 most commonly reported symptoms and signs had a sensitivity greater than 76% in one study.

Diagnostic criteria of Giant Cell Arteritis (3 of 5) – sensitivity of 93.5% and a specificity of 91.2% for GCA

- Age \geq 50
- New headache
- Temporal artery tenderness (or decreased pulse)

- ESR > 50 mm/h **OR**
- Positive biopsy

Fever is common in up to 50% of patients and can be misleading, taking the clinician down the path of infection as the cause of their symptoms. There is an overlap between GCA and Polymyalgia Rheumatica (PMR) of unknown etiology, so ask your patients in whom you suspect GCA about PMR symptoms and vice versa. Visual complications are varied and include ischemic optic neuropathy, amaurosis fugax and rarely, transient diplopia.

Pearl: *the ocular/visual manifestations of GCA are varied (ocular nerve ischemia, central retinal artery occlusion, diplopia, flashers, occipital lobe infarct), so a careful ocular exam recommended.*

Pitfall: *When GCA presents with the chief complaint of headache, it is easier to consider in the differential diagnosis, however it is easy to miss when the chief complaint is "weak and dizzy" or in the patient who presents with fever, or in the patient in whom you are querying an occult cancer as the cause for their constitutional symptoms and weight loss; ie. Keep GCA in the differential diagnosis of "weak and dizzy", fever of unknown source and unexplained weight loss.*

- The sensitivity of ESR > 50 for GCA is as high as 90% ie. It is possible to have GCA with an ESR <50 so in those patients with a high pretest probability and an ESR <50 consideration should still be given to referral for arterial biopsy; be aware that **ESR increases with age** and that sensitivity increases when combined with CRP; do not rely on ESR alone to rule out GCA; one study of 119 patients with GCA showed a sensitivity 97.5% for CRP (< 0.5 mg/dL).

Pitfall: A common pitfall is relying on ESR alone to rule out GCA, which can occasionally be near normal. When considering the diagnosis it is recommended to order both an ESR and a CRP.

ED management of suspected Giant Cell Arteritis – do not delay steroids!

GCA, left untreated, may lead to blindness from a variety of mechanisms as well as ischemic stroke, so steroids should be initiated for all patients in whom you have a high pretest probability before the diagnosis is confirmed by temporal artery biopsy. Patients without any visual symptoms or stroke symptoms may be safely started in oral prednisone (before confirmation of the diagnosis with arterial biopsy) which is highly effective in preventing visual loss, with outpatient follow-up for arterial biopsy within 2 weeks; the accuracy of the biopsy decreases with time, however, even with prednisone on board, it is adequate if done within 2 weeks. Patients with visual and/or ischemic symptoms should be started on IV Methylprednisolone and admitted for further workup.

Pitfall: neglecting to start steroids in those patients in whom you have a high clinical suspicion for GCA from the ED and before they get their biopsy. As long as the biopsy is performed within 2 weeks of starting the steroids, the accuracy of the biopsy is spared. Nonetheless, the sooner the biopsy, the better.

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