

Incidence and Pattern of Headache in Cerebral Venous Thrombosis

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Abstract

Cerebral venous thrombosis (CVT) is a challenging condition because of the variability of clinical presentations. CVT can present at all ages, but is seen more in young and middle-aged women. CVT does not necessarily occur only when there is an obvious underlying etiology. In almost 30% of cases, the etiology cannot be established. CVT can present with an acute thunderclap headache, fever, seizures, focal deficits, impaired sensorium, or papilloedema. Headache is known to be the most frequently associated initial complaint, and is present in more than 80% of patients, but it is not always remembered that headache can be the sole presenting complaint of CVT and even when early papilloedema is absent. Headache can occur in isolation in up to 5% of CVT cases. There is no identifiable, uniform, recognizable pattern of headache in CVT, but this article discusses the "Headache Profile" that is seen more commonly in this setting with an illustration of one such case where the innocuous headache turned sinister.

Magnetic resonance imaging with venography is the investigation of choice to diagnose CVT; computed tomography alone will miss a significant number of cases. One must keep in mind the possibility of CVT in every patient who presents with new-onset headache of any type, any severity, and in any location, particularly when there is worsening in spite of analgesics. Earlier the diagnosis, earlier the treatment, better is the outcome.

Introduction

With the help of modern neuroimaging techniques like MR scan and MR venogram (MRV), cerebral venous thrombosis (CVT) is being diagnosed much more frequently than in the past and is not so uncommon as was generally portrayed. Since there is no proper epidemiological data, the true incidence cannot be correctly assessed. However, in the past 40 years, there have been some clinical series from India that have suggested that the incidence of CVT may be higher in the subcontinent than in western countries.^{1,2}

A high index of clinical suspicion is required to diagnose this uncommon condition so that appropriate treatment can be initiated at the earliest. If diagnosed early and treated early, the prognosis and outcome are favorable, as there is extensive collateral circulation in the cerebral venous system.

CVT - Clinical Profile

CVT can present at all ages^{3,4}, but is seen more in young and middle-aged women and in association with pregnancy, puerperium and oral contraceptive use.⁵ Systemic illness and malignancies, which predispose to CVT would include the very young and old persons.³ CVT does not necessarily occur only when there is an obvious underlying etiology.⁶ In almost 30% of cases, the etiology cannot be established.⁷

Thrombosis usually affects the large cerebral venous sinuses and cerebral veins. The superior sagittal sinus (SSS - 72%) and the lateral sinus (LS - 70%) are most frequently affected. The vein of Galen and the lateral petrosal sinus are uncommonly involved.⁸

CVT is a challenging condition because of its variability of clinical presentations⁹, which is mainly determined by the site, extent and rate of thrombosis. When thrombosis affects a cortical vein, focal deficits and/or seizures may be the presenting features, but if thrombosis is restricted to the SSS and LS, it may present with symptoms of idiopathic intracranial hypertension. Likewise the extent and speed of occlusion due to thrombosis may decide the severity of the deficit, the change in sensorium and rise of intracranial pressure.

CVT - Headache Profile

The diagnosis of CVT should be suspected in any patient who presents with new onset headache that may or may not be associated with Fever, Seizures, Focal deficits and impaired sensorium and Papilloedema.^{10,11}

Headache is known to be the most frequently

associated initial complaint, and is present in more than 80% of patients¹⁰, but it is not always remembered that headache can be the sole presenting complaint of CVT. Headache can occur in isolation in up to 5% of CVT cases. This article mainly discusses patients who may present with headache as the only presenting feature of the CVT.

There is no identifiable, uniform, recognizable pattern of headache in CVT, but more commonly the "Headache Profile" is as follows:

Onset: Could be variable but most commonly it is new onset subacute to rapidly progressive over a few days.¹¹ Only a few patients have sudden onset or even a thunderclap headache.^{11,12}

Frequency: Could be intermittent initially to become a constant headache later on.¹³

Location: Could be unilateral¹³ or localized to any region of the head; need not necessarily be occipital.

Nature of Pain: Usually throbbing.¹³

Severity: Of variable severity, usually above 8/10 in a visual analogue scale (VAS).¹³

Accompaniments

When the headache of CVT is accompanied by focal deficits, seizures, drowsiness or papilloedema, the patient generally presents to the emergency department and is investigated more aggressively with MRI, MR angiography and MR venography¹⁴ in order to establish the diagnosis as well as to rule out other conditions. But when the CVT presents with headache alone, in isolation, without accompanying papilloedema or other signs and symptoms, or when the patient with new-onset headache consults in an office setting, then investigations may stop short with a CT scan. Although the CT scan may demonstrate changes suggestive of CVT - the 'empty delta sign' - pathognomonic of SSS thrombosis¹⁵, we need to realize that the CT scan may be normal in up to 30% of patients with CVT.^{3,7} It would be wiser to do an MR scan in all patients with recent onset headache and include an MRV^{16,17} in the protocol for all patients with new onset headache of whatever location and severity; then the likelihood of missing this potentially treatable condition is minimized.

It would be worthwhile remembering some of the other conditions that can present with new-onset headache and have a normal CT scan. These would need to be considered as the differential diagnosis of CVT (Table).

Case study

Given below is a case study¹⁸ which is illustrative of the dilemma that surrounds CVT when it presents with headache only.

This 42-year-old businessman was seen in the Headache Clinic with complaints of continuous head pain for the past two weeks. This was the first time in life that he had experienced such a headache. To begin with, the pain was restricted to the right hemicranium, mildly throbbing in nature, and then gradually progressed to a generalized headache that was unbearable. There were no specific relieving factors and analgesics only afforded mild relief.

There was no history of vomiting, photophobia, phonophobia, fever or other neurologic/systemic accompaniments. There was no history of head or neck trauma and there were no pointers to suggest an extracranial infection or systemic illness that would explain this new-onset headache in an otherwise healthy individual. There were no trigger links, no aggravating factors and no postural worsening. He was not a known hypertensive or diabetic and there was no past history of migrainous headaches in him or his family members.

His general physical and neurological examination was normal. Blood pressure was 130/90 mmHg and temperature was normal. Pupillary examination and fundoscopy were normal, there was no papilloedema; cranial nerves and rest of the neurological examination were normal. In view of the new-onset of headache and clinical examination being non-contributory, he was advised a CT head scan on an out-patient basis, which was reported to be normal.

Three days after his out-patient consultation, he was rushed to the intensive care unit in an unconscious condition with a series of generalized convulsions and mild right hemiparesis.

On examination in the ICU, the patient was unconscious and poorly responsive to painful stimuli, temperature was normal, blood pressure was 130/90 mmHg, there was no neck stiffness, pupils were equal but sluggish in reaction, there was early papilloedema, which was not seen earlier. He had mild right hemiparesis and both plantars were extensor. Systemic examination was normal.

The convulsions were brought under control with parenteral phenytoin and diazepam. He was started on antioedema measures and antibiotics. His repeat CT scan, keeping in mind this new development, was reviewed and found to be non-revealing. Finally, an MR scan was ordered, which showed thrombosis of the left sigmoid and transverse sinus with a fairly large temporoparietal acute haematoma and multiple fluid levels within it suggestive of a haemorrhagic venous infarct with areas of recurring bleed. There was significant adjacent mass effect with peri-lesional oedema causing a subfalcine herniation. There was evidence of bleed within the right lateral ventricle.

Table. Differential Diagnosis of New-onset Headache and Normal CT Scans.

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| * | Carotid or vertebral dissection |
| * | Central nervous system vasculitis |
| * | Isodense subdural hematomas |
| * | Encephalitis |
| * | Meningitis |
| * | Infiltrative gliomas |
| * | Pituitary tumor |
| * | Spontaneous intracranial hypotension |
| * | Benign intracranial hypotension |

With a confirmed diagnosis of cerebral venous thrombosis (CVT), he was investigated extensively keeping in mind the various underlying causes that could contribute to venous occlusion. He was anticoagulated routinely first with intravenous heparin and then with low molecular weight heparin (LMWH). His level of consciousness improved but he was left with dysphasia and residual right hemiparesis. After a prolonged hospital stay he was discharged on oral anticoagulants.

Conclusion

CVT can present with any type of headache. When the presentation is one of thunderclap headache to the emergency room, then usually, but not always, the diagnosis is made, even if not initially considered. CVT can present with headache alone, it does not always present with a background setting, and may be mistaken for primary headache disorders. CT with or without contrast is not sensitive enough to rule out CVT in patients with progressive headache. The remarkable advent of MRI and in particular the use of MRV has made this diagnosis a lot easier. CVT should be looked at as a secondary headache that can mimic a primary headache. If CVT is given the same level of scrutiny as subarachnoid bleed, then few will be missed in the future.

In conclusion therefore, one must keep in mind the possibility of CVT in every patient who presents with new-onset headache of any type, any severity, and in any location, particularly when there is worsening in spite of analgesics. An MR scan + MR venogram would be the only way to make the diagnosis at an early stage in such patients, and earlier the diagnosis, earlier the treatment, better is the outcome.

In Summary

The pathogenesis of isolated headache in CVT in the absence of a setting is unknown but may involve changes in the walls of the occluded sinus.¹³

Magnetic resonance imaging with venography is the investigation of choice to diagnose CVT; computed tomography alone will miss a significant number of cases.

Not all patients with CVT have risk factors for venous thrombosis. The diagnosis is easy when there is unexplained headaches with risk factors like oral contracep-

tives, systemic lupus erythematosus (SLE), or recurrent venous thrombosis. This justifies asking for an MRV to rule out CVT.

It is important to remember that CVT can present with headache alone when early papilloedema may be absent.

We do not have evidence to conclude whether the prognosis of patients with CVT where headache is the only symptom is similar to patients with focal neurological signs or epileptic seizures¹⁹, so whether these patients should be treated with IV heparin²⁰, and if so for how long the anti-coagulation should be sustained - we really do not have answers to all these questions.

CVT is treatable and it is truly a clinical diagnosis that most neurologists, who miss it the first time, remember for a lifetime!

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