HEADACHE AS THE ONLY PRESENTATION OF CEREBRAL VENOUS THROMBOSIS: A CLINICAL CASE

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ABSTRACT

Cerebral venous thrombosis (CVT) is an uncommon cerebrovascular disease presenting with a remarkably wide spectrum of signs and mode of onset. Headache is the most frequently occurring and the commonest initial symptom. In this article we present a case of CVT occurred in a middle-age female patient, with headache as its only clinical presentation, and oral contraceptive use as only risk factor. In spite of a radiological evidence of parenchymal softening in the right temporal lobe, she had a neurological examination substantially normal. There was a delay of diagnosis, but clinical course and prognosis were favourable.

Key words: Headache-Cerebral venous thrombosis- Venous infarct.

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Introduction

Headache is by far the most frequent symptom and also the inaugural feature of cerebral venous thrombosis (CVT), which is an uncommon brain vascular disease presenting with a remarkably wide spectrum of symptoms. Headache in CVT patients has no specific features and is associated usually with papilledema, focal deficits, disorders of consciousness, seizures, cranial nerve palsies. Some Authors grouped these symptoms into specific main syndromes: isolated intracranial hypertension, diffuse encephalopathy, cavernous sinus syndrome.

In rare cases the headache is not accompanied by any clinical finding and only computed tomography (CT) scanning or cerebrospinal fluid (CSF) examination reveals conditions related to CVT aetiology, such as cerebral venous infarction, subarachnoid haemorrhage, or meningitis. Headache in the absence of such conditions as the only symptom due to CVT is extremely rare but reported.

In this article we report the case of a patient with CVT and venous infarction revealed by CT of the head and confirmed with magnetic resonance imaging (MRI), but who had headache as only clinical sign of presentation of the disease.

Case report

A 49-years-old woman came to emergency complaining severe headache without fever and other neurological or systemic accompanying signs or symptoms. Headache was described as near-daily, continuous pain restricted to right hemi cranium, mildly tightening in nature, for two weeks prior. This was the first time in her life that she had experienced such a headache. Initially analgesics afforded mild relief, but the night before admission the patient complained of a much-increased pain, which gradually progressed to a generalized headache that was unbearable.

Patient had no history of tobacco, alcohol, and had no significant family history. She was taking combined oral estrogens and progestin for contraceptive use; she didn’t suffer of high blood pressure or diabetes, neither there were indicators for extracranial infection or systemic illness. Patient denied any spontaneous abortions.
Physical examination was unremarkable, with a normal blood pressure and body temperature. Cranial nerves were intact, no neck rigidity, and strength, sensation, reflexes, and results of gait testing were normal; Babinski’s sign absent. No focal deficits were noted except for a mild difficulty to perceive a finger in the extreme side of the left visual field. Ocular examination and pupillary reflexes were normal, and fundoscopy no revealed papilledema. Abstract thinking was intact and spontaneous language use while providing the history was fluent.

Laboratory testing showed blood chemistries and complete blood count to be normal. Prothrombin time and partial thromboplastin time were not increased. An urgent CT of the head showed a right temporal infarction in the posterior region associated with spontaneous hyper-density of the right lateral sinus. No haematoma or midline shift was appreciated.

A possible thrombosis of the transverse sinus was suspected, and a MRI was ordered which confirmed (fig. 1) the diagnosis. Magnetic resonance venography (MRV) demonstrated decreased flow signal in the right transverse and sigmoid sinuses consistent with venous thrombosis (fig. 2).

Patient underwent to systematic aetiological work-up in which we looked for the main causes and risk factors of CVT such as systemic diseases, malignancies, haematological disorders, antiphospholipid syndrome, and local infectious or non-infectious causes. None specific cause was found, and birth control pills use remained the unique risk factor associate.

Treatment was immediately started based on subcutaneous low molecular weight heparin. During hospitalization patient did not show clinical complications such as seizures, intra-cranial hypertension, neither consciousness deterioration. Headache significantly improved and after two weeks patient was discharged with anticoagulant oral therapy.

Discussion

In CVT there is not a uniform pattern or specific characteristics of headache. Most often it is diffuse, progressive, severe and associated with other signs of intracranial hypertension. It can also be unilateral and sudden, and sometimes very misleading, mimicking migraine, or primary thunder-clap headache.

Headache can be the only manifestation of CVT but in the usual presentation is associated with focal signs (neurological deficits or seizures) and/or signs of intracranial hypertension, sub acute encephalopathy or cavernous sinus syndrome. According the new classification of the International Headache Society (ICHD-II criteria) it is classified as headache attributed to cranial or cervical vascular disorder (see table 1)\(^\text{6}\).

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**Fig 1**: T2-weighted FLAIR axial magnetic resonance of the brain showing the ischemic lesion in the right temporal lobe, without mass effect, and the “hyper signal” of the right transverse sinus suggestive for venous thrombus (white arrows).

**Fig 2**: MRV with TOF technique showing no visualization of the right transverse and sigmoid sinuses, confirming the diagnosis of thrombosis; note that jugular vein is also not seen.

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**Table 1**: Diagnostic Criteria of the International Headache Society (2004)
There are some interesting considerations about the case of our patient. The apparent innocuous presentation of the headache, initially responsive to analgesic treatment, with a normal clinical examination represents a diagnostic puzzle. In our case only the abrupt worsening of the cranial pain advised for further investigations and CT head scan.

Computed tomography without contrast remains the technique of choice for screening patients with non specific clinical presentation, but it not is sensitive enough to rule out CVT because in 10-30% of cases the findings on either un-enhanced or contrast-enhanced CT are negative.

Although CVT can present a vast array of clinical presentations, headache is present in more than 80% of patients and can be the sole presenting complaint of CVT. Therefore in a patient with headache without a background setting one can be missing diagnosis of CVT or need for a high index of suspicion. We think that in patients with dubious or unexplained headache, especially in association with risk factors like oral contraceptives or systemic lupus erythematosus, the use of MRI imaging is mandatory.

In a perspective series of 123 patients, Cumurciuc et al. have found 17 patients (14%) with headache as the only manifestation of CVT. A remarkable feature of this case was the unusually high frequency (85%) of lateral sinus involvement, and in general the prognosis was good.

Another more recent study reported 28 patients out 62 with isolated headache and lateral sinus thrombosis. By analogy with our case this allow us to speculate about the importance of the site of thrombosis: in contrast, for example, headache in superior sagittal sinus thrombosis is more frequently associated with intracranial hypertension, focal deficits, or consciousness disorders.

In the cases with only headache and no parenchymal lesions, another aspect still unexplained is if patients would have a spontaneous recovery or whether anticoagulant therapy play an essential role. Again, it is interesting to know whether these patients generally should be treated with iv heparin, and how long anticoagulation should be sustained. The underlying pathogenesis is essential in that cases of CVT “no complicated”. For example in some patients spontaneous intracranial hypotension is reported as cause of CVT, and treatment with epidural blood patch seems enough to re-canalisate occluded venous sinus. In our case oral contraceptive and the venous infarction are certain indication for a anticoagulant therapy.

Headache in our patient is described as progressive until a “top” of severity, tightening in quality, diffuse and with nausea. In a recent prospective study (11) of patients diagnosed with CVT, this pattern of headache is the most frequently reported (80%). Nonetheless, the Authors have not found a statistically significant association between the characteristics of headache and extension of CVT or involved sinus; neither the pattern of headache is suggestive for diagnosis. Further, time from onset to diagnosis was significantly delayed in these patients presenting only with headache, when comparing to those that presented with other symptoms and signs.

In conclusion CVT, although relatively rare, should be included in the differential diagnosis of unexplained headache. In fact, isolated headache can be the only clinical sign of CVT with no evidence of increased intracranial pressure or focal neurological signs, and as demonstrated in our case report, may be the only presenting symptom. The mechanism of isolated headache in CVT is unknown, but considering the variability of presentation, ranging from a simple progressive pain to real “thunderclap”, the key to diagnosis lies in neuroimaging.

References


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