Chronic, Recurrent Headache in Cerebral Venous Sinus Thrombosis

She-Qing Zhang,¹ Xiao-Long Ma,² Yang-Tai Guan¹ & Hui-Min Zheng¹

¹Department of Neurology, Changhai Hospital, Shanghai 200433, China

²Department of Neuroradiology, Changhai Hospital, Shanghai 200433, China

Keywords

Cerebral venous sinus thrombosis; Chronic; MR imaging; Recurrent headache; Stroke.

Correspondence

She-Qing Zhang, M.D., Department of Neurology, Changhai Hospital, No. 168, Changhai Road, Shanghai 200433, China.
Tel.: +86 21 8187 6719;
Fax: +86 21 5523 7129;
E-mail: sheqingzhang@126.com

doi: 10.1111/j.1755-5949.2011.00270.x

Headache is the most frequent (80–90%) and often the presenting symptom of cerebral venous sinus thrombosis (CVST) [1]. It has no specific features in the mode of onset, quality of pain, extent and duration, and can be the sole clinical manifestation at the onset and during the course [2]. Headache attributed to CVST is sometimes misleading, mimicking migraine, cluster-like headache, even thunderclap headache [3–5]. Here, we report two patients with chronic, recurrent, atypical headache, who were finally diagnosed as CVST and underwent a benign course after anticoagulation treatment.

Case 1. A 48-year-old Chinese woman complained of headache in the last 2 years before admission. The headache was diffuse and dull, lasting several days to a week, not accompanied by fever, nausea, and vomiting, then it could resolve spontaneously or after taking analgesic drug. However, the headache had never been really cured. After a few days of headache free, it may recur. And with the time passing by, the headache became increasingly severe and longer in duration. CT scan was performed for several times, but no obvious abnormality was found. During the past 6 months, she even experienced several attacks of thunderclap headache with nausea and vomiting, which was aggravated by standing upright. This drove her to our emergency department. She did not experience visual problem during the course; and she did not have headache attack 2 years before, with no history of oral contraceptive pills.

Neurological examination revealed no apparent abnormality except for the papilledema. Subsequent lumbar puncture showed opening pressure 30 cm, but the cerebral spinal fluid was normal

in components. Routine haematological results, including cancer markers, PANCA, CANCA, ESR, antiphospholipid antibody, were normal or negative. Her blood D-dimer was 0.71 $\mu\text{g/mL}$ (normal range $<0.5 \mu\text{g/mL}$), and the blood homocysteine level was 85.10 nmol/mL (normal range 5–15 nmol/mL). MRI showed normal brain parenchyma, except for the delta sign in the superior sagittal sinus on T2 image, and MR angiography showed extensive CVST with filling defects in the superior sagittal sinus, with extensive cortical venous collateral formation (Figure 1). After administration of low molecular weight heparin (4000 u, hypodermic injection, bid) for 3 weeks, the headache dissipated gradually and did not recur in the next 6 months follow-up.

Case 2. This 78-year-old Chinese woman was admitted for 4 months of episodic headache. She had no prior history of hypertension. The headache was deep and localized around the left temporal bone, not accompanied by fever, nausea, and vomiting. It is episodic and pin-prick in quality at first, lasting from half an hour to 4 h each time. The symptom was mild initially, but aggravated occasionally. Two months later, the headache gradually became persistent, pulsatile, sometimes drilling, and severer at night. She even could not go into sleep in the nights before seeing the doctor. She did not experience visual problem during the course.

Neurological examination was normal. Bilateral fundascopy did not reveal papilledema. A noncontrast brain CT showed no abnormal signs. Routine haematological results were normal or negative. Her blood D-dimer was 0.91 $\mu\text{g/mL}$ (normal range $<0.5 \mu\text{g/mL}$), and blood fibrin degradation products (FDP) was 3.96 $\mu\text{g/mL}$ (normal range $<6 \mu\text{g/mL}$). The patient refused

Figure 1 MR angiography revealed (A) superior sagittal sinus thrombosis with cortical venous collateral formation (indicated by arrowhead) and (B) delta sign in T2 MR imaging (indicated by arrow).

lumbar puncture throughout the course. MRI and MR angiography showed CVST in the left transverse sinus with filling defects on GD-DTPA enhanced imaging (Figure 2). After administration of low molecular weight heparin (4000 u, hypodermic injection,

bid) for 3 weeks, the headache gradually disappeared and did not recur in the next 6 months follow-up.

Headache occurs in about 80–90% of patients with CVST. Agostoni E. reviewed 49 patients and found that the onset was acute

Figure 2 MR angiography revealed (A) left lateral sinus thrombosis (indicated by arrow) and (B) filling defect in left lateral sinus and superior sagittal sinus on GDDTPA enhanced MR imaging (indicated by arrowhead).

(<48 h) in 45% of cases, subacute (48 h–1 month) in 40% and chronic (>1 month) in 11% [1].

The International Headache Society (IHS) Classification defined criteria of the diagnosis of headache attributed to CVST: (A) Any new headache (with or without other neurological signs) fulfilling criteria C and D. (B) Neuroimaging evidence of CVST. (C) Headache and other neurological signs (if present) develop in close temporal relation to CVST. (D) Headache resolves within 1 month after appropriate treatment [6]. How to explain the “new onset” in our patients?

The mechanism of headache in CVST is thought to be caused by local involvement of the pain-sensitive fibers of the dura mater by distension of the sinus wall, cerebral venous dilation or leakage of blood on the surface of the brain, as well as intracranial hypertension. The mechanism of this chronic, recurrent headache in our patients may reflect the course of thrombosis and spontaneous thrombolysis at the early stage of the disease. As this mild type undergoes a chronic course, there is enough time for the collateral veins in the brain to compensate. Thus, these patients only exhibit isolated intracranial hypertension, even isolated headache

[2,7,8]. As the disease progresses, the compensatory mechanism becomes no longer sufficient, and the headache becomes increasingly frequent and severe.

Are there any other causes that can explain this chronic, recurrent headache in our patients? This was impossible. In case 1, the headache is not in accordance with migraine and tension type headache, or with cluster headache. In case 2, the headache is initially episodic and pin-prick in nature and gradually became persistent, pulsatile, sometimes drilling, and severer at night. This profile in the older often raises the possibility of giant cell arteritis, but the patient had no temporal tenderness, and no elevated ESR. These features can exclude the diagnosis of giant cell arteritis. Moreover, the headache in our patients resolved in 1 month after appropriate treatment, and it did not recur ever since.

Our observations have important practical consequences. CVST should be considered when chronic, recurrent headache becomes increasingly frequent and severe. As the typical signs of CVST (delta sign and cord sign) are seldom found in plain CT scan, the best investigation is MRI and MR angiography [9,10].

References

1. Agostoni E. Headache in cerebral venous thrombosis. *Neurol Sci* 2004;**25**:S206–S210.
2. Cumurciuc R, Crassard I, Sarov M, Valade D, Bousser MG. Headache as the only neurological sign of cerebral venous thrombosis: A series of 17 cases. *JNNP* 2005;**76**: 1084–1087.
3. Slooter AJC, Ramos LMP, Kappelle LJ. Migraine-like headache as the presenting symptom of cerebral venous sinus thrombosis. *J Neurol* 2002;**249**:775–776.
4. Rodriguez S, Calleja S, Moris G. Cluster-like headache heralding cerebral venous thrombosis. *Cephalalgia* 2008;**28**:906–907.
5. Bruijin SFTM, Stam J, Kappelle LJ. Thunderclap headache as first symptom of cerebral venous sinus thrombosis. *Lancet* 1996;**348**:1623–1625.
6. Classification and diagnostic criteria for headache disorder, cranial neuralgias and facial pain of the Headache Classification Committee of the IHS. *Cephalalgia* 2004;**24** (suppl 1N):9–160.
7. Quattrone A, Bono F, Oliveri RL, et al. Cerebral venous thrombosis and isolated intracranial hypertension without papilledema in CDH. *Neurology* 2001;**57**:31–36.
8. Biousse V, Ameri A, Bousser M-G. Isolated intracranial hypertension as the only sign of cerebral venous thrombosis. *Neurology* 1999;**53**:1537–1542.
9. Virapongse C, Cazenave C, Quisling R, Sarwar M, Hunter S. The empty delta sign: Frequency and significance in 76 cases of dural sinus thrombosis. *Radiology* 1987;**162**:779–785.
10. Bousser M-G, Jose MF. Cerebral venous thrombosis: An update. *Lancet Neurol* 2007;**6**:162–70.