CASE REPORT

Headache in a young male: the clot thickens

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Charles Philip Miller, charles.miller04@imperial.ac.uk**SUMMARY**

Cerebral venous sinus thrombosis is a rare but serious cause of headache. In this report, we present a young man with poorly controlled Crohn's disease who presented with a 2-week history of headache and fluctuating left-sided sensory and motor symptoms. CT demonstrated changes consistent with either a subarachnoid haemorrhage or venous sinus thrombosis. The ensuing magnetic resonance venogram confirmed superior sagittal venous sinus thrombosis and an infarct of his right superior frontal lobe. The patient was started on low-molecular weight heparin and steroids. He required multi-disciplinary input from the stroke physicians, neurologists, gastroenterologists, dieticians and physiotherapists. He made a full neurological recovery and is now on long-term azathioprine. The purpose of this report is to highlight the consideration of venous sinus thrombosis in the diagnosis of headache and as a rare extraintestinal complication of Crohn's disease.

BACKGROUND

Headache is a common presentation and is encountered in all areas of medicine. Cerebral venous sinus thrombosis (CVST) is a rare condition^{1 2} that can present with headache. It often presents non-specifically with wide-ranging variable symptoms and signs.³ Without careful consideration of CVST misdiagnosis can occur without correct imaging.⁴ Not only does it represent a diagnostic dilemma but can have delayed diagnosis⁵ and hospital admission.⁶ There are many associated risk factors including head trauma, malignancy, puerperium and inflammatory bowel disease.^{1 7} It is an important diagnosis to make and may result in increased morbidity if diagnosis is delayed at least in certain subsets.⁸ Furthermore it is a treatable condition, both with anticoagulants and, potentially, thrombolysis.⁹ Crohn's disease affects many organ systems and suboptimal control can result in numerous complications including CVST.^{10 11}

CASE PRESENTATION

A 27-year-old man presented to our accident and emergency department with a 10-day history of headache and fluctuating mild left-sided motor and sensory symptoms.

He had a medical history of ileocaecal Crohn's disease diagnosed within the last year for which he had taken a prolonged course of steroids but had subsequently been lost to follow-up.

He had been experiencing an intermittent headache in both the vertex and retro-orbital regions. He had vomited twice in the last week with no

obvious precipitant, though had not experienced visual disturbance, meningism or photophobia.

Subsequently, he had developed sensory symptoms with paraesthesia in his left hand. This had fluctuated and at times affected the whole of his left arm and torso. He had also experienced left-sided heaviness. At its worst he was unable to move his left arm or get out of the car, though this was short-lived. There was no major systemic disturbance of note except that he had an increased bowel frequency. His regular medications included mesalazine. He had no relevant family history.

On examination his observations were within normal limits and he was fully orientated. There were no positive neurological findings except slight sensory numbness on his left torso and left arm in a non-dermatomal distribution.

Given his neurological symptoms and his young age there was significant diagnostic uncertainty although the possibility of a stroke was being considered. A discussion with the on-call stroke consultant was sought to help determine what imaging would be most appropriate. It was felt a stroke was unlikely in his age group but CT imaging would help rule out intracerebral haemorrhage. CVST was not initially considered as a diagnosis.

INVESTIGATIONS

Blood tests revealed C reactive protein 49 and albumin 13 g/l. He was initially sent for non-contrast CT imaging (see figure 1). This had initially been reported as suspicious for subarachnoid haemorrhage, however, on discussion with the neurosurgical team it was felt that a CVST ought to be considered in light of fluctuating symptoms and radiological findings. Figure 1A shows two findings suggestive of venous sinus thrombosis, the positive delta sign and a thrombosed anterior cortical vein. Of note there is an area suggestive of infarction in the right frontal lobe (figure 1B).

He subsequently underwent MRI head and MR venography to further establish the pathology (figure 2). This demonstrates a filling defect in the superior sagittal sinus suggestive of thrombosis. MRI head also confirmed a right frontal lobe infarction.

DIFFERENTIAL DIAGNOSIS

This was particularly tricky given he was in such a young age group with soft neurology at presentation. A vascular cause including cerebrovascular accident was considered. Other differentials being considered included space occupying lesions, migraine with aura and meningitis. Another possibility included seizure activity as a cause for his symptoms given the transient nature of his neurology.

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Figure 1 (A) Selected axial image from non-contrast CT of the brain on brain windows. Red arrow: positive delta sign. Green arrow: thrombosed anterior cortical vein. (B) Selected axial image from non-contrast CT of the brain on stroke windows. This demonstrates a wedge shape area of low attenuation in the right frontal lobe consistent with infarction.

TREATMENT

He was initially treated with low-molecular weight heparin and rehydration. The gastroenterologists reviewed him and in view of his low body mass index (BMI), low-serum albumin (13 g/l) and increased bowel frequency, steroid therapy and dietetic support were started. He was anticoagulated with warfarin for a provisional duration of 6 months.

OUTCOME AND FOLLOW-UP

He had a good clinical response to anticoagulation with almost full neurological recovery. Steroid therapy was tapered down and azathioprine was started. His Crohn's disease has become much less symptomatic and he has put on 10 kg weight. Most importantly he feels that his quality of life has remarkably improved.

DISCUSSION

Headaches are a common presentation in all clinical settings. When associated with neurology there is a wide differential including infection, cerebrovascular accidents and malignancy.

CVST is rare and can present subtly, sometimes with headache as the only symptom.¹² It would usually be low down on a physician's differential list. In this case it was not initially considered until the neurosurgical team reviewed the imaging. Fortunately there were clues from the CT imaging but this is not always the case and they can be normal.¹ Furthermore, It must also be remembered CT imaging and MRI alone without venography can be insufficient for diagnosis.^{4 13 14}

The aetiology is similar to that of venous thrombosis in other body areas and is often described with reference to Virchow's triad. Multiple risk factors have been identified including hypercoagulable states such as inflammatory bowel disease, thrombophilias and pregnancy.^{7 13} Other temporary conditions including dehydration, infection and head trauma are also associated.^{7 13} Presence of these risk factors should raise the clinician's index of suspicion.

Inflammatory bowel disease is a precipitant of CVST 2–3% in one literature review.¹¹ Broadly speaking it is ulcerative colitis that is typically associated with thrombotic complications. Interestingly this study showed that the proportion of CVST in inflammatory bowel disease was attributed to Crohn's disease 75% of the time.¹¹

In this case it would appear that poorly controlled Crohn's disease, as evidenced by a low BMI, low-serum albumin and increased bowel frequency, was a contributing aetiological factor. Interestingly he was also predisposed being heterozygous for factor V Leiden.

Symptoms can be subtle and variable and are dictated by the anatomical location. These include symptoms of raised intracranial pressure and focal neurology. The fluctuation of neurology, as in this case can be suggestive of venous thrombosis. Patients can also present with symptoms relating to complications of venous thrombosis including infarction, haemorrhage or seizure.⁵

Various clinical signs can be apparent dependent on anatomical location. These signs include evidence of papilloedema and focal neurology of the central and peripheral nervous system.¹

The key investigation is radiological imaging. CT imaging can be diagnostic but MRI and MR venography is the current diagnostic modality of choice.¹³ The primary MRI techniques used to detect CVST are time-of-flight MR venography (TOF-MRV) and contrast-enhanced MR venography. Both techniques have well-recognised limitations. Highlighting just one example, TOF-MRV may give false positive results if there is venous flow in the plane of image acquisition, leading to signal dropout.^{15 16} CT venography also plays a role as it remains widely available and the imaging technique is considerably quicker.

There are various factors which produce imaging interpretation pitfalls irrespective of the imaging modality and these include variants or normal anatomy, hypoplasia/atresia of venous sinuses and arachnoid granulations, all of which may give rise to false positive results.¹⁶

It can pose management dilemmas as with this patient his neurology subsequently fluctuated. He underwent further inpatient head imaging twice. A clinician can never be certain if complications such as haemorrhage or further infarction have occurred. This can pose significant management difficulties especially in a patient who has been anticoagulated.

Figure 2 Selected maximal intensity projection images from contrast-enhanced MRI venogram. Sagittal and coronal projections: this demonstrates a complete lack of filling in the superior sagittal vein.

Learning points

- ▶ Cerebral venous sinus thrombosis (CVST) is a complication of Crohn's disease.
- ▶ CVST is a rare cause of headache and should be considered in the differential especially in the presence of risk factors.
- ▶ CVST can present variably and subtly.
- ▶ A normal CT head does not exclude the diagnosis of CVST.
- ▶ MRI and MR venogram are the gold standards for diagnosis.
- ▶ The underlying precipitant must be considered and treated.

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