Reminder of important clinical lesson

Intracranial hypertension: was it really idiopathic?

Manon Wyn Williams,¹ Mandagere Vishwanath²¹University of Manchester, Manchester, UK;²Manchester Royal Eye Hospital, Manchester, UK**Correspondence to** Manon Wyn Williams, manon-wyn@hotmail.co.uk**Summary**

A healthy 44-year-old slim female presented with a constant, non-specific dull headache of subacute onset. The examination showed papilloedema but no other problem. On further questioning she revealed transient visual obscurations and pulsatile tinnitus. The CT scan of the brain was normal, the lumbar puncture showed increased pressure but normal cerebrospinal fluid, and the MR scan and MR venogram was reported showing a small area of developmental venous anomaly in the left parietal region but no venous thrombosis. Benign intracranial hypertension (BIH) was diagnosed but later in the course further tests were done as the patient was atypical for BIH, which revealed essential thrombocythaemia. A CT venogram was arranged which showed partly recanalised chronic sigmoid sinus thrombosis with dilation of collaterals. The patient was treated with acetazolamide, warfarin and hydroxycarbamide. The headache resolved quickly and the papilloedema reduced significantly over 6 months.

BACKGROUND

This case highlights the importance of thorough investigation in atypical cases, and demonstrates the superiority of CT venogram in detecting cerebral sinus thrombosis.

CASE PRESENTATION

A healthy 44-year-old slim female presented to the medical unit with a subacute onset generalised, constant, dull headache of 6 months duration. The headache was not associated with any nausea, vomiting, photo or phonophobia and not altered by posture or valsalva manoeuvre. Apart from analgesics she was on no medication. The examination revealed bilateral swollen optic discs but no other deficits. On specific questioning she revealed visual blur of a few seconds on standing up which she could not localise, and hearing a pulsatile noise in the ears sounding like a 'whoosh' on lying down.

INVESTIGATIONS

Her blood pressure and CT scan of the brain were normal. The lumbar puncture showed cerebrospinal fluid (CSF) opening pressure of 27 cm of H₂O and normal CSF constituents. Further neuroimaging by MR scan and MR venogram (figure 1), were reported as showing a small area of developmental venous anomaly in the left parietal region but no venous thrombosis. A diagnosis of benign intracranial hypertension (BIH) was made and treatment started with acetazolamide. She was referred to the neuro-ophthalmology unit of another hospital for long-term monitoring. During her review at the neuro-ophthalmology clinic it was felt that although she fitted with the clinical criteria for BIH her age and slender build were atypical, and further test were arranged, which revealed an elevated platelet count of $615 \times 10^9/L$. Bone marrow studies and a positive Jack 2 enzyme assay led to the diagnosis of essential thrombocythaemia (ET). In the light of this finding the

scans were reviewed again and a CT venogram (figure 2) was performed which showed chronic, partly recanalised sigmoid sinus thrombosis with dilation of collaterals. The visual fields did not show any abnormality.

DIFFERENTIAL DIAGNOSIS

- ▶ Optic disc drusen related pseudopapilloedema.
- ▶ Malignant hypertension.
- ▶ Sequential or bilateral optic neuropathy.

TREATMENT

Acetazolamide was continued but warfarin and hydroxycarbamide were added once the diagnosis of ET was established.

OUTCOME AND FOLLOW-UP

Her headaches resolved soon after starting acetazolamide, the disc swelling reduced significantly by 6 months. The warfarin and acetazolamide were stopped after 6 months and the hydroxycarbamide was stopped 2 months later. She remains asymptomatic and under review.

DISCUSSION

Headaches are the most common medical problem although not all patients with headache seek medical attention. The majority of the headaches are benign in nature. Headaches can be broadly grouped in to primary headache syndromes for example, migraine, cluster headache etc and secondary headaches. Secondary headaches are caused by identifiable pathology and hence need investigating. The critical tool in proper diagnosis of the headache is a thorough history. It is important to remember that occasionally secondary headaches may develop in a patient with well known primary headache syndrome and some secondary headaches may resemble primary headaches. The warning signs of a serious pathology are: new onset headache in a patient who

Figure 1 Axial MR venogram showing reduced circulation in the left transverse sinus. The radiologist reported that this may be a normal variant accounted for by dominance of circulation on the right side or a congenital anomaly as a clear venous thrombosis was not present.

has never suffered headache, change in the pattern of an existing headache, severe headache, change in personality or headaches associated with neurological signs, fever or trauma.

Bilateral disc swelling, especially if associated with headache or neurological deficits is presumed to be due to raised intracranial pressure (ICP) unless proved otherwise. Malignant hypertension may present in a similar way with headache and swollen optic discs. Swollen optic discs in an asymptomatic patient is often detected by optometrists during routine check ups. Pseudo papilloedema due to buried optic disc drusen can be the cause in these patients. However, occasionally, some of these patients may have unrelated headache causing diagnostic confusion. Ultrasound B-scan can show the bright echo of calcified drusen. Drusen can occasionally coexist with papilloedema. Hence any patient with swollen discs and symptoms of raised ICP need further investigation even if the ultrasound scan shows drusen.

The symptoms of raised ICP are: generalised headaches, worse in the morning and on valsalva manoeuvre and blur of sight lasting no more than a few seconds and noticed on sitting up from a recumbent position. Occasionally, diplopia is present if there is VI nerve palsy secondary to raised

ICP. Pulsatile tinnitus is a symptom usually revealed by the patient only on specific questioning. This feature is most often seen in BIH. Raised ICP occurs due to obstruction to the drainage of CSF by pressure from tumours, adhesions secondary to infective or inflammatory pachymeningitis or cerebral venous thrombosis (CVT). If the above causes are not seen BIH is diagnosed. The clinical criteria to diagnose BIH are elevated ICP, normal CSF constituents and normal neuroimaging. BIH commonly occurs in young, obese females. Vitamin A metabolism abnormality has been implicated in the condition.

After ruling out malignant hypertension, all patients with symptoms and signs of raised ICP should have neuroimaging. CT scan is the usual first choice due to easy availability and low cost. This will help to rule out any space occupying lesion (SOL) and hydrocephalus, and sometimes identify CVT. Lumbar puncture can be safely done once SOL has been ruled out for measuring CSF pressure and analysis. If it shows normal ICP, then ophthalmological referral to rule out drusen needs to be done and other causes of headache explored. If it shows raised ICP but normal CSF, additional imaging is required to rule out CVT.

CVT was first described by Ribes in 1825.¹ It is an uncommon condition affecting 3–4 cases/million/year

Figure 2 CT venogram showing the thrombus and dilation of collateral vessels.

with a mean age of 37–38, though any age may be affected.² The presentation is varied and depends on several factors, such as the site and extent of the thrombosis, the rate of propagation of the occlusion, the availability of collateral channels, the age of the patient and the nature of the underlying disease. The superior sagittal sinus is commonly affected, followed by lateral sinuses and cavernous sinus.³ The onset of symptoms is acute in 30%, subacute in 50% and chronic in 20% of the cases, an acute onset is common with infective and obstetric causes where as inflammatory and coagulation disorders tend to have a subacute or chronic onset.⁴

Headache is the most frequent and starting symptom, occurring in over 80% of patients.^{3–4} The headache can be of any grade of severity, diffuse or localised and chronic or intermittent. It is generally associated with other neurological signs such as papilloedema, focal deficits, seizures, disorders of consciousness or cranial nerve palsies. Occasionally though the headache may be the only presenting feature,⁵ the causes of CVT include infections, pregnancy, puerperium and oral contraceptives, dehydration, nephrotic syndrome, systemic malignancy, inherited thrombophilia and acquired thrombotic states, anaemia, polycythaemia, thrombocythaemia, vasculitis and head trauma. Despite extensive investigations no cause can be found in 20–30% of cases.²

The diagnosis of CVT is by neuroimaging. CT scan without contrast shows brain parenchymal changes secondary to CVT and some changes within the sinuses, where as contrast CT scan can show the low density clot surrounded by enhanced collateral veins, the so called ‘empty delta’ sign. However in up to 30% of cases the CT is normal, particularly in patients with isolated intracranial hypertension.⁴ MR scan is better than CT scan but can be false negative in very early or late in the course of the disease. MR scan can also show false positive results in slowed circulation without actual thrombosis. MR venography helps to overcome these problems but is prone to artefacts. CT venogram on the other hand is less prone to these problems. Ozsvath *et al*,⁷ compared CT venography with MR venography in the assessment of dural sinus stenosis and the detection of cerebral veins and dural sinuses and concluded that CT venography was a superior modality. The CT venography was better at identifying smaller cerebral veins, with slower flow and sinuses and it was found to be at least equivalent to MR venography at diagnosing dural sinus thrombosis.^{6–7}

Treatment of CVT depends on the underlying cause and its manifestations, and prevention of further formation and propagation of the thrombus. Heparin is the first line agent and its advantages have been proved by randomised trials.^{8–9}

Our patient presented to the medical unit of another hospital with a subacute onset of a generalised headache, papilloedema and other symptoms related to increased ICP. The patient was investigated first with a CT of the brain which revealed no abnormalities, and the MR scan and MR venogram did not reveal a thrombus. A diagnosis of BIH was made as she fitted with the criteria. However, during neuro-ophthalmological review further investigations were carried out as she was felt to be slightly atypical. A diagnosis of ET was revealed. The blood tests which revealed thrombocythaemia may have been done at her initial visit but as she was seen in a different hospital this information was not available. Then after the diagnosis of ET, the scans were reviewed and a CT venography was arranged which revealed the sigmoid sinus thrombosis. As the thrombosis was deemed chronic anticoagulation with warfarin was started, her thrombocythaemia treated with hydroxycarbamide, and acetazolamide given to reduce the ICP.

In our case, reviewing the scans again and performing a CT venogram helped to make the correct diagnosis. This highlights the importance of reviewing the history, examination and investigation reports carefully when the presentation is atypical. In the UK, CT scan is the most commonly used emergency neuroimaging modality as it is easily accessible and cheaper compared to MR scan. MR scan and MR venogram are often carried out later to rule out CVT. Our case demonstrates the superiority of CT venogram over MR venogram and in our unit CT venogram is the preferred modality of imaging to rule out CVT. A CT venogram can be done at the same time as the initial CT scan without needing additional resources. This would avert the need to do any further scans, reducing the cost and radiation exposure. It is important to discuss and get an agreement with the radiologists if both CT scan and CT venogram are to be done at the first instance. It also highlights the need for accurate and complete transfer of information when patients are looked after in different institutions.

Learning points

- This case highlights the importance of thorough investigation in atypical cases, and the need to revisit any tests or reports with expert help, when the clinical index of suspicion is high.
- This case also confirms the superiority of CT venogram over MR scan or MR venogram in detecting CVT.
- Investigation of papilloedema needs to be carried out in a systematic way, we suggest CT scan to be combined with CT venogram while investigating papilloedema.
- New onset and unremitting headache needs thorough investigation including neuroimaging.

Acknowledgements This case report has been supported by the Biomedical Research Centre of Manchester Academy of Health Sciences.

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Williams MW, Vishwanath M. Intracranial hypertension: was it really idiopathic?. *BMJ Case Reports* 2011;10.1136/bcr.08.2010.3274, date of publication

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