

CASE REPORT

Hypertensive encephalopathy mimicking cerebral vasculitis with pontine oedema, cerebellar white matter lesions and multiple cerebral infarctions

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SUMMARY

A 47-year-old man with poorly controlled hypertension presented with headaches, right-sided weakness and dysarthria. CT and MRI scans of the brain showed widespread abnormalities including significant pontine oedema, basal ganglia and corona radiata infarctions and cerebellar white matter high signal. Imaging of the intracerebral vasculature also demonstrated wall irregularities. Initially a central nervous system inflammatory disorder was thought to be the most likely diagnosis, possibly acute demyelinating encephalomyelitis or cerebral vasculitis, and the patient was treated with high-dose intravenous steroids. The diagnosis of hypertensive encephalopathy was made because (1) the patient was hypertensive and (2) the patients MRI findings resolved with antihypertensive treatment.

Blood pressure treatment was instigated from admission, and the patients symptoms improved with resolution of the radiological abnormalities.

BACKGROUND

This challenging case highlights how hypertensive encephalopathy (HE) can present with radiological features that can mimic CNS vasculitis and emphasises how important is to consider this differential diagnosis even in the presence of vascular wall irregularities, a less common feature of posterior reversible encephalopathy syndrome (PRES).

CASE PRESENTATION

A 47-year-old man presented with slurred speech and mild right-arm weakness. The week prior to admission he suffered four episodes of bifrontal, gradual onset headaches relieved by paracetamol. His wife also reported uncharacteristically irritable and aggressive behaviour for 2 weeks.

His medical history included poorly controlled arterial hypertension diagnosed 6 years earlier and angina.

He reported smoking 10–15 cigarettes/day. He denied rash, fever, fatigue, weight loss, alcohol intake and use of intravenous or other drugs.

Physical examination on admission revealed a blood pressure of 199/110 mm Hg which fell to 160/78 mm Hg a few hours after presentation.

On neurological examination, he was alert and with GCS 15/15. Cranial nerve examination revealed slow movements of the tongue with

dysarthria. There were no other cranial nerve signs. He had a minimal loss of power of the right upper limb (4/5 with pronator drift) but no weakness elsewhere. Sensory examination was normal.

INVESTIGATIONS

A CT brain was performed which showed periventricular changes consistent with previous lacunar infarcts and low attenuation in the pons.

MRI brain scan and time-of-flight MR angiography (day 1) revealed widespread abnormalities: multiple white matter and grey matter lesions, pontine oedema (figure 1 and figure 2) and arterial wall irregularities in the basilar artery and middle cerebral artery. (figure 3)

Two discrete areas of restricted diffusion were noticed in the right basal ganglia and left corona radiata in keeping with small acute lacunar type infarcts (figure 4).

Routine laboratory investigations did not reveal any significant abnormalities. In particular CRP and ESR were not raised and ANA and ANCA antibodies were negative. HIV testing was negative. C3 and C4 complement levels were normal.

DIFFERENTIAL DIAGNOSIS

Initially, an uncomplicated ischaemic stroke, perhaps of the lacunar subtype, was suspected.

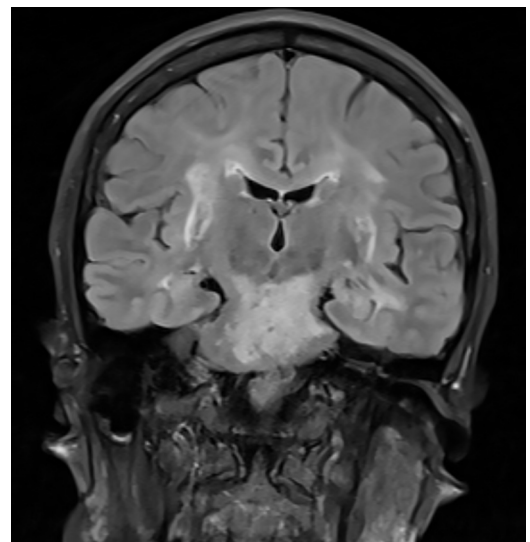


Figure 1 Pontine oedema on fluid attenuated inversion recovery MRI.



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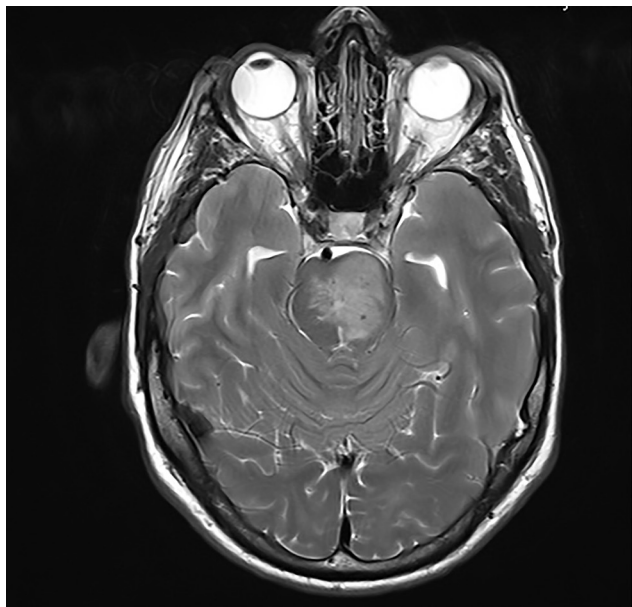


Figure 2 Pontine oedema on fluid attenuated inversion recovery MRI.

However, the asymmetrical pontine lesion and the cerebellar white matter high signal (figure 5) were felt on radiological grounds to suggest an inflammatory cause such as acute demyelinating encephalomyelitis (ADEM) or the consequences of vasculitis. MR angiography showed irregular wall appearances, especially of the basilar artery, consistent with a central nervous system vasculitis with secondary microinfarcts.

TREATMENT

The patient was started on methylprednisolone 1 g for 3 days (thereafter, it was discontinued on the basis of negative vasculitis

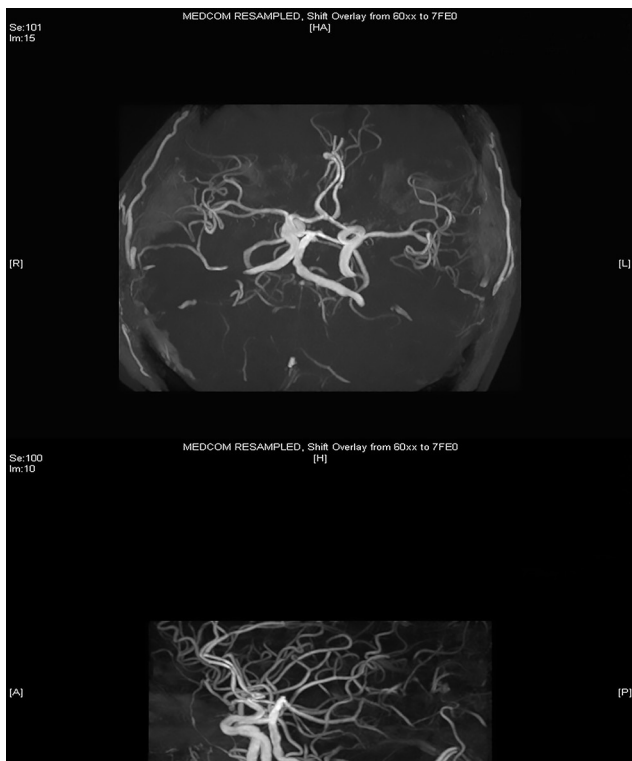


Figure 3 Time-of-flight MR angiography showing arterial wall irregularities, in particular in the basilar artery.

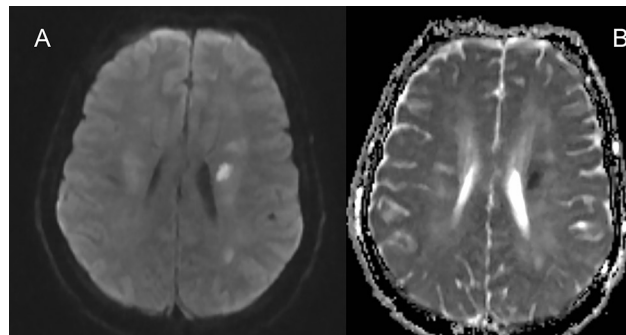


Figure 4 (A) Diffusion-weighted MRI. (B) Apparent diffusion coefficient map suggesting subacute stroke. Restricted diffusion of water appears as decreased signal on ADC and increased signal on DWI.

screening), aspirin 300 mg and glyceryl trinitrate infusion intravenously for blood pressure control. Antihypertensive treatment was then continued with oral agents including amlodipine, bisoprolol, indapamide and lisinopril.

OUTCOME AND FOLLOW-UP

The blood pressure improved over the first days following admission although he continued to have occasional readings of up to 170/100 mm Hg.

The patient was transferred to a neurology unit for further assessment and management. A lumbar puncture was considered but there were concerns about the safety of this due to the pontine oedema.

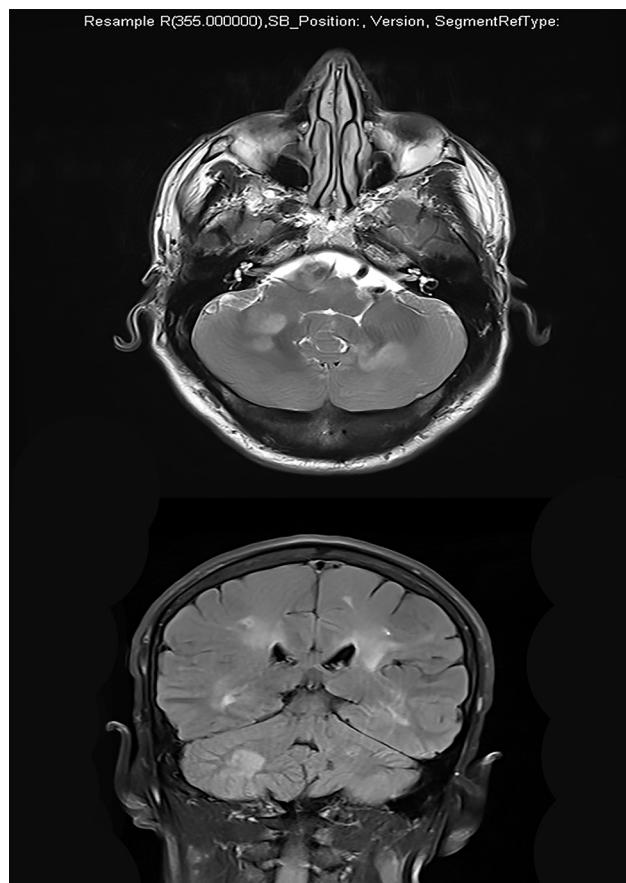


Figure 5 Cerebellar white matter high signal changes.

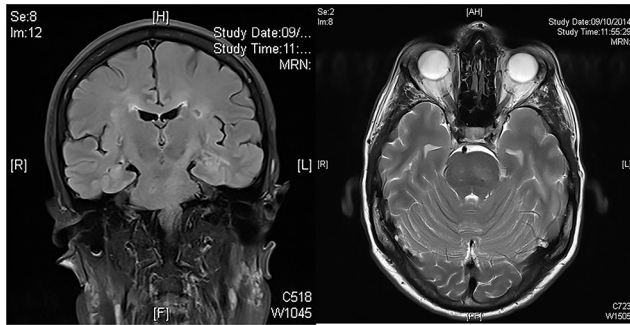


Figure 6 Follow up MRI (day 54) showing improvement of vasogenic oedema.

His imaging was repeated and the diagnosis was changed to hypertensive encephalopathy in light of clinical improvement and previous case reports describing pontine oedema in PRES.

At day 12 postadmission, the patient was discharged on antihypertensive medications, clopidogrel 75 mg and nicotine patches. Seen at 8 weeks after discharge, he continued to have mild dysarthria but was otherwise well. His blood pressure was 143/82 mm Hg.

On the third MRI (day 54), the pontine oedema had frankly improved (figure 6) although the restricted diffusion in the centrum semiovale remained.

DISCUSSION

Features of HE can be found in 16% of patients presenting with hypertensive emergencies,¹

HE is characterised by a heterogeneity of clinical presentation with frequent marked clinicoradiological dissociation (relatively mild symptoms in comparison to extensive imaging abnormalities)²⁻⁴

Common clinical manifestations of HE include headache, visual disturbance, altered mental status, seizures and focal neurological signs.³

On admission, our patient complained of multiple episodes of moderate bifrontal headache with minor neurological findings, namely slurred speech and right-arm pronator drift.

Initially, the focal neurology prompted immediate investigations for possible stroke.

Although some of the features on initial imaging supported uncomplicated ischaemic stroke in the context of hypertension, the significant pontine oedema and cerebellar white matter lesions in combination with large vessel wall irregularities led us to consider alternative more unusual causes. In retrospect, these seem to have been a combination of uncommon, but recognised, features of HE perhaps with atheromatous changes leading to vessel wall irregularity.

Treatment instigated included antihypertensive agents and intravenous methylprednisolone. We hypothesise that treatment with steroids might have favoured a rapid clinical and radiological improvement for their antioedematous and anti-inflammatory once the vasogenic oedema, caused by poorly controlled hypertension, was established.

However, the evidence of clinical and radiological improvement at follow-up after antihypertensive agents without steroids favoured a diagnosis of PRES.

Over 2 years of follow-up, the patient did not report recurrent symptoms or development of further neurological symptoms.

ADEM and cerebral vasculitis were included in our initial differential diagnoses. Unlike PRES, ADEM has a monophasic course requiring longer immunosuppressive therapy to achieve and maintain recovery; vasculitis has a higher rate of flares months or years after the first presentation, particularly when intravenous immunosuppression treatment is not followed by maintenance regimen.

An increasingly broad spectrum of clinical and radiological cerebral sequelae of severe hypertension is recognised. The most commonly reported are the the posterior circulation (often parieto-occipital) white matter changes which can be seen in the PRES.⁵ Pontine oedema is a rarer but recognised feature of HE usually occurring in the context of other typical HE radiological changes. Cases of isolated pontine oedema making the diagnosis more challenging have also been reported.^{3,6,7} Pontine oedema associated with hypertension can mimic central pontine myelinolysis. The differential diagnosis of pontine high signal changes on MRI includes also neoplastic causes such as glioma as well as inflammatory causes such as ADEM and multiple sclerosis.⁸

The clinical and radiological features of HE probably reflect loss of cerebral autoregulation leading to vasogenic oedema which appears to have a predilection for the posterior circulation.⁴ Often, though not always, such changes are reversible, at least in early stages, making prompt identification particularly important.

Learning points

- ▶ Hypertensive encephalopathy (HE) is characterised by a combination of focal and non-focal neurological symptoms usually in the setting of acute and severe hypertension.
- ▶ The posterior reversible encephalopathy syndrome may occur with normal or only moderately elevated blood pressure.
- ▶ The heterogeneity of radiological findings and the clinicoradiological dissociation can make the diagnosis even more challenging as HE can mimic other conditions.^{4,8}
- ▶ MRI findings in HE are diverse and may include lacunar infarctions, reversible white matter changes (especially in the posterior circulation) and more rarely pontine oedema.^{2,3,6,7}
- ▶ Awareness of the spectrum of clinical and radiological features of HE can help prompt treatment and avoid unnecessary investigations or treatment.

Contributors DC lead author: case selection, data collection, literature review, case writing, images selection. DH: proof reading. IB: proof reading. TW: main co-author and supervisor, draft correction and proof reading, main advisor.

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Reminder of important clinical lesson

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