Unusual presentation of more common disease/injury

Posterior reversible encephalopathy syndrome mimicking a cerebral tumour

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Posterior reversible encephalopathy syndrome (PRES) is a syndrome characterised clinically by headache, confusion, seizures, vomiting and visual disturbances with radiographic vasogenic oedema. CT imaging is typically normal, non-specific or suggestive of PRES or stroke. MRI usually shows symmetrical parietal and occipital lobe vasogenic oedema. The authors discuss a 58-year-old man presenting with right homonymous haemianopia, hypertension and ataxia. CT imaging suggested a left occipital lobe space occupying lesion (SOL). Surprisingly, subsequent contrast enhanced MRI showed characteristic bilateral vasogenic oedema in occipital and temporal lobes indicative of PRES. Hypertension, an associated predisposing factor, was present in this case and symptoms improved with antihypertensive therapy. This case highlights PRES may present with asymmetrical CT imaging findings mimicking a SOL. PRES is a potentially reversible condition with prompt treatment. MRI is essential in diagnosing both PRES and the cause of acute visual loss.

BACKGROUND

Posterior reversible encephalopathy syndrome (PRES) is a relatively recently described clinical-neuroradiographic entity, with typical symptoms of headache, confusion, seizures, vomiting and visual disturbance, including associated visual loss.^{1 2} CT imaging is frequently negative in PRES and when positive, can be difficult to differentiate from acute stroke.²

This case highlights PRES may present with asymmetric findings on CT imaging mimicking a space occupying lesion such as a tumour. Acute physicians, ophthalmologists and neurosurgeons not aware of subtle or asymmetric CT findings may lead to delays or misdiagnosis of the potentially reversible condition. Importantly, symptoms are potentially reversible with prompt treatment or by reversal of the triggering factor. Systemic hypertension is most commonly associated but other factors include immunosuppressant medication use, classically in transplant patients, eclampsia, autoimmune diseases and acute sepsis.^{3 4}

CASE PRESENTATION

A 58-year-old male manual worker presented to accident and emergency with visual loss, diplopia, ataxia and mild frontal headache. On systemic review, there was no history of nausea, vomiting, hearing disturbances, weight loss or loss of appetite. The patient had no medical history nor took any regular medication. There was a family history of asthma. He was an ex-heavy smoker and consumed 40 units of alcohol per week.

Examination revealed lateral nystagmus, diplopia and a right-sided homonymous haemianopia. Visual acuity was 6/9 and 6/12 in the right and left eye, respectively. Both upper and lower limb tone, power, coordination, reflexes and sensation were normal. Romberg's sign was negative and his gait was normal. Cardiovascular and respiratory

examination was unremarkable except for an elevated blood pressure of 170/115 mm Hg.

INVESTIGATIONS

Initial full blood count, clotting function, prostate specific antigen and urea and electrolytes were unremarkable.

Plain head CT imaging revealed the presence of a space-occupying lesion and surrounding vasogenic oedema in the left occipital lobe (figure 1). Grey matter around the oedema gave the impression of a space occupying lesion.

Figure 1 CT imaging appearance of space occupying lesion with surrounding vasogenic oedema in left occipital lobe.

Vasogenic oedema was also noted in the right temporal lobe.

Since contrast was not given for CT head imaging, referral for urgent MRI was advised. Surprisingly, MRI 5 days later revealed an abnormal signal in the sub cortical white matter in both occipital lobes, left more than right, in keeping with vasogenic oedema (figure 2).

In addition, there were similar albeit less pronounced changes in both temporal lobes (figure 3). There was no mass effect or enhancement.

Gradient echo sequences revealed widespread haemorrhagic tiny foci throughout the cerebral hemispheres and some old parenchyma haemorrhage within the left sylvian

Figure 2 T2-weighted MRI showing bilateral vasogenic oedema in occipital lobes.

Figure 3 T2-weighted MRI showing bilateral vasogenic oedema in the temporal lobes.

fissure in keeping with a history of hypertension. On post-gadolinium contrast imaging no mass lesion was identifiable (figure 4).

Diffusion-weighted imaging showed no evidence of acute infarction. The ventricles and cerebrospinal fluid spaces appeared normal. Additional CT imaging of the thorax, abdomen and pelvis with contrast were unremarkable with no evidence of malignancy or significant pathology including in the axial skeleton.

DIFFERENTIAL DIAGNOSIS

A preliminary diagnosis of a primary malignant tumour of a space occupying lesion was made on initial presentation following CT imaging. However, this was revised post MRI to PRES. Amyloid angiopathy and other coagulopathies appeared less likely possibilities. Other possible neuroimaging differential diagnoses include encephalitis, inflammatory or infectious processes, demyelinating pathology and stroke.²

TREATMENT

The patient was treated with amlodipine with symptoms promptly resolving after 24 h. The patient was discharged with full resolution of neurological symptoms.

OUTCOME AND FOLLOW-UP

On follow-up 3months later, the patient remained asymptomatic. His hypertension was controlled and MRI showed resolution of the oedema in the right temporal lobe and some persisting high signal in the left occipital lobe. The gradient echo sequence revealed extensive punctuated haemosiderin deposits, consistent with previous imaging, scattered through both hemispheres, thought likely to persist.

DISCUSSION

PRES was first described in 1996 by Hinchey *et al*,¹ formerly known as reversible posterior leukoencephalopathy

Figure 4 Contrast MRI revealed no identifiable mass lesion in the left occipital lobe.

syndrome. Radiologically, PRES displays a pattern of predominantly posterior cerebral oedema on CT and MRI⁵ but more recently involvement of other brain regions has been increasingly acknowledged.

CT imaging in PRES has been described as frequently normal, and when positive, as similar to acute stroke. Sparing of the calcarine and paramedian occipital lobe structures is a distinguishing feature of PRES from bilateral infarction of the posterior cerebral artery.¹

However, other series have described the majority (45%) of CT imaging as consistent with PRES.³ CT imaging was non-specific in 33% and normal in 22% of cases. Bartynski noted that when MRI was performed within 1 day of initial CT imaging, approximately two-thirds of scans were normal or non-specific.³ Interestingly, this case demonstrates that on non-contrast CT imaging PRES may mimic a space occupying lesion and prompt referral for neurosurgical review.

On MRI, PRES can display three primary patterns identifiable in the cerebral hemispheres; holohemispheric watershed, superior frontal sulcus or dominant parietal-occipital patterns.³ Partial or asymmetric expression of these primary patterns was found in approximately one quarter of cases. Atypical distributions other than symmetric parieto-occipito oedema, including a unilateral variant, and manifestations such as enhancement, restricted diffusion and haemorrhage are also recognised.⁶

The vast majority (94%) of patients improve clinically, with 58% showing evidence of reversal of the overall PRES pattern, including vasogenic oedema, on follow-up imaging studies.³ Residual areas of encephalomalacia remained apparent as focal areas of haemorrhage or restricted diffusion.

MRI findings are typically negative or non-specific when performed on the same day or 1 day after the initial CT imaging and toxic event, but diagnostic of PRES when MRI was obtained 2 or more days later.² MRI was essential in making the diagnosis in this case.

Although PRES is commonly seen in systemic hypertensive states, eclampsia, immunosuppressive therapy use in organ transplantation, acute sepsis and various autoimmune conditions (Wegner granulomatosis and systemic lupus erythematosus) are also associated.^{1 3}

The cause of PRES is multifactorial and the exact mechanism is uncertain. Sudden elevations in systemic blood pressure lead to forced hyperperfusion and/or failed autoregulatory capability of the brain vasculature resulting in blood brain barrier breakdown.^{1 3 7} Both vasospasm and

hypoperfusion have been identified radiologically resembling a watershed distribution.^{3 8} Other potential causative factors include endothelial injury from the cytotoxic effects of immunosuppressive medications such as cyclosporine and endothelial dysfunction in eclampsia.^{1 3 5}

While diagnosis of PRES is reliant on clinical manifestations and typical neuroradiological findings, an awareness of the variability in the patterns is important.

Learning points

- ▶ PRES is a relatively new disease entity characterised clinically by headache, confusion, seizures, vomiting and visual disturbances and vasogenic oedema on neuroradiological imaging.
- ▶ CT imaging can be normal, non-specific or suggestive of PRES or an acute stroke.
- ▶ This case demonstrates PRES can mimic a space occupying lesion on CT imaging.
- ▶ Prompt contrast enhanced MRI is essential in diagnosing PRES and facilitating prompt treatment of any associated triggering factor.
- ▶ It is classically associated with hypertension and frequently reversible with prompt treatment.

Competing interests None.

Patient consent Obtained.

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