Posterior reversible encephalopathy syndrome: a rare cause of seizure and headache presenting to AMU

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Introduction
Posterior reversible encephalopathy syndrome (PRES) is characterized by headache, confusion, seizures and visual loss as well as radiological findings of focal reversible vasogenic oedema, best seen on magnetic resonance imaging (MRI) of the brain.1,2

Case Report
- A 73 year old patient was seen in status epilepticus. She had suffered three seizures during the night and remained post-ictal.
- New left sided weakness was apparent.
- Examination, blood tests and computed tomographic (CT) brain scan were normal at clerking.
- Blood pressure was elevated at 198/105mmHg.
- The patient had been admitted the previous day with occipital headache. She had been seen with similar symptoms 48 hours earlier and discharged following a normal CT brain.
- Our patient was undergoing palliative chemotherapy with oxaliplatin and 5-fluorouracil for metastatic caecal carcinoma.
- A third CT brain showed no interval change.
- Dramatic improvement was made in 24 hours.
- MRI exhibited cortical and subcortical deep white matter high signal changes in the occipital and parietal lobes bilaterally. Findings were consistent with PRES.
- Our patient was able to walk out of hospital 10 days later.

Clinical features
- PRES is identified through combination of clinical and radiological features.
- First described by Hinchey et al in 1996 the clinical picture manifests as headache, altered level of consciousness, visual disturbance and seizures.3
- The majority of patients present with seizures.4
- PRES is recognised in a number of conditions such as pre-eclampsia/eclampsia and organ transplantation.
- A variety of immunosuppressant and anti-neoplastic drugs have also been implicated as causative.5
- Over 20 cases of cisplatin-associated PRES have been described in the literature, more than any other chemotherapy agent.6

Pathophysiology
- The pathophysiology of PRES remains unclear.
- One theory cites deranged cerebral vascular auto-regulation due to unmanageable rise in mean arterial pressure.
- The resulting vasodilation and endothelial dysfunction leads to vasogenic oedema.
- A second theory suggests a progression of cerebral vasoconstriction, hypoperfusion and ischaemia, causing cytotoxic oedema.2
- This does not explain the reversible nature of both the clinical symptoms and changes on imaging.1,2

Imaging
- MRI is essential to diagnosis.
- Areas of focal oedema are characteristically symmetrical and appear in the parietal and occipital cerebral white matter.8
- It is common to find corresponding areas elsewhere in the brain.8
- The regions most conspicuous on (fluid attenuated inversion recovery) FLAIR sequences.
- Also typically found as areas of high-intensity signal on T2 weighted images.1,6

Bilateral cortical and right parietal high signal changes. Highlighted by arrows on FLAIR (top) and T2 (bottom) MRI imaging.

Hypertension in PRES
- Hypertension appears to play some role in PRES.
- 70-80% patients show moderate to severe hypertension.
- However a significant 20-30% patients demonstrate normal or only mildly elevated blood pressure.1,4
- This challenges the theory of deranged cerebral vessel auto-regulation causing vasogenic oedema.

Oxaliplatin and Fluorouracil related to PRES
- Femia et al reviewed the literature in February 2012.
- There are seven cases of PRES induced by chemotherapy using oxaliplatin and fluorouracil in conjunction.
- Five patients experienced complete reversal of symptoms.
- One went on to have her chemotherapy successfully reintroduced without recurrence.
- Two patients died; one of advanced metastatic disease and the second case was complicated by severe co morbidities.1
- Other publications also associate significant co-morbidities with poor outcome.1

Core messages
- Clinical suspicion of PRES is warranted for all patients presenting with seizures and a history of recent chemotherapy.
- MRI is essential to diagnosis. CT imaging may be normal.
- Acute management should be supportive, aiming to manage seizures and control any associated hypertension.1
- Long-term anti-epileptic drugs are not required. Review by neurology is recommended for the timing of such therapy.1,7

References