PRES SYNDROME: A RARE CLINICAL PRESENTATION

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PRESENTATION OF CASE

21 years old female, housewife, was wheeled in to the Emergency Department by Husband with chief complaints of seizure (3 episodes) since last 30mins. Patient was apparently alright 4 days back when she developed pain in abdomen (not associated with- vomiting, loose motions, constipation) with high grade fever with no history of chills and rigor. The intensity increased since last 2 hours. While on the way to hospital she had 2 episodes of seizure (GTCS) followed by loss of consciousness. While examining she had 1 episode of GTCS. History of skin rash all over the body since last 14 days.

General Examination

- Pulse-112/min, Regular, Good Volume
- BP-160/100 mmHg
- HGT-134 mg/dl
- SPO2-92% on RA
- RR-24/min
- Chest- Clear, AE is B/L Equal, No Adventitious Sounds
- CVS- S1 S2 M0
- P/A- Soft, BS +
- CNS-Drowsy, Disoriented, Pupils B/L 3 mm and SRTL, Plantars-Flexor response,
- GCS-8/15

DIFFERENTIAL DIAGNOSIS

Vascular

- Cerebral Venous Sinus Thrombosis
- Intracranial Haemorrhage
- Posterior Circulation Stroke
- Primary Central Nervous System Vasculitis

Non-Vascular

- Infective Encephalitis
- Autoimmune Encephalitis
- Metabolic/Toxic Encephalopathy

CLINICAL DIAGNOSIS

1. High blood pressure and seizure point toward Eclampsia in this case.
2. However, there was no history of elevated BP in the past.

PATHOLOGICAL DISCUSSION

MRI Findings

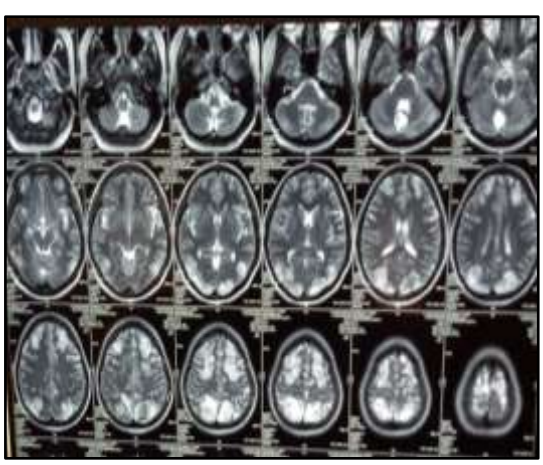

Figure 1- MRI Brain (Plain with Contrast)

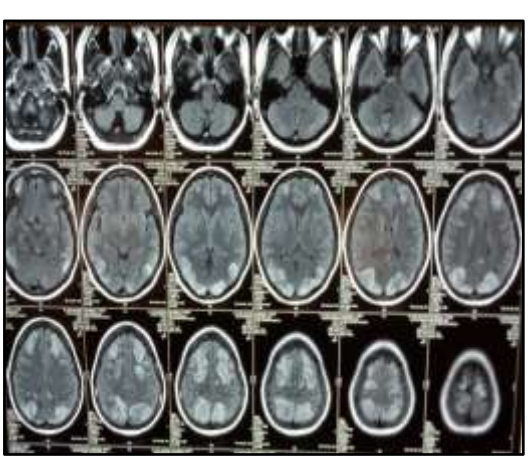

Figure 2. MRI Brain (Plain Plus Contrast)

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Symmetric areas of altered signal intensity are seen in bilateral medial cerebellar hemispheres and in posterior temporo-occipital cortexes, showing altered signal intensity, mild diffusion restriction and no significant contrast enhancement. (S/O- Reversible Encephalopathy Syndrome)

The cause of PRES remains controversial, but the most popular theory is that severe hypertension causes interruption to brain autoregulation.[1,2] Cerebral blood flow is usually regulated by dilatation and constriction of vessels to maintain adequate tissue perfusion[2] and to simultaneously avoid excessive intracerebral hypertension. Breakdown in autoregulation occurs above a mean (two thirds diastolic, plus one third systolic) arterial blood pressure of 150 - 160 mmHg; in chronic hypertension, it occurs at higher pressures.[2] Uncontrolled hypertension leads to hyperperfusion and cerebral vessel damage, resulting in interstitial extravasation of proteins and fluids, causing vasogenic oedema.[2] Irreversible damage is seen at mean arterial pressures above 200 mmHg.[2] Conditions commonly co-existing in PRES, such as chronic hypertension and atherosclerosis, are known to reduce the effectiveness of autoregulation.[2] However, the autoregulation theory does not explain why blood pressure in PRES does not usually reach the upper limit of autoregulation,[3] why PRES occurs in the absence of hypertension,[1,3,4] and why the extent of the edema is not directly related to the severity of the blood pressure.[2] Furthermore, although this theory suggests brain hyperperfusion, evidence from some positron-emission tomography studies actually demonstrates cerebral hypoperfusion.[2]

An alternative theory is that PRES is a result of a systemic inflammatory state causing endothelial dysfunction.[2] That postulate is supported by the observation that PRES is usually associated with a systemic inflammatory process such as sepsis, eclampsia, transplantation, and autoimmune disease.[2] Angiography in PRES demonstrates reversible focal and diffuse abnormalities,[2] which are thought to reflect endothelial dysfunction. When blood pressure is high, the vasoconstriction that occurs during autoregulation could exacerbate such a pre-existing inflammatory endothelial dysfunction, causing hypoxia and subsequent vasogenic edema. A mechanism of that kind would explain why control of hypertension allows for recovery. But the theory does not explain why some cases of PRES seem to occur in the absence of any inflammatory state.[4] It is clear, however, that further research is required to understand this potentially devastating but truly treatable condition.

DISCUSSION OF MANAGEMENT

Management-

No clinical trials have evaluated the management of PRES, but rapid withdrawal of the trigger appears to hasten recovery and to avoid complications: for example, aggressive blood pressure management (which may include increased ultrafiltration), withdrawal of the offending drug, or delivery in eclampsia.[1] Antiepileptic drugs should be used to treat seizures, and anaesthesia and ventilation should be instituted in generalized status epilepticus and to protect the airway in obtunded patients. Corticosteroids should theoretically improve vasogenic oedema, but there is no evidence for their use in PRES.[1]

DISCUSSION

- PRES (Posterior reversible Encephalopathy Syndrome) should be considered in patients who present with seizures, altered consciousness, visual disturbance, or headache, particularly in the context of acute hypertension.
- PRES has been associated with chronic and acute kidney disease, solid organ transplantation, and use of immunosuppressive drugs.
- Typical MRI findings include reversible, symmetrical, posterior subcortical vasogenic edema.
- If recognized and treated promptly, the rapid-onset symptoms and radiologic features usually fully resolve within days to weeks.
- A trigger is usually identifiable - most commonly, acute hypertension - but patients often have other comorbidities that may predispose them to developing PRES. Peak systolic blood pressure is usually between 170 mmHg and 190 mmHg,[1,2] but 10% - 30% of patients have normal or only mildly elevated blood pressure.[1,2] In PRES, the causes of acute hypertension are commonly acute kidney injury or eclampsia,[2] but hypertension is also reported in cases of autonomic disturbance, for example Guillain-Barré syndrome,[5,6] and after illicit drug use.[3] In a large series of cases recording the comorbidities of patients with PRES, more than half the patients had chronic hypertension, and 38% had chronic kidney disease.[2] Autoimmune diseases- including thrombotic thrombocytopenic purpura[2] and systemic lupus erythematosus - were present in 45% of the patients, and exposure to immunosuppressive drugs such as cyclosporine, tacrolimus, or chemotherapy agents was present in a similar number, particularly in the context of transplantation.[1,2,7]

Triggers and Associated Conditions

- Acute Hypertension
- Acute Kidney Injury
- Eclampsia
- Sepsis and Multi-Organ Failure
- Autoimmune Disease
- Immunosuppressive Drugs (e.g. tacrolimus, cyclosporine, chemotherapy)
- Illicit Drugs (e.g. cocaine)
- Organ Transplantation
- Chronic Hypertension
- Chronic Kidney Disease

The severity of clinical symptoms varies. For example, the visual disturbance can manifest as blurred vision, homonymous hemianopsia, or even cortical blindness. Patients may be mildly confused or agitated, but can become

comatose. Other symptoms less commonly seen include nausea, vomiting, and brainstem deficits. Seizures and status epilepticus are common, and non-convulsive status epilepticus may be more frequent than generalized status epilepticus. Non-convulsive status should be suspected in patients with prolonged states of altered consciousness and may be mistaken for postictal confusion. Signs of non-convulsive seizures include stereotypic movements such as staring, eye blinking, or head turning. Postictal confusion lasts for hours, but PRES and non-convulsive status can both persist for several days and be mistaken for psychosis, drug intoxication, or psychogenic states.

FINAL DIAGNOSIS

PRES (Posterior Reversible Encephalopathy Syndrome)

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