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POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME (PRES) IN A CHILD WITH SLE

Paediatrics

Anne Prewina Gurushekar Consultant Pediatrician, Kauvery Hospital, Tirunelveli, Tamil Nadu, India.

KEYWORDS

BACKGROUND

Posterior reversible encephalopathy syndrome (PRES) is a clinical condition that is characterized by intense headache and neurological deficits which are attributed to the vasogenic edema that occurs in the posterior cerebral cortex involving the occipital and parietal lobes.

Patients with SLE are more likely to experience headache due to vascular diseases such as cerebral venous sinus thrombosis, stroke, reversible cerebral vasoconstriction syndrome, posterior reversible encephalopathy syndrome, and vasculitis. Meningo - encephalitis, intracranial neoplasms, and intracranial hypertension or hypotension may also be a cause of headache in these patients

Case Presentation

A 13 year old girl X was diagnosed with SLE Nephritis 4 years back and was on treatment with immunosuppressive drugs (cyclophosphamide, tacrolimus), steroids and antihypertensives. She presented with a one day history of severe headache followed by unresponsiveness. On arrival she was unconscious and her BP was 190/140 mm Hg.

Evaluation

CT brain was taken urgently within the first hour which showed patchy hypodense areas in right parietal, bilateral occipito - temporal and right cerebellar regions, suggestive of PRES.

Treatment And Response

She was intubated on arrival, stabilized and her BP was gradually brought under control over the next 48 hours with IV infusions of Labetolol. She developed seizures on the first day and was given antiepileptic drugs.

Her sensorium improved and she was extubated after 48 hours. Initially , she had intermittent episodes of headache and her BP continued to fluctuate in spite of multiple anti hypertensive drugs. She also lost memory of several events which happened over the past four years and had a brief episode of delirium.

The Nephrologist, Rheumatologist and Neurologist were involved throughout in her care.

Over a period of 10 days, she gradually improved. MRI confirmed findings of PRES. Her seizures were well controlled. She no longer had headache. The memory loss of several events however remained even after discharge.

DISCUSSION

Posterior reversible encephalopathy syndrome (PRES) is a neurological condition characterized by reversible vasogenic edema on neuroimaging.

It is associated with various neurological manifestations, including headaches, vomiting, seizures, visual loss, altered mental status and focal neurological deficits. Endothelial dysfunction is a key element in PRES pathogenesis in lupus patients.

PRES mainly occurs in the setting of eclampsia, hypertension, uremia, malignancy, transplantation, autoimmune diseases and/or use of immunosuppressive drugs. This syndrome has been described in patients with systemic lupus erythematosus (SLE).

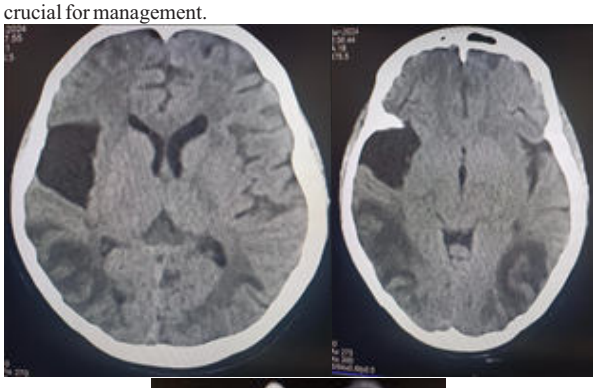
PRES can be complicated with vasculopathy, infarction or hemorrhage. Vasculopathy has been demonstrated to be a common finding in patients with SLE.

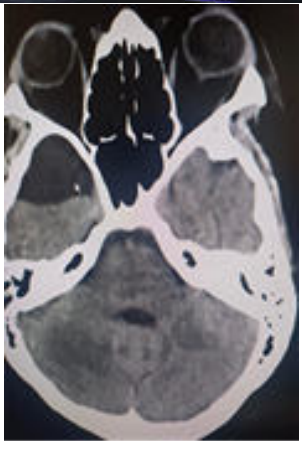
CONCLUSION

Juvenile lupus with PRES is considered an unusual neurologic manifestation triggered by multiple factors.

Early suspicion and treatment can result in a favourable outcome in these patients. Clinical and radiographic resolution of abnormalities within 1-4 weeks is typically seen. PRES is being increasingly recognized in association with autoimmune disorders.

The implications for differentiating PRES from stroke or cerebritis are crucial for management.





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