**Posterior reversible encephalopathy syndrome in Wegener’s granulomatosi: A rare occurrence**

Sir,

Granulomatosis with polyangiitis (formerly Wegener’s granulomatosis) is a rare disease with incidence of 29–65:1,00,000 in adults,[1] and even rarer in children with occurrence of 0.1 in 1,00,000.[2] We report the rare occurrence of a child with Wegener’s granulomatosis who presented with posterior reversible encephalopathy syndrome (PRES) to the ICU.

A 14-year-old male patient (known case of Wegener’s granulomatosis) was admitted to ER with multiple generalised tonic-clonic convulsions and loss of consciousness. Upon admission, patient’s blood pressure was 170/90 mmHg with a heart rate of 90/min and bilateral crepitations on auscultation in the chest (SpO2 of 90% on room air). Non-contrast computed tomography (CT) scan of the head showed bilateral symmetrical hypodensities in white matter and cortical areas of the parietal and occipital regions suggestive of PRES [Figure 1]. Blood investigations showed mild anaemia, leucocytosis, increased serum creatinine (4.9 mg/dl) and hyperkalaemia (5.8 mEq/l). The patient was already receiving oral prednisolone therapy. Tazobactam, teicoplanin, sodium valproate, amlodipine, clonidine, acetazolamide and syrup glycerol were initiated as per age-appropriate dosage. However, on the 5th day, respiratory distress with bilateral widespread crepitations developed for which trachea was intubated and patient shifted to ICU for mechanical ventilation.

In the ICU, the patient was unconscious, with uncompensated metabolic acidosis, PaO2/FiO2 ratio of 180, anaemia, leucocytosis, thrombocytopenia and deranged renal functions. A diagnosis of Wegener’s granulomatosis with pulmonary-renal affliction and PRES was made. In addition to earlier treatment, cyclophosphamide 600 mg IV single dose, methylprednisolone 1 gm IV once daily and mannitol (20%) 100 mL 12 hourly were initiated. During stay in ICU, patient’s kidney function continued to decline and renal replacement therapy was initiated. Left-sided ventilator-associated pneumonia developed by 3rd day of ICU admission. A repeat CT scan of head or MRI could not be done due to refractory hypotension not responding to fluid challenge and inotropic support (noradrenaline and dopamine infusions). On 6th day of ICU stay, patient had an arrest of cardiac activity and could not be revived.

Systemic autoimmune vasculitis is a major diagnostic and therapeutic challenge in critical care. Since our patient was a previously diagnosed case, there was no diagnostic dilemma but rarity of the disease remained a major challenge. In the available literature, reasons for ICU admission in patients with Wegener’s Granulomatosis have included a myriad of presentations, but never PRES. Wegener’s granulomatosis is rare in children and even lesser is known about PRES in these patients. Children (<18 years) may be more susceptible to PRES than adults given the narrower range of cerebral autoregulation.

PRES is a combination of radiologic findings of focal reversible vasogenic oedema in brain imaging by CT or magnetic resonance along with neurologic symptoms and signs of headache, seizures, encephalopathy and visual disturbances. The radiographic lesions in PRES typically involve the posterior parieto-occipital white matter bilaterally, but may also extend to the cortex, frontal lobes, basal ganglia and brainstem.[3] Our patient had a typical presentation of PRES with the seizures, and CT scan findings restricted to parieto-occipital areas. The commonest hypothesis for the development of PRES relates to hypertensive states, causing loss of cerebral autoregulation, resulting in compromise of...
blood-brain barrier leading to vasogenic oedema.\[^{[4]}\] In 20% cases of PRES with no apparent hypertension, other mechanisms including disruption of vascular endothelial layer, blood-brain barrier compromise or even vessel wall abnormalities have been implicated.\[^{[5]}\] Steroids and immunosuppressants have direct cytotoxic effect on the vascular endothelium, whereas vasculitis itself aggravates the risk of PRES by endothelial damage, disrupted blood-brain barrier, systemic inflammation, and treatment with immunosuppressants and/or cytotoxic agents.\[^{[6]}\] Our patient had multiple risk factors for PRES: accelerated hypertension, presence of autoimmune disease, vasculitis, renal affliction and corticosteroids.

Management of PRES remains supportive along with antihypertensives, antiepileptics and discontinuation of causative drugs. Despite known diagnoses and early initiation of treatment for PRES, the patient could not be salvaged.

To conclude, the aim of this letter is to highlight a rare disease (Wegener’s Granulomatosis) that may be present in ICU with a rare complication of encephalopathy due to PRES.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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**REFERENCES**

1. Watts RA, Al-Taiar A, Scott DG, Macgregor AJ. Prevalence and incidence of Wegener’s granulomatosis in the UK general practice research database. Arthritis Care Res 2009;15:61:1412-6.
2. Cassidy JT, Petty RE, Laxer RM, Lindsley CB. Granulomatous vasculitis, giant cell arteritis and sarcoidosis. Textbook of Pediatric Rheumatology. Philadelphia: Saunders/Elsevier, October 2010. p. 315-43.
3. Bartyinski WS, Boardman JF. Distinct imaging patterns and lesion distribution in posterior reversible encephalopathy syndrome. Am J Neuroradiol 2007;28:1320-7.
4. Staykov D, Schwab S. Posterior reversible encephalopathy syndrome. J Intensive Care Med 2012;27:11-24.
5. Rastogi A, Kaur J, Hyder R, Bhaskar B, Upadhyaya V, Rai AS. A case of post-operative posterior reversible encephalopathy syndrome in children: A preventable neurological catastrophe. Indian J Anaesth 2020;64:62-5.
6. Ni J, Zhou LX, Hao HL, Liu Q, Yao M, Li ML, et al. The clinical and radiological spectrum of posterior reversible encephalopathy syndrome: A retrospective series of 24 patients. J Neuroimaging 2011;21:219-24.

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