POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME IN ATYPICAL HEMOLYTIC UREMIC SYNDROME

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BACKGROUND

Atypical Hemolytic Uremic Syndrome (aHUS) is an autoimmune thrombotic microangiopathy caused by uncontrolled activation of the alternative complement pathway, causing hemolytic anemia, thrombocytopenia and acute renal failure. Posterior Reversible Encephalopathy Syndrome (PRES) is a clinic-radiological syndrome caused by malignant hypertension, renal disease, autoimmune disorders and endothelial dysfunction. PRES is featured by headache, seizures, visual disturbances, various degree of consciousness impairment, focal neurological signs. In this report we describe a case of PRES during aHUS.

CASE DESCRIPTION

A 30 years old man in February 2017 was admitted at Nephrology Unit of University Hospital of Bari referring nausea, emesis and asthenia since two months. Blood tests revealed anemia (6,2 g/dL), thrombocytopenia (89.000/mm^3), schistocytes on the peripheral blood film, increased serum level of Creatinine (4,5 mg/dL) and lactic dehydrogenase (569 IU/L), reduction of C3 (0,61 g/L) and haptoglobulin (<0.08 g/L). The renal biopsy detected thrombotic angiopathy; diagnosis of aHUS was proposed. Despite patient promptly underwent plasma exchange (PEX), he progressively developed malignant hypertension and after one week by PEX starting he presented a generalized tonic seizure followed by coma (Fig 1). Therapy with various intravenous antihypertensive drugs and phenytoin was rapidly started without improvement. EEG showed diffuse slow waves at all leads and focal isolated frontal spikes and sharp waves. The brain MRI revealed bilateral parietal-occipital lobe edema consistent with PRES (Fig.2-3), so the patient was treated with Sodium Nitroprusside with a prompt reduction in blood pressure and recovery of normal consciousness status. Immediately Eculizumab (weekly intravenous infusions of 900 mg for 4 weeks, followed by a maintenance dose of 1200 mg every 2 weeks) was started for the aHUS. Renal function was partially improved, although the damage is almost chronic and the patient is still periodically treated with dialysis. A detailed analysis of complement pathway and genetic screening for all currently known mutations is still ongoing.

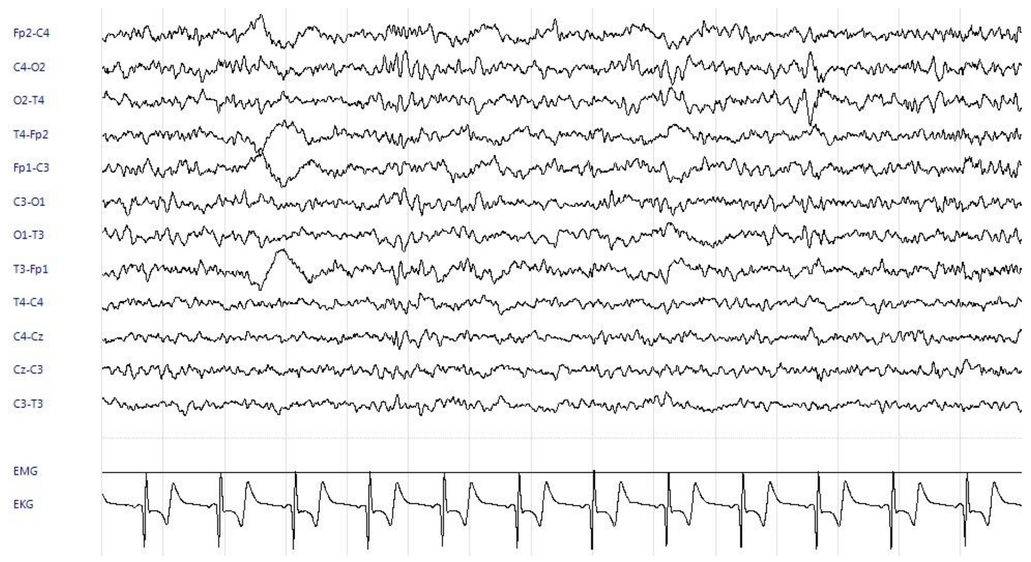

Fig.1

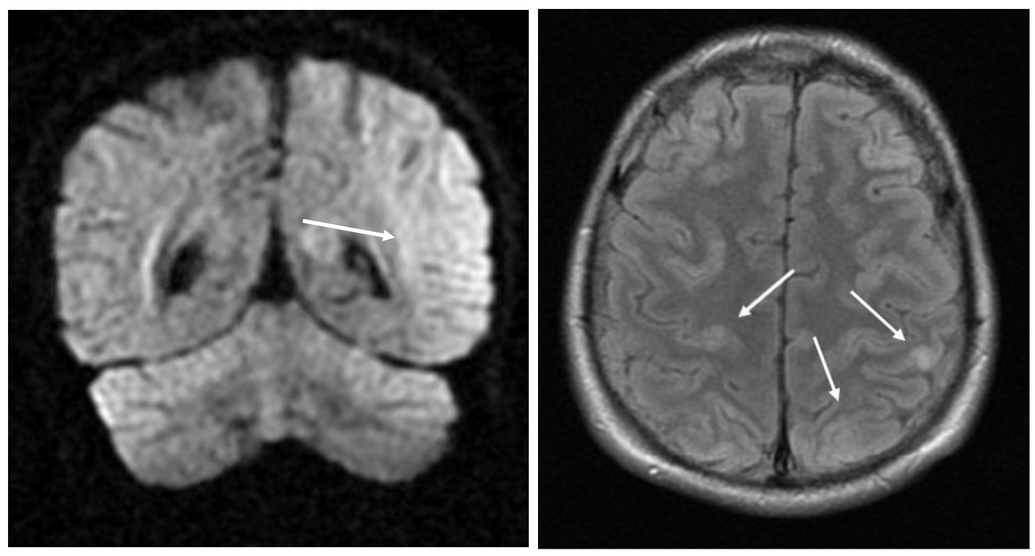

Fig.2-3

CONCLUSIONS

Few cases of this combined syndrome have been reported.

- Clinical symptoms suggestive of PRES should be considered in patients with primary nephrologic disease.
- The mainstays of PRES management include prompt control of blood pressure, treatment of seizures and removal of causative factors.
- Sodium Nitroprusside is confirmed as the best drug in treating PRES related malignant hypertension.
- Eculizumab is a "pathophysiologic-based treatment" since it binds the C5 preventing the activation of the C5a and the complex (C5b-9) and is the gold standard in aHUS tretament.
- The therapy should be promptly started to prevent chronicization of renal failure and to improve long-term prognosis.

References

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