POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME SECONDARY TO THROMBOTIC MICROANGIOPATHY OF SYSTEMIC LUPUS ERYTHEMATOSUS

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BACKGROUND

Posterior reversible encephalopathy syndrome (PRES) is a neurological complication that occurs in children and adults and progresses with headache, confused mental states, visual changes and seizures. It is a disorder that can be triggered by several causes, such as microangiopathic hemolytic anemia (thrombotic thrombocytopenic purpura and hemolytic-uremic syndrome), hypertensive crisis, renal failure and even the activity of systemic lupus erythematosus (SLE). In its pathophysiology, there is focal vasogenic edema, mainly in the occipital region, caused by the imbalance of brain self-regulation induced by the hypertensive peak or by the deposition of immune complexes to the cerebral vessels, resulting in neurological symptoms. Rarely, PRES syndrome may present with mydriasis due to secondary intracranial hypertension. Despite the tomography is suggestive, the diagnosis is classically confirmed by magnetic resonance assessment and the prognosis is usually favorable.

CASE REPORT

Female, 38 years old, with a previous diagnosis of SLE (arthritis, leukopenia, 1/640 nuclear speckled ANA, positive anti-Sm) was admitted to intensive care unit after suspension of drug treatment (hydroxychloroquine and methotrexate) with fever, hemolytic anemia

Figure 1. Brain tomography.

(elevation of reticulocytes, high levels lactate dehydrogenase, and indirect bilirubin, severe anemia (Hb 2.8 g/dL) thrombocytopenia (90,000 μ L), negative direct Coombs, decreased ADAMTS 13 activity, high creatinine (2.1 mg/dL), hemoglobinuria and normal diuresis. Additional investigation showed low complement C3 and C4 levels, positive anti-DNA, dosage of anticardiolipin, lupus anticoagulant and anti-beta2-GP I antibodies negative. Once excluded deficiency anemia, infectious causes and macrophage activation syndrome, she received methylprednisolone and immunoglobulin, showing improvement in renal function and platelets, but without improvement in hemoglobin, persisting schizocytes in peripheral blood and continued hemolysis. It was started plasmapheresis. On the seventh day in plasmapheresis, after hemoglobin improvement (8.5 g/dL), she presented mental confusion, mydriasis and decreased level of consciousness, and was submitted to mechanical ventilation and urgent brain tomography was made (Figure 1). Her blood pressure was normal. The tomography showed absence of focal expansive lesions and acute bleeding, indirect signs of intracranial hypertension (decreased cerebral sulcus and fissures), hypodense lesions in the occipital region, bilateral, compatible with syndrome PRES. Intensive care and plasmapheresis was maintained with a gradual improvement in the neurological and hematological condition.

CONCLUSION

Posterior reversible encephalopathy syndrome is a rare condition, present more frequently in lupus patients than in the general population, benign and reversible. In cases of deposition of immune complexes due to thrombotic microangiopathy, plasmapheresis is the best option of choice for treatment, being associated with intensive care for clinical improvement status.

KEYWORDS

Lupus, Posterior reversible encephalopathy, Thrombotic microangiopathy.