Abstract

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A case of thrombotic thrombocytopenic purpura (TTP) relapses in systemic lupus erythematosus (SLE): a correct management?

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Background

Thrombotic thrombocytopenic purpura (TTP) is one of the thrombotic microangiopathic (TMA) syndromes, caused by severely reduced activity of the vWF-cleaving protease ADAMTS13. In rare cases, TTP may precede the diagnosis of systemic lupus erythematosus (SLE) or occur concurrently. We describe a case of SLE-TTP relapse.

Case report

A 32-year-old female was admitted to the hospital for abdominal pain associated with episodes of vomiting and nausea. Her past medical history includes SLE onset with severe refractory TTP diagnosed about 10 years ago, who was initially treated with a combination of steroids and plasma-exchange (PEX) and then with rituximab, resulted in the clinical improvement; joint involvement, seizures and hypertension. Currently on azathioprine therapy and low dose steroids. Her emergency laboratory test showed thrombocytopenia, normal apt globin, negative Coombs's test and normal peripheral smear. She was diagnosed with thrombocytopenia in SLE due anti-platelet autoantibodies and she was initiated on steroids, namely methylprednisolone 40 mg IV twice daily and Ig 0,4 g/kg/die ev for 5 days. During hospitalization her condition continued to deteriorate. She presented purpura and fever with worsening thrombocytopenia and evidence of haemolytic anemia and schistocytes in the new peripheral smear. ADAMTS13 activity was severely reduced 1.4% (normal >67%). She was diagnosed with TTP relapse in active SLE, supported by high anti-ANA and anti-ENA titers, decreased complement C3/C4 counts, evidence of hematuria, proteinuria and pyuria in the absence of urinary tract infection. Her active SLE disease activity (SLEDAI) was >10. She started therapy with PEX, high dose steroids and caplacizumab. Azathioprine was suspended. Platelet counts and ADAMTS13 activity normalized. This treatment induced sustained remission of TTP, but not of SLE.

Conclusion

The association of TTP and SLE is rare, but is frequently fatal. Therefore, it is important to report the clinical features of patients with SLE associated with TTP elucidating the pathophysiology that underlies each condition and improving its management and therapeutic strategies. It is necessary a multidisciplinary approach and a continual follow up with Rheumatology.