Thrombotic thrombocytopenic purpura preceding systemic lupus erythematosus

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Abstract
The case of a patient admitted with thrombotic thrombocytopenic purpura nine years after developing systemic lupus erythematosus (SLE) is reported. Thrombotic thrombocytopenic purpura associated with SLE has been described on other occasions, but in most patients the diagnosis of SLE precedes that of thrombotic thrombocytopenic purpura. The unusual sequence and the chronological separation of the two diseases is emphasised.

Thrombotic thrombocytopenic purpura is a rare clinical syndrome characterised by the presence of microangiopathic haemolytic anaemia, thrombocytopenic purpura, neurological abnormalities, fever, and renal dysfunction. In most patients there is no known aetiological agent or related underlying disease. On rare occasions, thrombotic thrombocytopenic purpura has been described in association with systemic lupus erythematosus (SLE). A relation between SLE and thrombotic thrombocytopenic purpura has not been shown; however, similar clinical features may be observed in the two diseases. Chronologically, thrombotic thrombocytopenic purpura presents in patients previously diagnosed with SLE. An inverse sequence of events is rare. A patient who presented with thrombotic thrombocytopenic purpura and who then developed SLE nine years later is described in this paper.

Case report
A 30 year old woman with no relevant medical history was admitted in January 1980 with general weakness, headache, jaundice, and petechiae. Physical examination also revealed skin and mucous membrane pallor and hepatosplenomegaly.

A chest radiograph was normal. Laboratory data included severe anaemia with haemoglobin 70 g/l (normal <120 g/l), packed cell volume 0.2, abundant schistocytes, platelet count 17×10^9/l (normal 150-450×10^9/l), direct bilirubin 136.8 µmol/l (normal 6-84 µmol/l), haptoglobin 0 g/l, and reticulocyte count 250×10^9/l (normal <75×10^9/l). Coagulation tests were normal. Coombs’ and sucrose lysis tests were negative. A test for antinuclear antibodies was positive at a titre of 1/2560 with a speckled pattern; antibodies to double stranded DNA could now be seen at 139 U/l (normal <15 U/l). Anticardiolipin antibodies and lupus anticoagulant were negative. Sucrose lysis and Coombs’ tests were negative. A renal biopsy sample revealed diffuse proliferative glomerulonephritis. A diagnosis of SLE with renal involvement was made and treatment with methylprednisolone (60 mg/day) and bolus cyclophosphamide (15 mg/kg/day) was initiated. The patient improved with this treatment.

Discussion
The diagnosis of thrombotic thrombocytopenic purpura is usually based on the clinico-biological syndrome (microangiopathic haemolytic anaemia, thrombocytopenic purpura, neuro-
Thrombotic thrombocytopenic purpura (TTP) is a rare, but life-threatening, autoimmune disorder characterized by microangiopathic hemolytic anemia, thrombocytopenia, and organ dysfunction. TTP is often associated with autoantibodies against a specific adhesion molecule, the von Willebrand factor (vWF), which plays a crucial role in platelet adhesion and aggregation. The association of TTP with systemic lupus erythematosus (SLE) and other autoimmune diseases is well-documented, and this association is often characterized by specific clinical manifestations, laboratory findings, and response to treatment.

In cases of SLE, the development of TTP may be preceded by fever, renal dysfunction, and other clinical manifestations. The presence of autoantibodies against vWF is a hallmark of TTP, and their detection can help in the diagnosis of this condition. Treatment of TTP in SLE often involves plasmapheresis, steroids, and immunosuppressive agents, with varying degrees of success.

The pathophysiology of TTP in SLE is complex and involves both immune and non-immune mechanisms. The role of autoantibodies is central to the pathogenesis of TTP, and their targeting by monoclonal antibodies has shown promise in the treatment of this disorder. Further research is needed to elucidate the mechanisms underlying the development of TTP in SLE and to optimize treatment strategies for these patients.


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