Treatment of Systemic Lupus erythematosus-Associated Thrombocytopenia with Intravenous Gamma Globulin

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Dear Sir,

Thrombocytopenia occurs frequently in systemic lupus erythematosus (SLE), and it may sometimes be resistant to the immuno-suppressant treatment [1]. We describe a patient diagnosed as having SLE with antiplatelet antibodies, who had acute thrombocytopenia associated with intravascular hemolysis and digestive hemorrhage during the course of an acute lupus relapse. There was no response to the treatment with methyl-prednisolone (MP) and cyclophosphamide (CPM). After the treatment with intravenous γ -globulin (IVGG) there was a recovery of the platelet count and the extrarenal affection disappeared.

A 17-year-old woman was diagnosed as having SLE with skin, articular and renal involvement at 13 (diffuse proliferative glomerulonephritis type IV according to the WHO classification) and started treatment with prednisone and oral CPM. In August 1992 she was admitted to hospital because of a new lupus relapse and treatment with a 0.7 g/m² monthly dosis of CPM bolus was started. In December 1992, 15 days after the fifth bolus of CPM, she had a new lupus relapse with polyarthritis, pleuropericarditis, microangiopathic hemolytic anemia, acute thrombocytopenia and rapidly progressive renal insufficiency. Analysis: Serum Creatinine 1,325 μ mol/l, leukocyte count 2×10^9 /l (4% metamyelocytes, 2% myelocytes, 14% band forms), platelets 10×10^9 /l, hemoglobin 51 g/l, hematocrit 0.14, lactic dehydrogenase 1,640 U/ml, abundant schistocytes, C3 28 mg/dl (normal 85-170), C3pa 16 mg/dl (normal 11-50), C4 7 mg/dl (normal 9-40), antinuclear antibodies (+) 1/2,560 (homogeneous pattern), antiDNA 89 U/l (normal

< 15), positive antiplatelet antibodies, negative anti-Sm, anti-Ro, anti-La and ANCA antibodies. Although for the next 3 days a new dose of 1,000 mg of MP and 2 mg/kg/day of CPM was given, she was still thrombocytopenic and had a digestive hemorrhage. Soon after her admission, she needed a substitutive hemodialysis treatment through a femoral catheter. No plasmapheresis treatment was recommended as there was an active hemorrhage. Four days after her admission, IVGG (0.4 g/kg/day for 5 days) was given. The platelet count increased progressively from the 1st day after finishing the treatment and stabilized at 250×10^9 /l 10 days later. The signs of lupus involvement remitted, although the renal function was not recovered. Now she is treated

with oral pred-nisolone (0.5 mg/kg/day) with no activity signs, and requires a substitutive treatment with hemodialysis.

IVGG was introduced as a treatment of the primary immunodeficiency syndromes. It can also prevent infections in immunocompromised patients [2]. Recently their effect on the treatment of several autoimmune diseases has been described, such as myasthenia gravis, Guillain-Barré disease, Kawasaki disease, vasculitis associated with neutrophil anticytoplasmic antibodies, autoimmune cytopenia, among other entities [3, 4]. It is also an effective treatment of the idiopathic membranous nephropathy and of the nephropathy associated with SLE [4-7]. In 1981 Imbach et al. [8] found out that IVGG was effective in the treatment of idiopathic thrombocytopenic purpura when high doses were given. However, this therapeutic effect has not been proved so far in acute thrombocytopenia as-

sociated with SLE. The action mechanism of IVGG is now controversial. IVGG has antibodies directed against idiotypes expressed by antibodies which are in the serum of the patients with different types of autoimmune diseases, but its beneficial effect is not only due to the passive transference of these antibodies. IVGG may interact with the basic structure, function and dynamic of the immune system, which might account for the clinical response persisting beyond the half-life of the injected immunoglobulin [3]. Although greater experience is needed to prove the effectiveness of IVGG, we believe that, when high doses are given, it may be useful to control specific manifestations of SLE, especially in the cases in which the treatment with plasmapheresis may not be indicated (infection, thrombocytopenia, active hemorrhage).

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