Evans Syndrome in Renal Transplantation: Correlation between Drops in Platelet and Red Blood Cell Counts and Rejection

C. Campieri
F. Zanchelli
S. Stefoni
V. Bonomini

Nephrology Department, S. Orsola University Hospital, Bologna, Italy

Dr. Claudio Campieri, Nephrology Institute, S. Orsola University Hospital, V. Massarenti, 9, I-40138 Bologna (Italy)

Dear Sir,

Immune thrombocytopenic purpura (ITP) can occur in patients with kidney transplantation in whom a ‘drop’ in platelet count can be the only clinical expression of this disease [1]. ITP can be associated with autoimmune hemolytic anemia in Evans syndrome [2]. In a transplanted patient with ITP we detected features of Evans syndrome and found a highly significant correlation between the increase in serum creatinine associated with rejection on the one hand, and platelet and RBC drops on the other hand.

A 59-year-old woman who had undergone cadaveric renal transplantation in January 1979 presented a serum creatinine of 2 mg/dl while under therapy with azathioprine 50 mg/day and prednisone 25 mg/day. In October 1995, an immune thrombocytopenic crisis with a platelet count of 80,000/mm3 was diagnosed in the presence of direct and indirect antiplatelet antibodies. A previous drop in platelet count (50,000/mm3) dated back to June 1987 during an episode of rejection (serum creatinine up to 2.3 mg/dl). By that time, RBC count was 2,910,000/mm3 with a Hb of 9 g%. A cross-match was positive for HLA-A33 antigen. Enhanced antirejection therapy resolved the episode of rejection (creatinine 1.1 mg/dl) and increased the platelets (up to 180,000-220,000/mm3) as well as RBCs (up to 3,870,000/mm3) within 5 days.

In August 1987, a thrombophlebitis of the right leg led to hospital admission associated with thrombocytopenia (60,000/mm3) and autoimmune hemolytic anemia (3,125,000/mm3) with a positive direct and indirect Coombs’ test.

In this patient we analyzed the serum creatinine values and platelet and RBC counts during 3 hospital admissions for rejection episodes and 5 hospital admissions not related to rejection (ureteral stenting, acute gastroenteritis, pulmonary infection, cyclosporine toxicity), as shown in table 1.

Asymptomatic drops in platelet count (80,000-120,000 platelets/mm3) and hemolytic anemia (< 3,000,000/mm3) have been detected during hospital admissions for rejection, and an enhancement in immunosuppressive therapy either resolved the rejection or increased platelet and RBC count.
A significant correlation between creatinine increase and RBC and platelet drops in rejection has been documented (platelets 60,890 ± 19,405 during rejection episodes and up to 225,363 ± 18,147 out of rejection; RBCs 3,496,000 ± 138,254 during rejection and up to 4,694,545 ± 1,658,722 out of rejection; p < 0.001).

When hospital admission was not related to rejection, we could not detect such a correlation. In this transplanted patient, we detected either ITP or hemolytic anemia featuring together in Evans syndrome [2-4] as originally described in 1949. This case seems to be the first description of Evans syndrome in a renal transplant patient and therefore we recommend a careful analysis of mild immunohematological episodes in renal transplant patients. Only such an approach could lead to the definitive and correct diagnosis of Evans syndrome.

Thrombophlebitis that is an clinical hallmark of the syndrome had been present in this patient, as a single episode, 9 years before the definitive diagnosis [5].

The data we have reported suggest that rejection and immunohematological events may occur together [6]. ‘Momentary immunological breakdowns’ in the therapeutic effectiveness of immunosuppressive therapies can cause an imbalance between production and destruction of blood cells.

Even mild hematological events can herald the full-blown symptoms of renal rejection, and this observation is worth this report.

Table 1. Platelet and RBC counts during rejection and outside rejection episodes

Student’s t test for platelet and RBC counts. A = Rejection-related data; B = data not related to rejection; n = number of determinations.