sTfR levels in the REF group (ANOVA, p=0.96).

Raised sTfR levels have been reported in cases with polycythemia[3,4] although it is unclear whether this increase is related to red cell mass, disease activity or iron status. Iron deficiency is closely associated with high sTfR values and sTfR levels progressively increase in parallel with the different iron deficiency stages, from the earliest stages with only low ferritin values to fully expressed iron deficiency anemia.[5] In our study, the difference in sTfR levels between polycythemia and REF groups was probably due to iron status. Thus, at variance with serum erythropoietin,[6] the possible role of sTfR in evaluating erythroblastic mass in polycythemia remains unresolved.

Key words

Polycythemia, serum transferrin receptor.

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Elective splenectomy in relapsing thrombotic thrombocytopenic purpura

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Between 20 and 40% of surviving patients with thrombotic thrombocytopenic purpura (TTP) have relapses. Plasma exchange therapy is usually effective in treating relapses, but this treatment does not prevent TTP recurrence. The role of splenectomy in relapsing TTP is still controversial. We describe a patient with multiple relapses of TTP who was successfully treated with elective splenectomy.

Thrombotic thrombocytopenic purpura (TTP) is a rare, life-threatening disorder of unknown pathophysiology. Without treatment, TTP is a rapidly progressive and fatal disease, with 90% of patients surviving less than 3 months. Treatment with plasma exchange, often used in combination with corticosteroids, antiplatelet agents, splenectomy and vincristine has reduced mortality to 20%.[1-3] However, between 20 and 40% of surviving patients have relapses. Relapses can occur as early as a few weeks after recovery, but also after an interval of many years.[4,5] Plasma exchange therapy is usually effective in treating relapses, but this treatment exposes the patient to blood products from numerous donors and does not reduce the relapse rate. Various interventions, including antiplatelet agents, corticosteroids and splenectomy have been used to prevent relapses.[2] Some studies have suggested that splenectomy has a role in the management of relapsing TTP, since it seems to reduce the frequency of relapse.[6-8] We describe a patient with multiple relapses of TTP who was successfully treated with elective splenectomy during remission.

A 27-year-old female was diagnosed as having TTP in May 1992. Clinical manifestations in the initial episode of TTP were fatigue, headache, hematuria, petechiae and hematomas. At admission, biological findings were as follows: hemoglobin 8.2 g/dL, platelet count 15×10^9/L, LDH 897 U/L, bilirubin 1.8 mg/dL and serum creatinine 1 g/dL. The initial episode and two early recurrences through 1992 were successfully treated with fresh plasma transfusions, plasma exchange, corticosteroids and vincristine (Figure 1). The patient remained in remission until 1994. Between March 1994 and February 1996, this patient had five relapses (incidence of 2.5 relapses/year). The disease-free interval varied from 3 weeks to 9 months (Figure 1). Presenting signs and symptoms during relapses were similar but less severe than those observed in the first episode. The patient repeatedly responded to therapy with plasma infusions and prednisone. Splenectomy was performed 18 days after the last relapse when the platelet counts and LDH levels had returned to normal values with the treatment schedule mentioned before (Figure 1). One week before splenectomy pneumococcal vaccine was given. There were no perioperative complications. The spleen was not enlarged (weight 110 g). The histologic study showed a moderated hyperplasia of follicles in the white pulp. Microthrombi were absent. In the 2 years of follow-up since the splenectomy, the patient had no further relapses.

The pathophysiology of TTP is still poorly under-

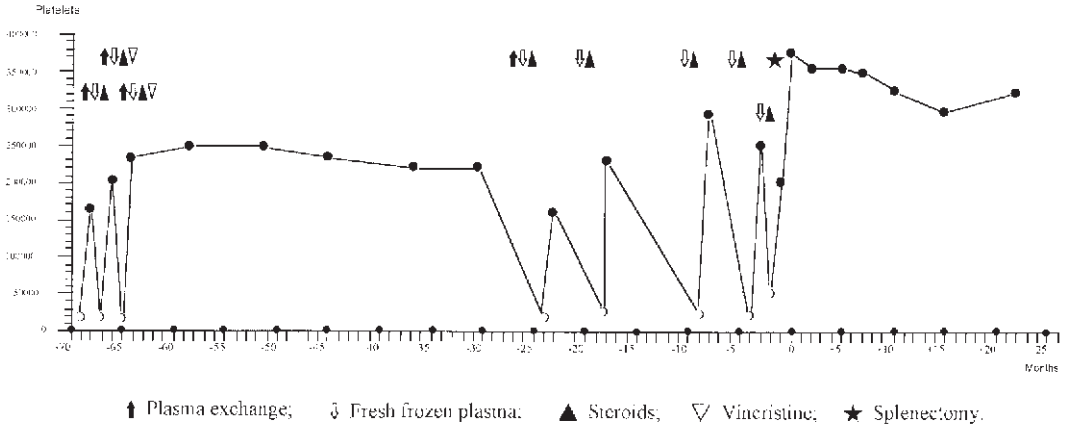

Figure 1. Platelet counts before and after treatment.

stood. The current view includes endothelial damage, presence of abnormally large factor VIII/von Willebrand multimeric structures and a platelet aggregating factor.[9] The contribution of the spleen in this process is unknown. Despite this fact, splenectomy remains an important part of multimodality therapy for TTP. Splenectomy has been used primarily in patients in whom plasma exchange failed to improve haematologic values, but it has been associated with a high mortality.[1,2] Much better results have been obtained when splenectomy was performed at the time of relapse in patients with relapsing TTP.[6,7] This seems to increase disease free intervals with less morbidity and mortality. Recently it has been suggested that splenectomy for relapsing TTP would be more convenient while in remission than splenectomy performed during the relapses. In a small series, splenectomy performed during remission at least postponed relapses in all cases with no surgery-related deaths or major complications.[8] Elective splenectomy was done in our patient after remission of the seventh relapse, and two years later the patient remains in remission. Our result supports evidence that elective splenectomy increases disease-free intervals in patients with relapsing TTP with a minimal risk of perioperative complications. Further studies are warranted to elucidate the role of elective splenectomy in relapsing TTP.

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