MASSIVE HEPATOMEGALY FOLLOWING SPLENECTOMY FOR MYELOID METAPLASIA

To the Editor:

We read with great interest the article by Towell and Levine (Am J Med 1987; 82: 371–375) concerning a case of massive hepatomegaly after splenectomy for myeloid metaplasia. We have recently observed a similar case. A 51-year-old man was first hospitalized in our department in October 1986, complaining of postprandial fullness and mild weight loss (3 to 4 kg). He had a past medical history of typhoid fever and malaria, but denied recent fever. On admission, physical examination showed a protuberant abdomen due to massive splenomegaly, which spanned the entire left abdomen; the liver was not enlarged on palpation; no superficial lymph node enlargement was observed. Laboratory data on admission were as follows: sedimentation rate normal, hemoglobin 10.2 g/dl, hematocrit 29.6 percent, white blood cell count 13,900/mm^3, platelet count 350,000/mm^3. The peripheral blood smear showed many anisocytes, teardrop poikilocytes, and giant platelets. Other laboratory values included serum glutamic oxaloacetic transaminase 24 units/liter, serum glutamic pyruvic transaminase 40 units/liter, lactic dehydrogenase 236 units/liter, alkaline phosphatase 352 units/liter, and bilirubin 0.9 mg/dl. A bone marrow biopsy specimen showed diffuse severe depletion of the myeloid and erythroid elements, abundant fibrosis, and clusters of megakaryocytes. ^{59}Fe and ^{51}Cr erythrokinetic study showed reduced skeletal erythropoietic activity with major splenic uptake. A diagnosis of myelofibrosis with myellid metaplasia (MMM) was made and the patient was discharged without therapy. In June 1987, the patient was re-hospitalized for increasing abdominal pain, with major weight loss and abdominal swelling. Physical examination showed impressive splenomegaly with the median edge palpable in the right lower abdomen. No changes in laboratory data were observed. On June 20, 1987, the patient underwent elective splenectomy and a 6,000-g spleen was removed; at laparotomy the liver was noted to be not enlarged. Pathologic examination of the spleen showed myeloid metaplasia. (Liver biopsy was not performed.) On immediate postoperative days, the platelet count rose to 1,000,000/mm^3 and white blood cell count increased to 31,700/mm^3 with 64.3 percent neutrophils, 23 percent lymphocytes, and 8.5 percent monocytes. Abdominal echoscan revealed a silent portal vein thrombosis. The patient was discharged on the ninth postoperative day; he was well, taking no medication, and on a normal diet. In September 1987, the patient complained of asthenia, malaise, and abdominal swelling. On readmission, physical examination showed pallor, mild pedal edema, and liver edge palpable 15 cm below the right costal margin, no ascites, no pleural effusion, and no lymph node enlargement was present. His hemoglobin level was 5.6 g/dl, hematocrit 19.6 percent, white blood cell count 198,000/mm^3 with 27 percent neutrophils, 44.8 percent lymphocytes, 1.7 percent eosinophils, 1.5 percent basophils, 11.0 percent metamyelocytes, 7 percent myelocytes, and 35.0 nucleated red cells per 100 white blood cells. Other laboratory data included the following: albumin 3.4 g/dl, plasma iron 69 μmol/liter, ferritin 174.65 ng/ml, total cholesterol 138 mg/dl, total serum bilirubin normal, serum glutamic oxaloacetic and glutamic pyruvic transaminase normal, prothrombin time 14.8 seconds, partial thromboplastin time 29.4 seconds, lactic dehydrogenase 1,382 units/liter (normal, 230 to 460), alkaline phosphatase 1,806 units/liter (normal, 98 to 279). Chromosomal pattern was normal. A liver scan with ^{99}Tc revealed massive hepatomegaly with heterogeneous uptake. The patient underwent transfusion of two units of blood and received oral therapy with folinic acid, vitamin B_6, and iron, with prompt hematologic recovery. At discharge, his hemoglobin level was 10.1 g/dl, hematocrit 38 percent, white blood cell count 181,000/mm^3 with normal peripheral smear, platelet count 1,000,000/mm^3, and reticulocyte count 22 percent. Bone marrow aspiration demonstrated stromal proliferation with normal myeloid series, erythroid hyperplasia, and mild dyserythropoiesis. One month later the patient was in good health. The hemoglobin level was 13.0 g/dl, hematocrit 48 percent, white blood cell count 35,000/mm^3, platelet count 600,000/mm^3, and alkaline phosphatase level 1,082 units/liter; results of liver function tests were normal, with persistent hepatomegaly.

Symptomatic splenomegaly is the most common indication for elective splenectomy in MMM [1]. Development of hepatomegaly after splenectomy in patients affected by MMM is less common; when it occurs, the liver's growth develops over many months or years and any element could preoperatively predict the development of this event [2]. Post-splenectomy leukocytosis is a common feature in patients with MMM and may cause difficulties in diagnosis of infectious complications [3] or malignant evolution. In our patients, no infection or malignant disease was found: the leukemoid reaction and dyserythropoietic anemia, resolved by medical therapy, were probably due to a massive compensatory hepatic myeloid metaplasia.

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POOR SURVIVAL OF PATIENTS WITH IDIOPATHIC CARDIOMYOPATHY CONSIDERED TOO WELL FOR TRANSPLANTATION

To the Editor:

I read with interest the recent article by Stevenson et al (Am J Med 1987; 83: 871–876) on the survival of patients