Idiopathic Thrombocytopenic Purpura Occurring in a Subject Previously Splenectomized for Traumatic Splenic Rupture

Role of Splenosis in the Pathogenesis of Thrombocytopenia

ERIC M. MAZUR, M.D.
WILLIAM W. FIELD, M.D.
C. ELTON CAHOW, M.D.
FRED J. SCHIFFMAN, M.D.
THOMAS P. DUFFY, M.D.
BERNARD G. FORGET, M.D.
New Haven, Connecticut

A 25 year old woman presented with idiopathic thrombocytopenic purpura. She had undergone splenectomy 12 years previously for traumatic splenic rupture. Thrombocytopenia was ultimately resistant to steroid therapy. Howell-Jolly bodies were absent from the peripheral smear and ^{99m}TC-spleen scan demonstrated foci of increased uptake thought consistent with accessory spleens. However, splenosis alone was demonstrated at laparotomy, and all visible splenotic tissue was surgically removed. The patient responded and adequate platelet counts were maintained after discontinuation of steroid therapy. The functional capacity of splenic implants has been previously demonstrated both in animal and man. However, reports linking splenosis to hematologic disease are rare. In the present case, characteristic splenic function was demonstrated by both the ^{99m}Tc-spleen scan and the absence of the typical peripheral blood findings of asplenia. The hematologic response to the removal of the splenotic tissue attests to its importance in maintaining the thrombocytopenic state. In the setting of prior splenectomy for splenic trauma, splenosis may contribute to hematologic disease. Removal of this splenotic tissue may result in hematologic remission.

Splenectomy is usually necessary in the treatment of chronic idiopathic thrombocytopenic purpura (ITP) in the adult. Seventy to 90 per cent of patients so treated achieve a permanent remission [1,2]. Rarely, accessory spleens have been implicated in resistant or recurrent ITP following splenectomy. Subsequent removal of these spleens has usually resulted in clinical improvement and an increase in the platelet count [3–6].

Ectopic splenic tissue is also characteristic of the acquired entity of splenosis. Splenosis occurs when splenic fragments are autotransplanted following splenic trauma [7,8]. Despite the known functional capacity of these splenic nodules [9–12], associated hematologic disease has been only rarely described [13,14]. We report such a case here: steroid resistant ITP was present in a splenectomized patient later proven to have splenosis. The thrombocytopenia subsequently responded to surgical removal of the ectopic splenic tissue.

From the Departments of Medicine and Surgery, Yale University School of Medicine, and Yale University Health Services, New Haven, Connecticut. This study was supported in part by Grant HL07262 from the National Institutes of Health. Requests for reprints should be addressed to Dr. Bernard G. Forget, Department of Medicine, Yale University School of Medicine, 333 Cedar Street, New Haven, Connecticut 06510. Manuscript accepted April 17, 1978.

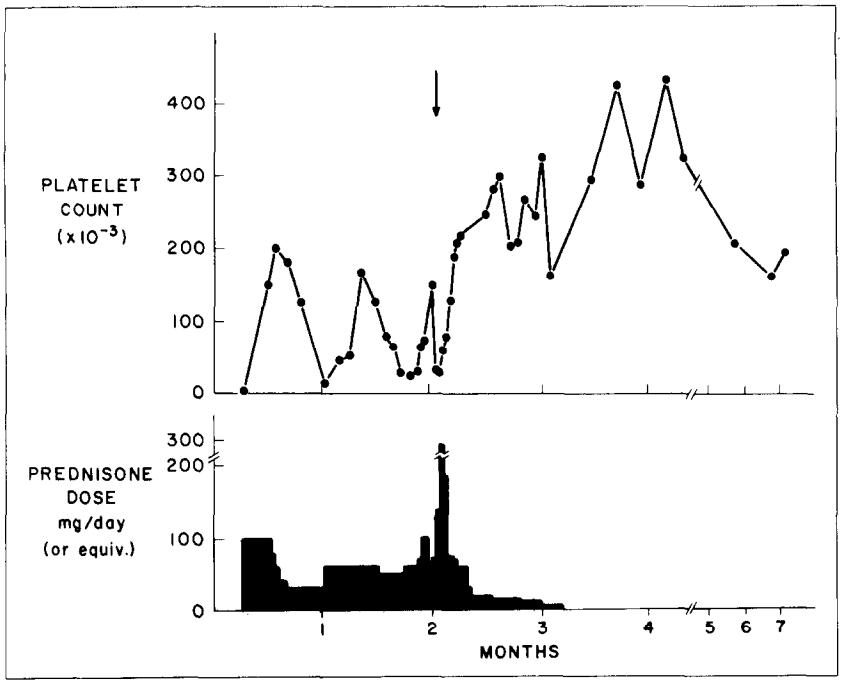

Figure 1. *Platelet count and corresponding daily prednisone dose (or equivalent) in the patient described. Arrow indicates date of surgery when all demonstrable splenotic tissue was removed.*

CASE REPORT

The patient is a 25 year old white woman. At age 12 she required emergency splenectomy following abdominal trauma. The removed spleen had been lacerated circumferentially around the lower pole with splenic pulp bulging from the surface in the area of the tear. An extensive hematoma surrounding the splenic pedicle and extending retroperitoneally was evacuated. No mention was made of residual intraabdominal splenic tissue.

She did well until age 25 when she presented suddenly with petechiae of the lower extremities and severe menorrhagia. Initial platelet count was less than 1,000/mm^3. Bone marrow aspirate contained abundant numbers of megakaryocytes. Peripheral blood smear demonstrated essentially normal red blood cell morphology with no Howell-Jolly bodies present. Antinuclear antibody was negative. A presumptive diagnosis of idiopathic thrombocytopenic purpura was made, and therapy was started with high dose prednisone, 100 mg daily. Within two weeks the platelet count increased to 150,000/mm^3, but it decreased to 50,000/mm^3 one month later as her dose of prednisone was tapered. An increased dosage produced only a transient increase in the platelet count. The platelets subsequently decreased to less than 30,000/mm^3 on a stable dose of 50 mg of prednisone a day (Figure 1).

Because of the steroid resistance and lack of Howell-Jolly bodies in the peripheral blood smear, a ^{99m}Tc liver-spleen scan was performed. It demonstrated two areas of increased uptake in the left upper quadrant consistent with accessory splenic tissue (Figure 2). These were thought to represent accessory spleens, and the patient was admitted for accessory splenectomy two months after the initial onset of thrombocytopenia.

At laparotomy, scores of splenic implants measuring from 1 mm to 1 cm in diameter were present. These involved primarily the greater omentum, but also the liver capsule with some large deposits implanted on the inferior vena cava below the liver. A large 2.5 by 1 by 1 cm nodule was present in the site of the splenic pedicle. The areas demonstrated on liver spleen scan corresponded to these large implants. An omentectomy was performed, and all grossly evident splenic tissue was removed.

The findings on pathologic examination were consistent

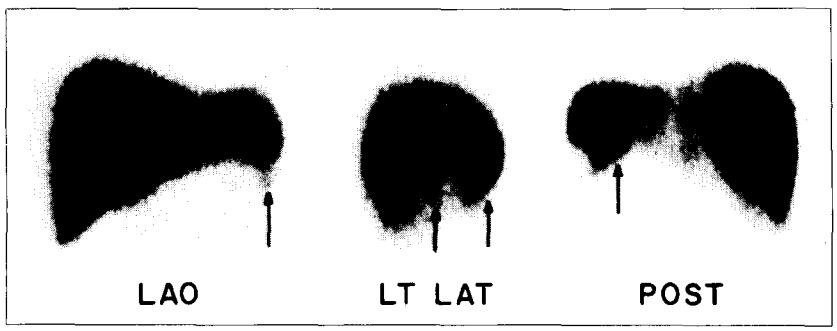

Figure 2. *Liver-spleen scan performed prior to laparotomy, left anterior oblique (LAO), left lateral (LT LAT) and posterior (POST) views. Arrows indicate two foci of abnormal radionuclide accumulation consistent with the splenotic tissue later found at laparotomy.*

with splenosis. None of the splenic nodules demonstrated the hilar arterial supply characteristic of accessory spleens.

Within five days after surgery, the patient's platelet count increased to over 100,000/mm^3 and remained greater than 150,000/mm^3 while her steroid dose was tapered. Occasional Howell-Jolly bodies appeared in the peripheral blood smear. Prednisone therapy was finally discontinued six weeks postoperatively. For the ensuing four months, the patient has been asymptomatic and her platelet count has ranged from 165,000 to 440,000/mm^3 (Figure 1).

COMMENTS

Splenosis is an acquired entity originating from the autotransplantation of splenic tissue following splenic trauma with rupture of the splenic capsule and spillage of splenic tissue into the peritoneal cavity. It is usually an incidental finding at surgery or postmortem examination. Clinical manifestations, when present, are most often those of intestinal obstruction or vague abdominal pain [15]. That these splenic nodules may function in a manner similar to the intact spleen has long been suspected. Williams [11], while examining rabbit spleen autografts, observed spontaneous erythrophagocytosis by the splenic macrophages in such autografts. Ingestion of intravenously injected India ink particles by the macrophages of the splenic implants was also seen. Some protection from Trypanosoma lewisi infection and Bartonella muris anemia has been demonstrated in rats receiving subcutaneous splenic autotransplants prior to splenectomy [16,17].

Splenic function has also been demonstrated in isolated cases of human splenosis. Shaw and Shafi [9] observed granulomas in 13 of 17 implants examined in a patient with splenosis dying with schistosomiasis and miliary tuberculosis. Both erythrophagocytosis and scattered mycobacteria were observed within the splenic nodules. Gill [10] described a splenectomized patient with a single subcutaneous splenic implant which subsequently enlarged during a relapse of malaria. Hemosiderin-laden splenic macrophages were described within the implant on pathologic examination. Splenic nodules with a maximum diameter of 3 cm have been visualized in a case of splenosis by spleen scan using ^{99m}Tc-sulfur colloid [12]. This visualization is dependent on the phagocytic function of splenic reticuloendothelial cells.

Other isolated reports have failed to demonstrate gross splenic function. Despite the known presence of more than 100 splenic nodules ranging from 2 mm to 2 cm in diameter, there was no intraabdominal uptake of ^{99m}Tc on spleen scan in a patient described by Trimble and Eason [15]. Mackie and Miller [18] described the presence of typical Howell-Jolly bodies in the peripheral smear of a patient previously demonstrated at laparotomy to have splenosis.

Splenosis has long been suspected of contributing to hematologic disease in previously splenectomized patients. This conclusion may be inferred from both the functional capacity of the splenotic nodules demonstrated in the isolated cases listed herein, and the experience relating recurrent disease to the presence of accessory spleens. Nevertheless, case reports implicating splenosis in hematologic disease are rare. An oft-quoted report by Stobie [13] purports to relate splenosis to recurrent congenital hemolytic anemia. However, this relationship is unclear in that no objective evidence of improvement following the original splenectomy is presented, and the case is further complicated by cholelithiasis and a biliary duct stricture.

In a report by Perry and Bayrd [14], Felty's syndrome recurred in a patient with splenosis. More important, however, was the presence of a massively enlarged accessory spleen weighing 1,040 g. The 41 smaller splenules ranging from 0.3 to 3 cm in diameter showed no uptake on ^{99m}Tc-spleen scan and probably contributed little to the neutropenic state.

Unique in the present case is the clear relationship of splenosis alone to steroid-resistant idiopathic thrombocytopenic purpura. Function of the splenotic nodules was demonstrated by both the ^{99m}Tc-spleen scan and the absence of the typical peripheral blood findings of asplenia.

The patient's response to the removal of the splenotic tissue attests to its importance. The appearance of Howell-Jolly bodies in the peripheral blood smear coincident with the increase in platelet count suggests that the functional capacity of the removed splenotic nodules had been sufficient to entrap both the antibody-coated platelets and the red cell nuclear remnants. In the present case, the peripheral smear was a sensitive indicator of splenic phagocytic function. The splenotic nodules may have further contributed to the thrombocytopenic state by the production of antiplatelet antibodies, a capacity which has been demonstrated previously in splenic tissue of patients with idiopathic thrombocytopenic purpura [19,20]. In the setting of prior splenectomy for splenic trauma, the absence of Howell-Jolly bodies should alert one to the possible presence of splenosis. In such a patient with resistant or recurrent hematologic disease, the removal of the splenotic tissue may result in hematologic remission.

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