Functional Asplenia

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Functional asplenia, or hyposplenism, has been described as impaired or absent splenic function despite presence of the spleen. The spleen's ability to accumulate technetium during a liver-spleen scan is impaired. Functional asplenia may be intermittent, reversible, or irreversible. Concurrently, the peripheral smear usually shows Howell-Jolly bodies, target cells, spherocytes, and poikilocytes. The diagnosis of functional asplenia is important clinically because it is often associated with an increased frequency of infections.

Case Report

A 16-year-old black patient with a history of sickle cell disease was admitted to the hospital because of chest pain and back pain. Physical examination revealed a well-developed, alert, icteric male patient in mild distress secondary to pain with a blood pressure of 120/80 mmHg. He was afebrile, and oral mucosae were dry. Cardiopulmonary examination was unremarkable. Abdominal examination did not show scars or ascites. The liver was felt 4 cm below the right costal margin, and the spleen was noted to be also enlarged 3 cm below the left costal margin. The rest of the physical examination was noncontributory.

Laboratory tests showed a hemoglobin of 14.8 g/dL and hematocrit of 44 percent. The white blood cell count was 6,700/mm^3. The reticulocyte count was 4.6 percent. The total bilirubin was 3.4 mg/dL with a direct fraction of 0.8 mg/dL. Blood glucose, electrolytes, liver chemistries, and urinalysis were normal. A chest x-ray film was also normal.

The patient was treated with vigorous hydration, supplemental oxygen, and analgesics. Two days later he developed pneumonia, which required the use of intravenous antibiotics. The patient continued to have diffuse pains despite hydration and analgesic therapy. Five days after admission he developed left upper quadrant ten-

Continued on page 407

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Continued from page 403

derness corresponding to the area of the enlarged spleen.

Ultrasound examination of the abdomen showed an enlarged spleen and liver. There were no gallstones or ascites. A liver-spleen scan using technetium 99 demonstrated the presence of an enlarged liver and absence of the spleen. The hematocrit, which was 44 percent on admission, began to fall progressively after hydration. Seven days after admission the hematocrit dropped as low as 23 percent, and it was decided to transfuse three units of packed red blood cells in an attempt to correct his anemia and alleviate his sickle cell crises. After the blood transfusions, the patient improved greatly and from that point on was essentially pain free. The patient became afebrile shortly thereafter and was discharged home in good condition.

One week after discharge, a follow-up liver-spleen scan showed the presence of the spleen.

Table 1. Diseases and Agents Associated With Functional Asplenia

Sickle cell disease[1,2,3]
Hemoglobin sickle cell disease[1]
Thalasemia trait[1]
Polycythemia vera[1]
Multiple myeloma[1]
Ulcerative colitis[2]
Dermatitis herpetiformis[2]
Celiac-sprue disease[1,4]
Reticulum cell sarcoma[4]
Amyloidosis[4]
Radiation therapy[5]
Discoid lupus erythematosus[6]
Systemic lupus erythematosus[6,7]
Crohn's disease
Thyrotoxicosis[8]
Graft-host disease[9]

Discussion

Sickle cell anemia is present in approximately 0.15 percent of black children in the United States. This disorder is characterized by the substitution of valine for glutamic acid at position 6 of the beta chain. Sickle cell anemia affects almost any organ, and it has been shown that deoxygenation may change the shape of the erythrocyte from a biconcave disk to an elongated form or "sickle-shaped" cell. Other factors, such as acidosis and increased erythrocyte 2,3-DPG (2,3-diphosphoglycerate), will promote sickling. Most of the symptoms related to this disorder are due to a vaso-occlusive phenomena, which may involve almost any organ.

The anatomic presence of the spleen without the ability to accumulate sulphur colloid tagged with technetium 99 has been termed *functional asplenia*. It was initially described in sickle cell disease,[1,2] and since then has been associated with diverse diseases and chemical agents (Table 1).[1-9] The presence of Howell-Jolly bodies, spherocytes, target cells, and poikilocytes in the peripheral smear correlates with the diagnosis of functional asplenia.

Although causative mechanisms of functional asplenia are unknown, a possible explanation would be an abnormal splenic circulation. Impaired circulation can be seen in sickle cell disease, where large numbers of sickle red cells produce obstruction of the blood flow. Patients with sickle cell disease also have an increase in blood viscosity, possibly because of deoxygenation of blood, contributing further to decreased blood flow. Obstruction of blood flow will cause a diversion of blood through intrasplenic shunts, thereby making the splenic flow bypass the phagocytic reticuloendothelial system.[2] Furthermore, a decrease of a plasma opsonizing factor, causing reticuloendothelial blockade, has also been reported as a possible mechanism in the development of functional asplenia.[10] Functional asplenia has been associated both with pneumococcal infections in patients with sickle cell anemia[2,3] and with gram-negative sepsis in postcolectomy patients with ulcerative colitis.[11] Infections may be secondary to a defective clearance of bacteria by the spleen.

The patient herein reported presented with the typical sickle cell crisis and later developed a pul-

monary infection. He was also found to have an enlarged and tender spleen that failed to be visualized with a liver-spleen scan. However, a follow-up liver-spleen scan revealed the presence of the spleen, establishing the diagnosis of functional asplenia.

The management of patients with sickle cell disease and functional asplenia may include hydration and prophylaxis with pneumococcal vaccine (Pneumovax). One of the most important considerations is the early recognition of infections and the use of appropriate antibiotic therapy. The use of blood transfusions in patients with sickle cell disease has been shown to restore splenic function[3]; however, this is achieved only temporarily.

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