Reversible Functional Asplenia
In Chronic Aggressive Hepatitis

Vijay M. Dhawan, Richard P. Spencer, and John J. Sziklas

University of Connecticut Health Center, Farmington, Connecticut, and Hartford Hospital, Hartford, Connecticut

A 61-year-old man presented with aggressive hepatitis. Howell-Jolly bodies were present in circulating erythrocytes and the spleen failed to accumulate intravenously administered Tc-99m sulfur colloid. The patient thus demonstrated functional asplenia. He was treated with high doses of steroids. Four years later, Howell-Jolly bodies were no longer present in circulating erythrocytes. In addition, the spleen had regained the ability to accumulate intravenously injected radiocolloid. Hence, the patient had reversed his functional asplenia. The reported cases of this disorder (reversible functional asplenia) were reviewed and a preliminary classification was proposed.


The concept of functional asplenia (anatomic presence of the organ but without the ability to accumulate intravenously administered radiocolloid) was first described in association with sickle cell disease in 1969 (1). Since then, functional asplenia has been identified in several disorders (2,3). It has also been recognized that some instances of functional asplenia may be transient or at least temporarily reversible (4,5). For example, functional asplenia associated with sickle-cell anemia could be transiently reversed by transfusion of normal red blood cells (4). Reversible functional asplenia has been demonstrated in some cases of cyanotic congenital heart disease (5), and episodically in hemoglobin SC disease (6). Recently a child with combined immunodeficiency disease has been identified as having reversible functional asplenia (7). In the present report we describe a case of reversible functional asplenia associated with chronic aggressive hepatitis. A classification of the disorders associated with this reversible splenic dysfunction is proposed.

CASE REPORT

A 61-year-old white man had been admitted to the hospital 4 years previously because of fever, anorexia, and jaundice. Liver-function tests at that time were grossly abnormal with total serum bilirubin of 4.2 mg/dl, alkaline phosphatase 106 units, SGOT 460, and SGPT of 502. Hemoglobin was 14 g/dl with a hematocrit of 40%. There were abundant Howell-Jolly bodies in the peripheral erythrocytes. A radiocolloid liver-spleen scan, performed with 3 mCi of Tc-99m sulfur colloid i.v. showed hepatomegaly with a pattern consistent with diffuse hepatocellular disease. The spleen was not visualized and was presumed to be functionally absent (Fig. 1, top). A liver biopsy showed hepatic necrosis with swelling of hepatocytes with granular cytoplasm. There were areas of inflammatory bridging of portal tracts, as well as early peripheral fibrosis. A diagnosis of chronic aggressive hepatitis with functional asplenia was made. The patient was started on 40 mg of prednisone daily. This was reduced to

---

Received June 6, 1978; revision accepted Aug. 3, 1978.

For reprints contact: Richard Spencer, Dept. of Nuclear Medicine, University of Connecticut Health Ctr., Farmington, CT 06032.
This admission, on consistent count able was phatase sulfur showed 30 mg/dl. Hemoglobin 14 g/dl, white cell count of 18,100, with a normal differential. A peripheral smear showed Pappenheimer bodies and toxic granules in white blood cells, but there were no Howell-Jolly bodies in erythrocytes. This was true on repeated examinations. A liver-spleen scan, performed following i.v. administration of 3 mCi of Tc-99m sulfur colloid again showed mild hepatomegaly with findings consistent with diffuse hepatocellular disease. The spleen was visualized, being 7 cm in the longest dimension. Liver-function tests showed total serum bilirubin of 4.2 mg/dl, alkaline phosphatase of 71 units, and SGOT 178. The patient’s dose of prednisone was returned to 40 mg and he was discharged home for followup in clinic.

DISCUSSION

The spleen is an organ not only of hematologic significance but also of importance (particularly in the young) in the defense against infections. Recognition of the incidence of infection in patients who had the spleen surgically removed, and also in individuals with congenital absence of the spleen, hypoplasia of the spleen, or functional asplenia in sickle cell disease, has served to emphasize this point. Functional asplenia has been described in association with several diseases, but there are only a few instances of reversible functional asplenia. The case described here is unique in that it is the first reported instance of reversible functional asplenia in an adult with chronic aggressive hepatitis. At the initial presentation, there were abundant Howell-Jolly bodies in the peripheral circulation, and the patient had no demonstrable spleen on the radiocolloid scan. Four years later, while the patient was being treated with steroids, follow-up radiocolloid scan studies showed a small but functioning spleen. Repeated peripheral smears failed to reveal any Howell-Jolly bodies. The cause of the initial episode of functional asplenia, and then spontaneous reversal, remains unclear.

Howell-Jolly bodies are nuclear remnants within circulating erythrocytes, and they are usually removed from red blood cells by the spleen. With few exceptions, a coupling exists between the presence of Howell-Jolly bodies in erythrocytes and the lack of splenic ability to extract radiocolloid. The exceptions occur in two instances. Sudden occlusion of the splenic vasculature (such as a massive embolus) causes abrupt loss of the ability to accumulate radiocolloid, but red cell inclusions have not had an opportunity to accumulate (8). At the other extreme, massive outpouring of erythrocytes from the bone marrow (following hematologic stress) may temporarily “overwhelm” the splenic ability to extract Howell-Jolly bodies, but the uptake of radiocolloid is unaffected (9). Neither mechanism was apparently operative in this case.

The presence of Pappenheimer bodies (iron granules within erythrocytes) in this patient is of uncertain significance. Such inclusions can be seen in the presence of liver disease. There was no clinical evidence of hemochromatosis, and both the serum iron (72 µg/dl) and total iron binding capacity (280 µg/dl) were normal on a prior admission.

We offer an initial classification, albeit tentative, of disorders resulting in reversible functional asplenia (Table 1). Patients in the first group have as a common basis the presence of splenic anoxia that can be reversed. The first three of these disorders are well documented. The fourth, transient occlusion or recanalization of major splenic vessels, has been observed clinically but is insufficiently documented at present. The second group of disorders (actions on splenic cells) is more tentative. Such patients might have an immunologic basis as the common factor. In celiac disease there is a high incidence of functional asplenia, and this disease is also associated with a leukocyte-migration-inhibition factor (10). Whether the functional asplenia of celiac disease is reversible after steroid therapy is as yet unknown. Since steroids also have an anti-inflammatory effect, they might be mediating a response in some instances by effects either on splenic cellular elements or blood vessels. It is apparent that continued surveillance is needed in order to identify cases of reversible functional asplenia and their causes.

ACKNOWLEDGMENT

This work was supported by USPHS CA 17802 from the National Cancer Institute.

| TABLE 1. INITIAL CLASSIFICATION OF DISORDERS ASSOCIATED WITH REVERSIBLE FUNCTIONAL ASPLEINIA |
|---|---|
| 1. Likely related to low splenic oxygen content: |
| a) cyanotic heart disease in infants, treated (6); |
| b) sickle cell disease, with transfusion (4); |
| c) Hemoglobin S.C. disease during and after “crisis” (6); |
| d) transient occlusion, or recanalization of major splenic vessels. |
| 2. Possibly related to effects on splenic cells: |
| a) combined immunodeficiency (7); |
| b) chronic aggressive hepatitis. |
REFERENCES


Detection of a Large Arteriovenous Fistula between the Internal Iliac Vessels by Radionuclide Angiography

Tatsuya Miyamae, Mutsumi Fujioka, Yoshihata Tsubogo, Yoshito Tonariya, Yutaka Dohi, Yoshihisa Akashi, and Hiroshi Watanabe

Saitama Medical School, Saitama, Japan

A patient evaluated for heart failure was found by routine radionuclide angiography to have a large internal iliac arteriovenous fistula of presumed postoperative origin. The value of radionuclide angiography is described with a review of the literature on such unusual cases.


Radionuclide angiography is an excellent procedure as an initial screening test for detection of aneurysm or other vascular diseases. We recently observed an internal iliac arteriovenous fistula (AVF) in a 60-year-old woman in which radionuclide angiography demonstrated an unusual pattern. This unusual case is presented here.

CASE REPORT

A 60-year-old woman was admitted to Saitama Medical School Hospital because of recurrent exertional dyspnea for the past 3 years. The patient had had a supracervical hysterectomy 34 years previously for a chorionic epithelioma and right upper abdominal surgery 15 years previously, the nature of which was obscure.

Examination revealed a blood pressure of 130/80 mmHg, pulse rate 90 per minute and irregular, respiratory rate 21 per minute, and bilateral distention of the jugular veins. A grade 3/6 systolic murmur was heard in the left third and fourth intercostal space. The abdomen was flat, soft, and without palpable masses. By auscultation, a continuous murmur with systolic intensification was heard in the lower abdomen. No adnexal mass was detected on vaginal examination, but an expansile pulsation was noted in the right uterine fornix. The electrocardiogram revealed an atrial fibrillation with left-sided axis deviation, the chest radiograph showed cardiomegaly with deviation to the left, and an abdomen radiograph revealed a circular-shaped calcification in the right pelvis.

Routine anterior radionuclide angiograms and whole body scintigram were obtained with a scintillation camera and low-energy all-purpose collimator, following bolus injection of 15 mCi of Tc-99m red blood cells. The angiograms demonstrated early filling of the inferior vena cava and indicated laminar blood flow from the