

# Accessory Spleens in the Thoracic and Abdominal Cavities After a Relapse of Idiopathic Thrombocytopenic Purpura: A Case Report

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This case report presents a highly unusual finding of ectopic splenic tissue in both the thoracic and abdominal cavities in a patient with recurrent idiopathic thrombocytopenic purpura (ITP).

**Key Words:** technetium-99m-sulfur colloid; accessory spleen; idiopathic thrombocytopenic purpura

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It is well documented that accessory spleens occur in 10% of the population and that their presence generally has no clinical significance (1,2). In the case of hematologic disorders, however, they appear to occur with a higher frequency (3) and are reported to be responsible for the recurrence of idiopathic thrombocytopenic purpura (ITP) (4). As cited by Appel (1), Finkelstein postulated this relationship as early as 1921. Recent studies have reported the occurrence of accessory spleens in the range of 6% to more than 25% after splenectomy for ITP (5–7).

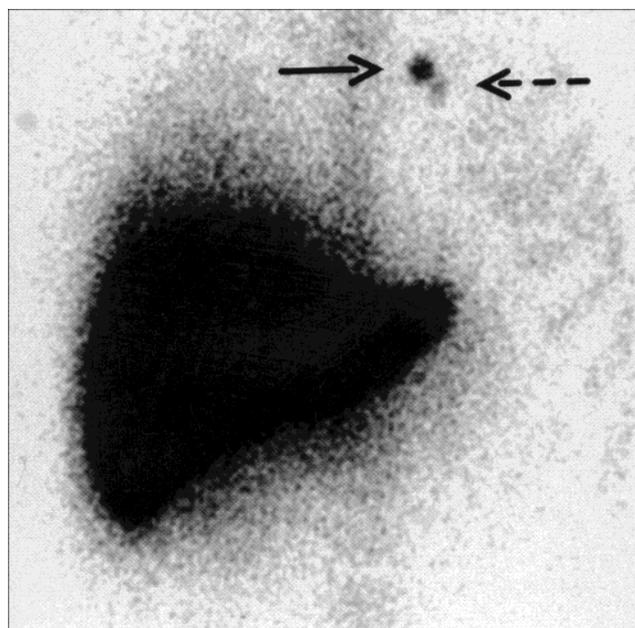
According to estimates, 1 in 10,000 people per year in the US develops ITP. This disease is characterized by immunologic platelet destruction that is believed to be humorally mediated by a gamma globulin (IgG) antibody directed against a platelet-associated antigen. Spontaneous bleeding occurs in the subcutaneous tissue causing the appearance of purple patches on the skin. Although corticosteroid management initially induces remission in many adults, eventual splenectomy is required in about 70%–80% of cases (8). Recurrent thrombocytopenia postsplenectomy for ITP presents a significant challenge clinically. It could represent persistence or recurrence of the primary disease process, functionally significant ectopic splenic tissue (accessory spleen or splenosis), or bacterial or viral illnesses. If the culprit is ectopic splenic tissue, accessory splenectomy generally is considered whenever the tissue can be localized.

Accessory spleens have been described in a wide variety of locations with the majority in a limited area around the main

spleen. The most common areas are the splenic hilus or the tail of the pancreas, with the splenic margin being the next most common area. They also may lie in the omentum, gastrocolic and gastrosplenic ligaments, ovaries or scrotum. Ectopic splenic tissue has been detected through various radiographic and scintigraphic procedures.

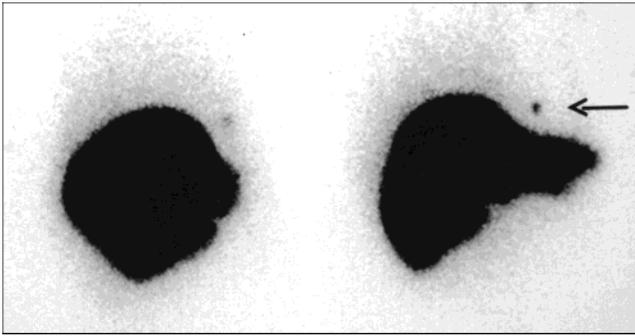
## CASE REPORT

A 47-y-old man presented with a platelet count of 51,000/mm<sup>3</sup> (normal range = 150 K–450 K/mm<sup>3</sup>) and easy bruising. He was originally diagnosed with ITP in 1973 at which time he had a splenectomy followed by an accessory splenectomy 1 y later. Aside from 2 visceral hernia repairs, he had been disease and trauma free for approximately 25 y. The current symptoms signaled a recurrence of ITP and he subsequently was referred



**FIGURE 1.** Anterior planar image acquired after administering technetium-99m-sulfur colloid. Two foci of radiocolloid accumulation are visible in the left thoracic cavity.

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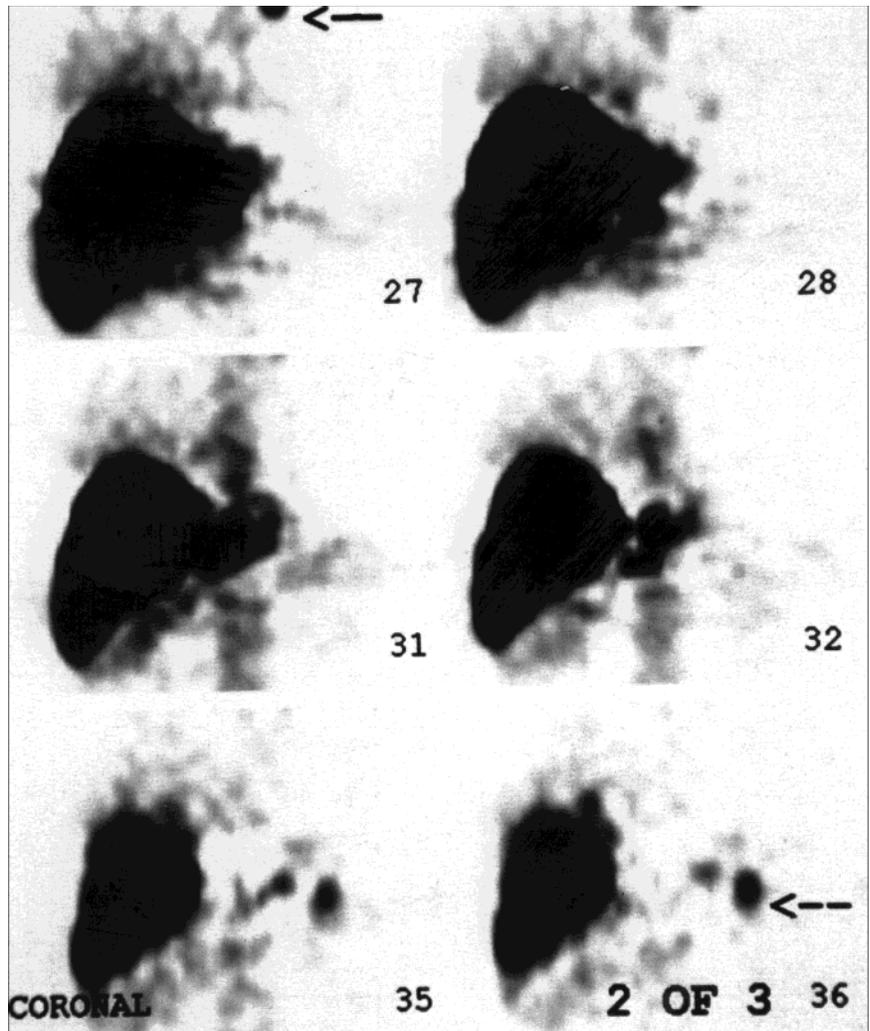
**FIGURE 2.** Right lateral (left) and right anterior oblique planar technetium-99m-sulfur colloid images revealed an accessory spleen, the third focus in this patient.

to the nuclear medicine department for a radiocolloid scan to localize functional ectopic splenic tissue, if present.

A routine planar liver/spleen procedure with  $^{99m}\text{Tc}$ -sulfur colloid was performed. Since unusual uptake was noted in the thorax, additional views incorporating this area were obtained. The images revealed 4 abnormal focal areas of increased radiotracer uptake. Two foci of different densities were in the inferior aspect of the left thoracic cavity (Fig. 1, solid arrow

shows the dense focus; broken arrow shows the less dense focus). A third focus was anterior and slightly superior to the cardiac notch of the liver (Fig. 2), and the fourth was in the splenic bed (planar image not included). CT also detected the 2 foci in the abdomen but failed to detect either of the thoracic foci.

A SPECT radiocolloid scan was performed approximately 1 wk later to rule out the unlikely possibility of physiologic clumping. Approximately 10 min after the injection of  $^{99m}\text{Tc}$ -sulfur colloid, transverse, sagittal and coronal slices were obtained using a dual-head camera (Vertex; ADAC Labs, Milpitas, CA) fitted with a very high-resolution collimator. Data was acquired in projections over a  $360^\circ$  orbit for 45 s per projection. All 4 abnormalities noted on the planar images also were identified on the tomographic slices. The coronal slices in Figure 3 show 1 abnormal area of uptake in the chest (solid arrow) and 1 in the splenic bed (broken arrow). The limited number of counts per projection resulted in poorer visualization of the lower density thoracic focus, although it was seen in several sagittal slices anterior to the more dense thoracic abnormality (images not included). Resolution of the abnormal focus anterior to the liver was best noted on the sagittal slices.



**FIGURE 3.** Coronal SPECT images acquired after administering technetium-99m-sulfur colloid revealed 1 thoracic focus (solid arrow) and the fourth accessory spleen in the splenic bed (broken arrow).

## DISCUSSION

Functional splenic tissue in the thorax is rare and is believed to be associated with splenosis rather than accessory spleen. Splenosis represents the autotransplantation of splenic tissue after accidental or iatrogenic injury. Disruption of the splenic capsule may result in fragments being seeded throughout the peritoneal cavity or less frequently to extraperitoneal locations, such as the thorax. As of 1993, there had been fewer than 20 cases of thoracic splenosis reported (9). Normand et al. (10) reported that cases of thoracic splenosis involved the visceral, parietal or mediastinal pleurae on the left side of the thorax.

Various imaging modalities have been used to determine the presence and location of accessory splenic tissue including  $^{99m}\text{Tc}$ -labeled sulfur colloid or heat-damaged autologous red blood cells,  $^{111}\text{In}$  oxine-labeled platelets, CT and ultrasound. Heat-damaged red blood cells have been shown to have a higher sensitivity (11) and specificity (12) than the radiocolloid in the diagnosis of accessory spleen. As demonstrated in this study, however, the ectopic splenic tissue was easily detected and localized with radiolabeled colloid. Although this technique does not differentiate between accessory splenic tissue and splenosis, it is believed that the focus found in the splenic bed of this patient represented an accessory spleen whereas the remaining 3 foci reflect splenosis.

SPECT imaging also should improve accuracy in localizing the ectopic tissue as compared with planar imaging. It is suggested that, regardless of the radiopharmaceutical, an initial planar study should be performed to survey the thorax, abdomen and pelvis. Once abnormal foci are detected, tomographic slices should be obtained over the area under consideration for better localization.

## CONCLUSION

The destruction and phagocytosis of antibody-coated platelets in ITP leads to thrombocytopenia and, if severe, may cause

iron-deficient anemia or life-threatening hemorrhage, including cerebral hemorrhage. Confirmation of ectopic splenic tissue in this disease is critical, as accessory splenectomy will be considered during patient management. As seen in this study, these ectopic structures may be multiple and widely disseminated, even crossing cavital borders into the thorax or lower pelvis. Additional thoracic and pelvic planar views followed by SPECT imaging should be obtained for more accurate detection and localization of ectopic splenic tissue when a recurrence of ITP is suspected.

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