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CT imaging of splenic sequestration in sickle cell disease

Received: 11 May 2000  
Accepted: 2 August 2000

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Abstract  Pooling of blood in the spleen is a frequent occurrence in children with sickle cell diseases, particularly in the first few years of life, resulting in what is termed “splenic sequestration crisis.” The spectrum of severity in this syndrome is wide, ranging from mild splenomegaly to massive enlargement, circulatory collapse, and even death. The diagnosis is usually clinical, based on the enlargement of the spleen with a drop in hemoglobin level by > 2 g/dl, and it is rare that imaging studies are ordered. However, in the patient who presents to the emergency department with non-specific findings of an acute abdomen, it is important to recognize the appearance of sequestration on imaging studies. We studied seven patients utilizing contrast-enhanced CT scans and found two distinct patterns – multiple, peripheral, non-enhancing low-density areas or large, diffuse areas of low density in the majority of the splenic tissue. Although radiological imaging is not always necessary to diagnose splenic sequestration, in those situations where this diagnosis is not immediately obvious, it makes an important clarifying contribution.

Introduction

Sequestration of blood in the spleen is a frequent occurrence in individuals with sickle hemoglobinopathies (SS, SC, and Sβ thal), resulting in what is termed “splenic sequestration crisis” [1, 2]. Sludging of the red blood cells in the microvasculature of the spleen results in trapping of blood in the red pulp spaces, which may be transient or lead to ischemia and infarction. Diagnosis is based on findings of acute or subacute enlargement of the spleen, often painful, associated with a hemoglobin that is more than 2 g/dl lower than the patient’s baseline. The spectrum of severity is wide, ranging from asymptomatic mild splenomegaly to massive enlargement, with circulatory collapse and even death. It is more common in younger children who have SS disease and older children and adolescents who have SC disease [3, 4], but may occur even in adults [3, 5]. Children who have had one episode are far more likely to have a recurrent event, and the morbidity from this complication is significant [4, 6–8]. Hospitalization and transfusions are almost always required. The diagnosis of splenic sequestration is usually clinical and it is rare that imaging studies are ordered. However, patients may present to their physicians or to an emergency department with pallor, lethargy, abdominal fullness and pain, tachycardia and tachypnea, non-specific symptoms of an acute abdomen. Without knowledge of the baseline physical examination and hemoglobin, the diagnosis may not be clear. An appropriate diagnosis may obviate the need for surgery. With this review we hope to alert the general physician and the radiologist to the varied appearance of splenic sequestration on CT imaging (the commonly used imaging technique in the evaluation of acute abdomen).
Table 1 Characteristics of seven patients

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Age (years)</th>
<th>Hgb</th>
<th>Presentation</th>
<th>Indication for imaging</th>
<th>CT findings (*indicates contrast study)</th>
<th>Follow-up/comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>8</td>
<td>SS</td>
<td>Left upper quadrant pain</td>
<td>Moderate splenomegaly</td>
<td>*Normal to slight enlargement; multiple peripheral low-density filling defects</td>
<td>Splenectomy</td>
</tr>
<tr>
<td>2</td>
<td>5</td>
<td>SC</td>
<td>Abdominal pain</td>
<td>Acute abdomen</td>
<td>Moderately enlarged, medial half showing diffuse low-density area</td>
<td>Splenectomy</td>
</tr>
<tr>
<td>3</td>
<td>2.5</td>
<td>SS</td>
<td>Fever, pain and abdominal distension</td>
<td>Massive, persistent splenomegaly</td>
<td>*Moderate-to-large spleen; large low-density non-perfusing filling defects</td>
<td>Follow-up CT 3 months later – normal</td>
</tr>
<tr>
<td>4</td>
<td>31</td>
<td>Sβ thal</td>
<td>Abdominal pain</td>
<td>Acute abdomen</td>
<td>*Normal to slight enlargement; multiple peripheral low-density filling defects</td>
<td>Unknown</td>
</tr>
<tr>
<td>5</td>
<td>16</td>
<td>Sβ thal</td>
<td>Recurrent sequestration, acute chest syndrome</td>
<td>Moderate splenomegaly</td>
<td>*Moderate enlargement; multiple peripheral low-density filling defects</td>
<td>Splenectomy</td>
</tr>
<tr>
<td>6</td>
<td>16</td>
<td>SC</td>
<td>Abdominal pain, acute chest syndrome</td>
<td>Acute abdomen</td>
<td>*Moderate enlargement; multiple peripheral low-density filling defects</td>
<td>Splenectomy</td>
</tr>
<tr>
<td>7</td>
<td>10</td>
<td>SS</td>
<td>Abdominal pain</td>
<td>Moderate splenomegaly</td>
<td>*Normal to slight enlargement; multiple peripheral low-density filling defects</td>
<td>Splenectomy</td>
</tr>
</tbody>
</table>

We will describe seven patients, three with SS, two with Sβ thal, and two with SC disease who presented with acute splenic enlargement or signs of acute abdomen and had different imaging studies. All seven had CT, three ultrasound, one MRI, and two radionuclide spleen scans. We will focus on the CT appearance of the spleen in patients with sequestration, because of the increased use of CT in the evaluation of the acute abdomen.

The diagnosis of splenic sequestration can also be made with sonographic evaluation. Many of the patients are sent for such an examination looking for gallstones when the diagnosis of the sickle hemoglobinopathy is well known to the referring physicians.

Sonography can readily show the splenic peripheral defects – hypoechoic – and also the larger hypoechoic areas. Its major drawback is the severe pain and abdominal tenderness.

Results

CT scans of the patients were reviewed with particular attention to the following characteristics:

1. Splenic size, which was graded as mild, moderate, or massively enlarged
2. The presence of filling defects on a contrast study or low-density areas on a non-contrast study
3. The size and location of these defects

Based on these criteria, two distinct patterns were identified. Five of the seven patients had mild-to-moderate splenic enlargement with multiple, small, peripheral areas of low density, which did not enhance with administration of contrast (Figs. 1, 2). The other two patients had larger more diffuse areas of low density, also non-enhancing on contrast administration. These larger, more diffuse defects appeared similar to those seen following splenic trauma. Follow-up scans of the spleen in two patients after recovery from the sequestration event demonstrated a return to a normal homogeneous appearance. These patterns were distinct from the single and persistent peripheral wedge-shaped defect that may be seen individuals who have splenic infarction [9].

Materials and methods

This review was performed in the setting of a Comprehensive Sickle Cell Center, where approximately 600 patients, both adults and children, with sickle cell disease are followed. We retrospectively reviewed the imaging studies of seven individuals with sickle cell disease who had severe splenic sequestration crises at various times in the past 10 years. Even though the total number of patients who had this complication, i.e., splenic sequestration crisis, is probably in the range of 10–15%, only a small number of cases are imaged because of clinical confusion as to the cause of the abdominal pain. Indications for imaging varied in these individuals, as shown in Table 1.