Acute massive splenic infarction with complete liquefaction of the spleen in sickle cell disease

To the Editor: The link between splenic sequestration and massive infarction has been noted by several authors. Large splenic infarcts have been reported in sickle cell trait patients with normal sized spleens, an event precipitated by hypoxia during high altitude flights in an unpressurised aircraft or during mountain climbing. Massive splenic infarction has also been reported in patients with other sickle cell variants, where splenomegaly is common mainly in Hb sickle cell disease (SCD), in Sβ+ thalassaemia and Hb SE disease. Many of these patients start with splenic sequestration syndrome, followed by clinical deterioration leading to splenic infarction. In SCD, infarction is reportedly induced by stressful conditions including severe generalized painful crisis, septicemia, the postoperative period, strenuous exercise and in the postpartum state. It has also been reported following high altitude travel, including flying in modern pressurized aircraft. Hence SCD patients with splenomegaly are advised against air travel when possible, and if deemed unavoidable, they are advised to have a pre-travel simple or exchange transfusion in addition to adequate hydration.

In a study of 134 SCD patients in Saudi Arabia, only two patients (1.49%) had massive splenic infarction. Repeated attacks of vaso-occlusion, which are usually small and often asymptomatic leading to splenic infarction, often occur in children and result in autosplenectomy. However a significant number of SCD patients in Oman (60%) have preserved splenic function with some splenomegaly, well into adulthood, making them liable for splenic complications including sequestration and infarction. The preserved spleen is due in part at least to the high prevalence of both alpha-thalassaemia (48%), and beta-thalassaemia (2.6%) in the country. Similar results in the temporal sequence of splenic dysfunction in SCD have been reported by others in the region. While splenic infarction is a well recognized complication of SCD, massive splenic infarction with complete liquefaction of the spleen is rarely reported. We report our experience with three patients with SCD who had massive splenic infarction among a large number of patients with SCD who had splenectomy (Table 1, 2).

All our patients with splenic sequestration had elevated HbF at a mean of 7.1% in general and 22% among the three patients with massive infarction. Noticeably, patients with massive splenic infarction had a significantly elevated HbF (P=.0001, unpaired t test). Although elevated HbF is usually compatible with a milder course for SCD, and in association with thalassaemia, helps to preserve the spleen into adulthood, high Hb F in these patients did not seem to protect them from getting splenic infarction or sequestration. On the contrary, this study suggest that a significantly elevated HbF may indeed be a predictor of high risk for massive splenic infarction when the spleen is preserved into adulthood, and under conditions that may lead to tissue hypoxia like stress, infections, hypoxia, strenuous exercise and high altitude. Sickling of erythrocytes occurs in efferent channels of the spleen, which sets a chain of reactions that progressively involves more efferent channels culminating in a large splenic infarction. Studies of blood flow through the spleen done during periods of acute splenic sequestration, verify that blood flow is greatly retarded and sluggish. Therefore, in the presence of the above precipitating factors, patients with SCD who develop acute splenic sequestration are at risk of

**Table 1.** Laboratory test results in sickle cell disease patients, including three patients with massive splenic infarction.

<table>
<thead>
<tr>
<th></th>
<th>Total, n=79</th>
<th>Pre-splenectomy</th>
<th>Post-splenectomy</th>
<th>Massive infarction group, n=3</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Hb (g/dL)</td>
<td>mean (SD)</td>
<td>mean (SD)</td>
<td>laboratory features</td>
</tr>
<tr>
<td>Pre-splenectomy,</td>
<td>6.4 (1.3)</td>
<td>6.2 (2.5)</td>
<td>156 (57)</td>
<td></td>
</tr>
<tr>
<td>(paired t test)</td>
<td>.001</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Post-splenectomy,</td>
<td>9.5 (1.0)</td>
<td>9.3 (2.2)</td>
<td>455 (238)</td>
<td></td>
</tr>
<tr>
<td>(paired t test)</td>
<td>.001</td>
<td>.001</td>
<td>.001</td>
<td></td>
</tr>
<tr>
<td>Massive infarction</td>
<td>7.33 (0.3)</td>
<td>8.8 (1.7)</td>
<td>139.7 (75)</td>
<td></td>
</tr>
<tr>
<td>group, n=3</td>
<td></td>
<td></td>
<td></td>
<td>22 (9.3), range, 11.6-29.4</td>
</tr>
<tr>
<td>Pre-splenectomy,</td>
<td>11.4 (1.04)</td>
<td>16.8 (5.4)</td>
<td>463 (122)</td>
<td></td>
</tr>
<tr>
<td>(paired t test)</td>
<td>.0137</td>
<td>.093</td>
<td>.009</td>
<td></td>
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</table>

*P value <.05 was considered statistically significant.
*p<.001 for mean Hbf between massive infarction group vs. rest of patients (unpaired t test)
developing acute splenic infarction, and those with a very high HbF and large spleen, are at a risk of developing massive splenic infarction.

Patients with massive splenic infarction usually present with severe left upper quadrant pain, fever and pallor. Frequently there is a progressive rapid fall in the hemoglobin level and platelet count. A high index of clinical suspicion is necessary to ensure early diagnosis and treatment of splenic infarction, especially when an SCD patient presents with the above symptoms, particularly following exposure to a stressful situation. This is now feasible by performing a simple ultrasound examination of the abdomen. Repeated ultrasound examinations would also facilitate the followup on the progress of this complication. Computerized tomography of the abdomen is of utmost help in demonstrating the extent of infarcts (Figures 1). While splenic infarcts due to other causes may appear as a wedge-shaped or multiple heterogeneous lesion, those in massive infarction present as a massive hypodense lesion nearly replacing the spleen and appear as poorly defined areas of low density to a normal appearing spleen. Furthermore, injection of a bolus of intravenous contrast sharply demarcates the area of infarction due to the contrast enhancement of the surrounding rim of normal tissue making the CT scan, one of the most useful modalities for the diagnosis.

Management of such complications is challenging to both physicians and surgeons. One of the major concerns is to rule out the possibility of a splenic abscess, as generally these patients are febrile. This could be achieved by performing an ultrasound-guided aspiration followed by culture of the aspirated fluid. These patients can be treated initially with a conservative

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Study group, n=79</th>
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<tbody>
<tr>
<td>Age (years)</td>
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<td>30</td>
<td>49</td>
<td>Mean age 13 years, range 2-49 years</td>
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<tr>
<td>Sex</td>
<td>F</td>
<td>M</td>
<td>M</td>
<td>Males, n=43 (54%) M:F:1:1.2</td>
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<td>Associated features</td>
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<tr>
<td>Fever at presentation</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
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<tr>
<td>Left upper quadrant pain</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>Pleural effusion</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>Consolidation</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
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<tr>
<td>Spleen size</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Dimensions (cm)</td>
<td>Not measured</td>
<td>20×40×5</td>
<td>17×11×9</td>
<td>Mean weight, 594 g, range 70-2200 g</td>
</tr>
<tr>
<td>Weight (g)</td>
<td>Not measured</td>
<td>2200</td>
<td>705</td>
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</table>

Figure 1. a. Contrast CT scan of patient 1 showing more than 80% splenic infarction, b. Contrast CT scan of patient 2 revealing a near complete infarction of the spleen enclosed by a rim of splenic tissue, c. Radionuclide scan of patient 1 showing very minimal splenic function, d. Splenectomy specimen of patient 2 showing a completely necrotic cystic spleen.
approach with appropriate broad spectrum antibiotics, and specific treatment initiated based on the culture report, followed by splenectomy after appropriate vaccinations. Since the functional residual splenic tissue in massive splenic infarction is minimal, splenectomy is not expected to worsen the hyposplenic state postoperatively. The recommended indications for splenectomy in patients with massive splenic infarction are persistent upper abdominal pain, pressure symptoms of a large spleen, accidental rupture and the concern of potential risk of infection, or the presence of an access of the infarcted splenic tissue. Splenectomy could, however, pose special technical problems due to the size of the spleen and surrounding perisplenic inflammation. Dense adhesions are usually seen between the spleen, diaphragm, stomach and perinephric region. This makes laparoscopic splenectomy a relative contraindication. Dissection could be partly facilitated by decompressing the cystic mass intraoperatively by aspiration, while taking precaution to avoid spillage of splenic tissue into the peritoneal cavity, as we did in our second case.

In conclusion, massive splenic infarction is rare, and constituted approximately 3.8% of patients who required splenectomy for splenic sequestration in this large single institution study. All our three patients with massive splenic infarction were adults with persistent splenomegaly well into adulthood, making them liable to this complication. An elevated HBf is an important risk factor that is significantly associated with massive splenic infarction. A potential complication of a massive infarcted spleen is splenic abscess. A high index of clinical suspicion supported by radiological evidence on ultrasound and CT scan would help early, prompt and appropriate management. Splenic aspiration may be warranted in some of these cases. These patients can be managed initially by conservative measures followed by splenectomy. The predominant indication for splenectomy would be persistent pain, pressure effects and the risk of infection of the infarcted splenic tissue.

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REFERENCES

Uterine sarcoma: a rare cause of uterine inversion
To the Editor: We describe a case of uterine inversion associated with endometrial sarcoma. Initially, the patient was thought to have a cervical mass, as she developed severe bleeding and the mass protruded outside the vagina. A CT scan was done before examination under anesthesia, with findings of a large uterine mass. We performed a total abdominal hysterectomy and bilateral salpingo-oophorectomy. A completely inverted uterus was found during surgery. Histology showed adenosarcoma of the endometrium. The tumor was limited to the endometrium. At the 5-year follow up, there was no clinical or radiological evidence of recurrent disease. Uterine sarcoma should be suspected in uterine inversion diagnosed in a postmenopausal woman. Most cases of uterine inversion are obstetric related and encountered during the puerperium. Non-puerperal uterine inversion is estimated to account for just 17% of all cases of uterine inversion. Benign uterine pathology may present as uterine inversion (submucosal myoma is a good example). Rarely, uterine inversion may complicate the presentation of uterine sarcoma. The differential diagnosis of uterine inversion in a postmenopausal woman should include uterine pathology, particularly sarcoma.

A 76-year-old woman was ad-