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Case Report

An unusual presentation of extra-hepatic portal venous obstruction

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Abstract

A 13-year-old girl presented with short duration of fever, left upper abdominal pain and massive tender splenomegaly. There was no evidence of liver dysfunction or significant history. Evaluation revealed multiple splenic infarcts with portal vein thrombosis. She developed splenic abscess and was managed conservatively with percutaneous drainage and antibiotics.

Key words: *Complications, Extra-hepatic, Portal hypertension, Splenic infarct*

Splenic infarct, an uncommon problem in children, is usually caused by hematological and thromboembolic phenomena [1]. Portal hypertension causes congestive splenomegaly, but splenic infarction is rare [2]. The clinical presentation of left upper quadrant pain and tender splenomegaly should prompt the suspicion of a splenic infarct. We report a child with extrahepatic portal venous obstruction (EHPVO) with an uncommon acute presentation.

CASE REPORT

A 13-year-old developmentally normal girl presented with complaints of fever, left upper quadrant abdominal pain and abdominal distension of 6 days duration. There was no history of jaundice, bleeding from any site, abdominal trauma or umbilical catheterization. History of contact with tuberculosis 2 years ago was present.

On general examination, she was toxic, pale and undernourished. On per abdominal examination, she had massive tender splenomegaly (8 cm below the left costal margin) with no hepatomegaly, ascites or dilated abdominal wall veins. A clinical diagnosis of splenic abscess was made, and she was started on broad-spectrum antibiotics with ceftriaxone and cloxacillin.

Laboratory investigations revealed mild anemia (hemoglobin [Hb] - 9.3 g/dl), and polymorphonuclear leukocytosis (total counts - 14,000 cells/mm^3 and polymorphs - 72%) with normal platelets (1.7 lakhs/mm^3) elevated erythrocyte sedimentation rate. Her liver function tests and coagulation profile was normal. Ultrasound (USG) abdomen with Doppler showed portal hypertension with cavernous malformation of portal vein and massive splenomegaly with hypoechoic areas in spleen, suggestive of splenic infarcts due to chronic venous stasis with no evidence of thrombosis in the hepatic veins, superior mesenteric vein and inferior vena cava on Doppler. Computed tomography (CT) abdomen confirmed the same and also showed multiple enlarged lymph nodes in the mesenteric, para-aortic, peri-portal and peri-pancreatic areas of insignificant size and patent splenic vessels (Fig. 1). Chest X-ray showed a hilar lymph node and montoux was strongly positive (20 mm). Hence, CT thorax was done which showed calcified mediastinal lymphadenopathy. Upper gastrointestinal (GI) endoscopy revealed Grade II esophageal varices, severe portal hypertensive gastropathy, and duodenopathy.

Etiological workup for splenic infarct was done. Prothrombotic studies including protein C, protein S, anti-thrombin III, lupus anti-coagulant and anti-phospholipid antibody were negative. Antinuclear antibody and anti- double-stranded deoxyribonucleic acid were negative, and Hb electrophoresis was normal. Viral markers including hepatitis B surface antigen, anti-hepatitis C virus, and human immunodeficiency virus were negative. Cardiac evaluation including electrocardiogram and echocardiography were normal. Septic screening done to rule out the cause of fever including serial blood cultures was negative. Bone marrow aspirate showed a reactive marrow and ruled out malignancy. She became afebrile and was sent home to review in 5 days.

Child was re-admitted within 1 week with 1 day duration of abdominal pain and abdominal distension. CT abdomen showed a splenic abscess of size 20 cm * 15 cm. Ultrasound guided pigtail catheter insertion was done, and 4 L of anchovy sauce colored pus was drained. Pus culture grew *Klebsiella pneumonia* (extended-spectrum beta-lactamases), and piperacillin-tazobactam was given for 2 weeks. Child improved

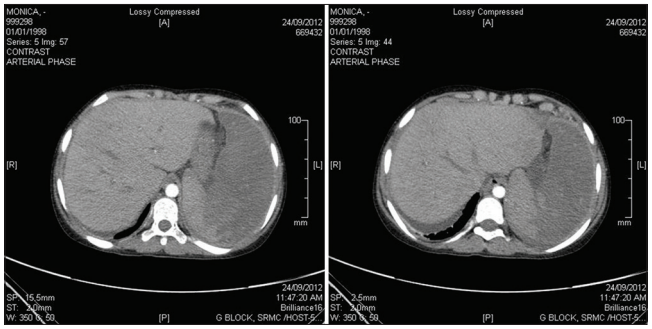

Figure 1: CT abdomen of the patient showing splenic infarct

significantly and was discharged on oral penicillin prophylaxis and oral propranolol. She was administered pneumococcal and meningococcal vaccines. At follow-up after 1 month, she was afebrile, gaining weight and doing well.

DISCUSSION

Extra-hepatic portal venous obstruction accounts for more than half of the cases of portal hypertension in children in India [3]. It usually presents as recurrent, well-tolerated upper GI bleed with no evidence of liver dysfunction and/or splenomegaly. The usual complications seen in EHPVO in children are severe hemetemesis, hyper-splenism and growth retardation. Our patient had an unusual presentation with the complication of splenic infarct and abscess.

Splenic infarcts per se are uncommon in children. The usual causes for splenic infarcts are hematological e.g. sickle hemoglobinopathies, malignancies such as acute and chronic myeloid leukemia and prothrombotic states including protein C and S deficiency, and anti-phospholipid antibodies [1,4,5]. Infections (e.g. malaria, Epstein–Barr virus infection) and embolic states like Infective endocarditis are also important causes. Splenic infarcts have been reported in adults with portal hypertension after cyanoacrylate injection for gastric varices [6]. Iatrogenic splenic infarction by partial splenic artery embolization, as an alternative to splenectomy to treat massive splenomegaly and hypersplenism, has also been reported [7].

The exact cause of splenic infarct in portal hypertension is unclear. The probable mechanism is hypoxia due to congestive splenomegaly with increased oxygen requirements. Also, anemia due to hypersplenism can lead to decreased oxygen carrying capacity [8]. Diagnosis can be confirmed by USG with Doppler and CT or magnetic resonance imaging abdomen. CT with contrast is the best non- invasive investigation to diagnose splenic infarction [8]. Etiological work up should be done to treat the underlying cause.

The complications of splenic infarct include abscess, pseudocyst, splenic rupture, and hemorrhage and sub-capsular hematoma. Treatment options include conservative or surgical management. Successful conservative management have been reported in both splenic infarct and abscess [8,9]. Conservative approach is useful especially, in critically ill patients. Also, preservation of some splenic tissue is possible. Surgical management by splenectomy is required in cases where complications such as abscess are present.

CONCLUSIONS

Splenic infarct and splenic abscess are rare complications and may be seen at presentation in portal hypertension. Clinical suspicion with appropriate imaging modalities will ensure early intervention.

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