Pediatric Splenic Torsion in an Orthotopic Spleen without Fixation Elements

A 4-year-old girl presented with acute left upper quadrant abdominal pain and anorexia of 4 days' duration. She had no relevant medical history and no trauma history was reported. Findings of the physical examination showed abdominal guarding and peritoneal irritation in the left upper quadrant. The rest of the examination was normal.

Blood tests showed leukocytosis, neutrophilia, moderate coagulopathy of the extrinsic pathway, and elevated acute phase reactants. An ultrasound study of the abdomen revealed the presence of a moderate amount of free fluid and splenomegaly with intraparenchymal hypoechoic areas. Computed tomography scan of the abdomen showed a massive splenic infarction with high suspicion of splenic hilum torsion; the image was compatible with a hilar plastron that seemed to pull on the tail of the pancreas and the stomach (Figure 1). An exploratory laparotomy was performed in which a partially medialized spleen was observed, without any type of fixation element, with a complete torsion of the splenic hilum (4 turns) (Figure 2). Despite the detorsion and tissue-reperfusion maneuvers applied, tissue necrosis was established and the organ could not be saved; therefore, a total splenectomy was performed. The patient is currently asymptomatic and was discharged following the prophylaxis guidelines for pediatric patients undergoing splenectomy.

Splenic infarction, which is infrequent in the pediatric population, may be attributable to multiple etiologies, such as

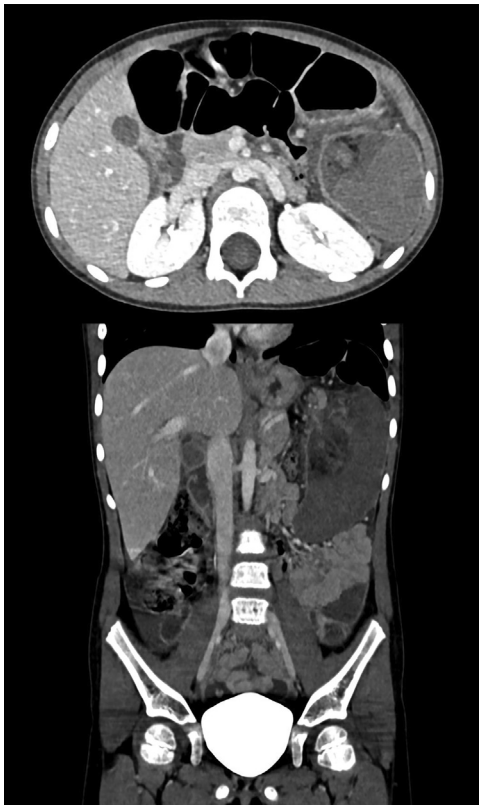

Figure 1. Computed tomography scan of the abdomen. *Top*, Axial view. *Bottom*, Coronal view. Spleen with homogeneous alteration in its density, with involvement of the hilum and with perisplenic free liquid.

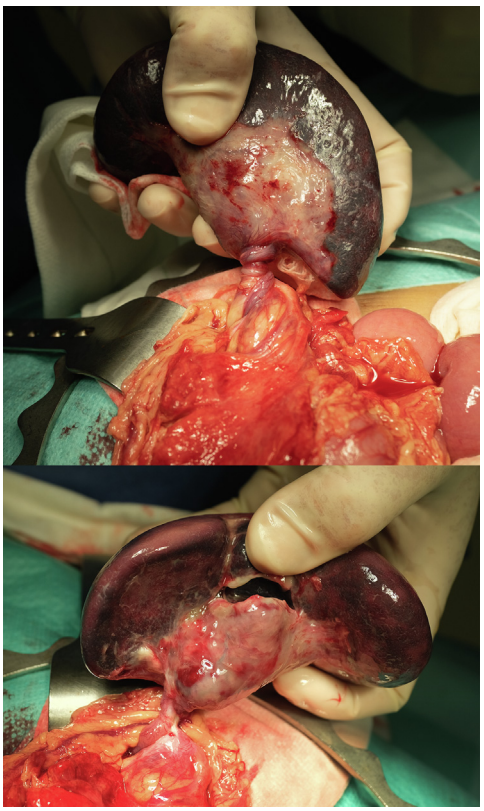

Figure 2. Intraoperative photograph. *Top*, Necrotic spleen, with hilar torsion of 4 turns. *Bottom*, After detorsion, cystification, and destruction of the splenic hilum is observed, without tissue recovery.

The authors declare no conflicts of interest.

J Pediatr 2022;251:209-10.

https://doi.org/10.1016/j.jpeds.2022.08.021

malaria, sickle cell disease,[1] and splenic hilum torsion, the latter usually in the context of a wandering spleen.[2] The spleen is located in the peritoneal cavity and is relatively fixed by different groups of ligaments. Weakening or absence of these ligaments often results in migration of the spleen through the abdominal cavity (splenoptosis or wandering spleen). This leads to an increased risk of splenic torsion. Although this phenomenon can be acquired, most cases of wandering spleen are congenital. Cases of spleen orthotopic torsion in spleen without fixation elements have been documented previously in adults.[3] In a patient with an acute abdomen and radiologic alteration of the spleen, the possibility of splenic torsion should be considered, even in an orthotopic position. The potential involvement of adjacent structures[4] and the fact that it is a time-dependent pathology[5] justify urgent surgical exploration in case of clinical and radiologic suspicion. ■

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Multiple Juvenile Xanthogranulomas

A 17-month-old boy presented with a 1-year history of asymptomatic, multiple yellow-orange papules and nodules that had gradually increased in number and size. Physical examination revealed numerous, discrete, yellowish-orange papules and nodules measuring 1-6 mm in diameter on the head, face (**Figures 1** and **2**), neck, and upper back. He had no signs of extracutaneous involvement. Laboratory investigations, including a complete blood count, serum lipid levels, renal panel, and liver tests, were all within normal range. Chest radiography, abdominal ultrasound examination, and an ophthalmologic examination revealed no abnormalities. Skin biopsy taken from his left forehead showed foamy histiocytes admixed with many Touton giant cells in the dermis (**Figure 3**), which were positive for CD68 and negative for S-100 protein and Langerin, thus confirming a diagnosis of juvenile xanthogranulomas (JXG). The patient did not receive further treatment, and was scheduled for annual follow-up. After 4 years of follow-up, most of his skin lesions regressed only leaving slight scar and hyperpigmentation, without internal organs affected.

JXG is a benign non-Langerhans cell histiocytosis of unknown etiology, most commonly occurring in infants and children.[1] It is characterized by solitary or multiple asymptomatic, yellowish cutaneous papules and nodules on the head and neck, trunk, and extremities.[1] Skin lesions of JXG often develop in the first year of life, although multiple lesions are more commonly present at birth or occur during the first 6 months of life.[1] Extracutaneous involvement of JXG including the eyes, central nervous system, liver, kidney, spleen, lungs, heart, and bone marrow is rare.[1,2] However, cutaneous manifestations of JXG may precede systemic involvement, which means regular periodic observation and systemic evaluation are essential.[1] A diagnosis of JXG is based on characteristic clinical features, histopathology, and immunohistochemistry. Typical histologic findings in JXG are dense infiltration of histiocytes and Touton giant cells with a wreath of nuclei surrounded by foamy cytoplasma in the papillary and reticular dermis.[2] Immunohistochemistry of JXG are typically strongly positive for CD68, and negative for S100 protein, CD1a, and Langerin.[3]

The authors declare no conflicts of interest.

J Pediatr 2022;251:210-11.

https://doi.org/10.1016/j.jpeds.2022.08.008