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An unusual case of abdominal pain in a 5-year-old with biliary atresia splenic malformation syndrome

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Describe role of Submitting/Presenting Trainee in this project (limit 150 words):

I, David Simon, the submitting author, was the primary resident responsible for the care of the patient, described in this case report, during her hospitalization. I obtained informed consent from the patient's mother, performed a comprehensive literature review regarding the patient's final diagnosis, and reviewed the relevant literature in order to create a case report documenting an unusual presentation of a rare pediatric condition (torsed accessory spleen).

Background, Objectives/Goal, Methods/Design, Results, Conclusions limited to 500 words

Background:
Accessory spleens or splenules represent a congenital focus of normal splenic tissue separate from the spleen. They are typically asymptomatic and observed in approximately 10-30% of patients at autopsy. Rarely, accessory spleens become symptomatic following torsion of their vascular pedicle, which compromises blood flow to the organ, leading to a potentially wide spectrum of clinical presentations. Cases may present with a variety of non-specific symptoms (e.g. vague abdominal pain, nausea, vomiting, and fever), recurrent abdominal pain, or with an acute abdomen. Diagnosis is typically made in the operating room as diagnostic imaging is often times unrevealing. Definitive treatment is almost always splenectomy. Herein, we describe an unusual presentation of a torsed splenule in a 5-year-old patient with biliary atresia splenic malformation syndrome.

Objectives/Goal:
The purpose of this case report is to highlight the variable clinical presentation and diagnostic challenge of a torsed accessory spleen. The goal is to raise awareness of this rare condition as well as to stress the importance of maintaining a broad differential diagnosis for pediatric patients with known laterality defects presenting with non-specific abdominal pain and symptoms. This case adds to the literature by being the first reported case of accessory spleen torsion in a patient with known biliary atresia splenic malformation syndrome.

Methods/Design:
Signed informed consent was obtained from the patient's mother for presentation of this case report. The medical record was then reviewed, and all relevant charts, radiology, and histology were utilized fin the creation of this report. Finally, a PubMed literature search was performed, and all relevant literature was reviewed.

Conclusion:
Accessory splenic torsion is a rare pediatric entity and should remain on the differential for pediatric patients presenting with both acute and non-specific abdomen pain, especially in the setting of known splenic malformation and/or other laterality defects. Once confirmed, conservative management could be considered, but operative management is most common.