

Case Report

# Torsion of an Accessory Spleen: A Rare Case and Review of the Literature

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#### Abstract

Torsion of an accessory spleen is a very rare entity that causes an acute abdomen in children, and it is difficult to accurately diagnose preoperatively. We herein report a case of acute abdominal pain in a 5-year-old boy caused by torsion of an accessory spleen. In this case, abdominal US detected the whorl of the twisted pedicle, which may be a hallmark to diagnose torsion of an accessory spleen. We must consider torsion of an accessory spleen as a differential diagnosis when encountering acute abdomen in children with a palpable mass.

Keywords: Accessory spleen; Torsion; Acute abdomen

## Introduction

An accessory spleen is a relatively common condition that is noted in 10-30% of autopsies [1]. Accessory spleens are asymptomatic in most cases [2]; however, once torsion of an accessory spleen occurs, it can be life-threatening, with severe infarction of the accessory spleen. Preoperative diagnosis is very difficult, even with modern imaging techniques [3]. We herein report a case of acute abdominal pain in a 5-year-old boy caused by torsion of an accessory spleen. We review the literature on this type of disorder, particularly concerning the etiology and diagnostic imaging of torsion of an accessory spleen.

### **Case Report**

A 5-year-old boy was admitted to our hospital with a 2-day history of left lower abdominal pain with fever and vomiting. His past medical history was unremarkable. On physical examination, a well-defined elastic hard mass with tenderness in the left lower quadrant was detected. Laboratory findings upon admission disclosed a white blood cell count of 11,600 /mm<sup>3</sup> and a C - reactive protein level of 7.50 mg/dL. The hematocrit was 29.0% and hemoglobin was 9.7 g/dL. Abdominal ultrasonography (US) revealed a well-defined, homogenous, hypoechogenic oval mass measuring 7.0 cm in diameter, and whorl of the twisted pedicle like the Whirlpool sign was also recognized (Figure 1). On color Doppler US, no flow within the mass or whorl could be detected. Enhanced CT demonstrated a normally enhanced spleen in the left upper quadrant and a  $7.0 \times 4.0$  cm low-density mass, surrounded with a thin wall which was weakly contrasted, in the left midabdomen adjacent to the intestine. The whorl of the twisted pedicle was located between the normal spleen and mass (Figure 2). We decided to perform emergent laparotomy on the day of admission because his abdominal pain gradually exacerbated.

At laparotomy, a dark-violet-colored mass  $(8 \times 5 \times 4 \text{ cm})$  with 720° torsion around a long vascular pedicle was surrounded by the greater omentum. On dissection of the surface of the tumor, the vascular pedicle was connected to the main splenic vessels. A normal spleen was located in the left upper quadrant, and we diagnosed the patient with torsion of an accessory spleen. After ligating the pedicle, we excised the accessory spleen (Figure 3). The postoperative course was uneventful. Histological examination of the mass was consistent with hemorrhagic and necrotic splenic tissues without malignancy.

### Discussion

Torsion of an accessory spleen is extremely rare. Patients range from infants to the elderly, but it mostly occur in children or in young adults and rarely in adults or the elderly. Alexander et al. [4] reported the first adult case with torsion of an accessory spleen in 1914 [4]. Settle et al. [2] reported the first pediatric case in 1940, and only 17 pediatric cases have been subsequently reported, including our case [2]. The youngest patient of the cases reported in the literature was a 14-dayold female infant [5]. In child cases, the average size of the accessory spleen with torsion was 6.6 cm in diameter, and the most common site was the greater omentum (58.8%). The clinical symptoms varied from vague abdominal pain in the case of a wandering accessory spleen with intermittent torsion, to fever, nausea, vomiting, and acute onset of severe abdominal pain in the case of infarction. The accompanying abnormalities found in the cases of torsion of an accessory spleen included left diaphragmatic hernia, situs inversus, and gut malrotation present in our case. All patients with torsion of an accessory spleen underwent excision of the strangulated accessory spleen, including laparoscopic excision [6,7].

The preoperative diagnosis of torsion of an accessory spleen is very difficult [8]. In all the reported cases the diagnosis was made at laparotomy. As in our case, detection of the whorl of the twisted pedicle by US, like the Whirlpool sign, may be a hallmark to diagnose torsion



**Figure 1:** Ultrasonography on admission. a) A well-defined, homogenous, hypoechogenic oval mass was noted, measuring 7.0 cm in diameter. b) The whorl of the twisted pedicle like the Whirlpool sign was also recognized (arrow).

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Received September 03, 2015; Accepted November 02, 2015; Published November 09, 2015

Citation: Kato K, Honda S, Minato M, Miklušica H, Okada T, et al. (2015) Torsion of an Accessory Spleen: A Rare Case and Review of the Literature. J Clin Case Rep 5: 632. doi:10.4172/2165-7920.1000632

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Page 2 of 2



Figure 2: Enhanced CT on admission. a) A normal spleen(S) was shown in the left upper quadrant. b) The whorl of the twisted pedicle was located between the normal spleen and mass(arrow). c) A  $7.0 \times 4.0$  cm low-density mass (M), surrounded with a thin wall which was weakly contrasted, in the left midabdomen adjacent to the intestine.



Figure 3: a)  $720^{\circ}$  torsion around a long vascular pedicle was noted. b) A dark-violet-colored mass (8x5x4 cm, 104 g) was surrounded by the greater omentum.

of an accessory spleen. Color Doppler US is useful to evaluate the degree of vascularization of an abdominal mass; however, the absence of flow does not differentiate a cystic mass from an ischemic lesion, such as torsion of an accessory spleen [4]. Enhanced CT findings are of a well-defined internal hypodense mass with marginal enhancement. However, these findings also cannot be used to differentiate cystic masses from ischemic lesions. Thus, detecting the whorl of the twisted pedicle is invaluable to diagnose torsion of an accessory spleen [9-15].

In conclusion, torsion of an accessory spleen is a very rare entity that causes an acute abdomen in children. It is difficult to accurately diagnose it preoperatively. Abdominal US should be performed to detect the whorl of the twisted pedicle to diagnose torsion of an accessory spleen. We must consider torsion of an accessory spleen as a differential diagnosis when encountering acute abdomen in children with a palpable mass [16-21].

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