Splenic Torsion in a Child with Polysplenia and Situs Inversus: A Very Rare Presentation

Manas R. Dash · Anand V. Upasani ·
Sudhir B. Chandna · Pradip B. Rathod ·
Kirti K. Prajapati · Dhiren N. Patel

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Abstract Polysplenia with situs inversus along with the torsion of spleen is a rare event. We report a case of a 12 year old child who presented with acute abdominal pain. There was an ill-defined mass in right hypochondrium and lumbar regions. The CT scan of abdomen revealed presence of multiple spleens with infarction of the largest spleen and features of complete dextroversion. Intraoperatively the infarcted spleen was found twisted around its vascular pedicle and removed.

Keywords Polysplenia · Situs inversus · Splenic torsion

Introduction

Polysplenia refers to presence of two or more spleens. Polysplenia syndrome refers to its association with various organ abnormalities in the abdomen and chest. Polysplenia syndrome was first described by Helwig in 1929 [1]. It is often recognized in childhood, though about 10 % cases present late during adulthood. Situs inversus is present in 20 % cases and more than 40 % have associated cardiac anomalies [1, 2]. Situs inversus indicates mirror image of the visceral location relative to situs solitus. Situs inversus with dextrocardia is more common in polysplenia, and 3–5 % of cases are associated with congenital heart disease [3].

Case Report

A 12-year-old boy presented with acute abdominal pain, nonbilious vomiting, and high-grade fever with chills and rigors. Abdominal examination revealed the presence of an ill-defined lump with tenderness and guarding in right hypochondriac and lumbar regions. He had leukopenia, thrombocytopenia, and presence of *Plasmodium vivax* schizonts in peripheral smear. X-ray of the chest/abdomen showed the presence of dextrocardia and reversed liver shadow. Ultrasonography revealed a mass of 108 mm×53 mm in size in the right hypochondriac region with avascular areas within it. Fever subsided in 2 days with antibiotics and antimalarials, but dull aching pain persisted. CT scan of the abdomen (Fig. 1) showed multiple spleens, largest measuring 70 mm×88 mm×110 mm in the right lumbar region and right iliac fossa. There was absence of postcontrast enhancement except in a small area at the upper pole, suggestive of infarction. Splenic and hepatic arteries were arising separately from the aorta. Echocardiogram confirmed dextrocardia without associated intracardiac anomalies.

On laparotomy, the liver was situated in the left hypochondrium, the cecum in the left iliac region, and the stomach on the right side. Multiple spleens (5 in number) were found on the right side of the abdomen adjacent to the stomach. The largest of these spleens which was occupying the right lumbar region was found to be dull and dusky in appearance with color changes suggestive of nonviability (Fig. 2). It had twisted around its vascular pedicle and was subsequently excised. The rest of the spleens appeared normal and were left intact. Postoperative recovery was uneventful and the patient was discharged after 6 days.

M. R. Dash · A. V. Upasani · S. B. Chandna (✉) · P. B. Rathod ·
K. K. Prajapati · D. N. Patel
Department of Pediatric Surgery, Smt. NHL Municipal Medical
College & Sheth Vadilal Sarabhai General Hospital,
36, Teenmurti Bunglows,
Thaltej, Ahmedabad 380 054, India
e-mail: sudhirchandna@gmail.com

Fig. 1 Preoperative CT scan of abdomen (with IV and oral contrast): **a** liver, **b** stomach with oral contrast, **c** viable spleen with postcontrast enhancement, and **d** nonviable congested spleen without postcontrast enhancement

Discussion

Polysplenia syndrome and situs inversus are associated with heterotaxy of various organs, congenital heart defects, malrotation, and other congenital anomalies [4]. Splenic torsion in polysplenia presenting with an acute abdomen is a rare event [5]. In a series of 1,413 splenectomies, only 4 cases were due to splenic torsion. Probably the first case of splenic torsion in a patient of polysplenia syndrome was reported around 1982 by Ackerman et al., which was managed conservatively [6]. Later on, our patient was operated for malrotation and during laparotomy the necrotic mass of splenic tissue was removed.

In polysplenia, the total splenic tissue is divided into two or more equal masses, whereas in accessory spleens there is a single normal size spleen and presence of one or more small

unequal masses of splenic tissue in addition. The location and number of spleens are variable; their number may range from 2 to 16. The common presentation is vague abdominal pain, nausea, and vomiting. A spleen with the narrow pedicle can twist around its pedicle, thus resulting in infarction [7]. Abdominal pain may be due to presence of malrotation and torsion of the spleen which may mimic acute appendicitis [8].

According to Moller, Nakib, and Anderson (1967), the polysplenic and asplenic syndromes are closely related developmental complexes due to a defect in early embryogenesis, resulting in abnormal body symmetry [6]. Although splenic torsion in polysplenia has been reported, we could not find any series or case report of splenic torsion in patients with polysplenia and situs inversus in spite of extensive review of English literature. Splenic torsion is a diagnostic challenge and the treatment of choice is splenectomy [8]. Patients with polysplenia are more prone to develop splenic torsion due to the presence of a narrow pedicle. In our case splenic infarction was confirmed after contrast-enhanced CT scan of the abdomen which showed nonenhancement of the infarcted spleen. The patient was also positive for vivax malaria which might have been responsible for splenomegaly in one of the spleens predisposing it to torsion. Splenic torsion commonly presents with acute abdominal pain, fever, and leucocytosis, but in this case in the initial period we found leukopenia and thrombocytopenia, which might be attributed to splenic sequestration.

A combination of malaria, right-sided abdominal lump, and abdominal pain should alert someone of the possibility of a right-sided spleen with torsion. Prompt splenectomy prevents late abscess formation and adhesions with the adjacent structures.

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Fig. 2 Intraoperative photograph showing **a** liver, **b** retracted right-sided stomach, **c** and **d** normal viable spleens, **e** twisted vascular pedicle, **f** nonviable congested spleen, and **u** umbilicus