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Case Report

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Asplenia Syndrome Complicated with Acute Gastric Volvulus: A Case Report and Review of the Literature

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ABSTRACT

Acute gastric volvulus in asplenia syndrome is a very rare presentation and only a few cases have been published in the literature. A 10-month-old baby girl with a history of complex congenital cyanotic heart disease presented with a history of repeated episodes of vomiting for 6 days duration. Her ultrasound scan showed a fluid-filled distended stomach, centrally placed liver and absent spleen. The upper gastrointestinal study showed right-sided stomach with mesentero axial volvulus. The gastric volvulus was complicated with a small perforation at the site of the obstruction that was identified during surgery. With the advancement of treatment for sepsis and surgical intervention for cardiac anomalies, the extra-cardiac complications related to other malformations in asplenia syndrome are becoming important issues. It is important to be vigilant about the common associations of asplenia syndrome and the possible rare complications related to visceral malposition.

Keywords: Asplenia Syndrome, case report, gastric volvulus

INTRODUCTION

The complicated cardiac anomalies and sepsis secondary to asplenia are identified causes for the early demise of patients with asplenia syndrome (1). With the advancement of treatment for sepsis and surgical intervention for cardiac anomalies, the extra-cardiac complications related to other malformations in asplenia syndrome are becoming important issues (1).

Acute gastric volvulus in asplenia syndrome is a very rare presentation (1) and only a few cases have been published in the literature (1-6). Due to the absence of the spleen and the associated splenic ligaments, patients with asplenia syndrome are thought to be more prone to gastric volvulus (1). Considering this predisposition, there are reports in the literature stating the importance of performing prophylactic gastropexy as the treatment for gastric malfixation in patients with asplenia syndrome (4). However, some are sceptical about routine screening for intestinal anomalies and recommend regular follow-up for asymptomatic patients (7).

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In this report, we describe a 10-month-old girl who presented with gastric volvulus as an extra-cardiac complication in asplenia syndrome. We also review the literature on previously reported cases of gastric volvulus in asplenia syndrome. This case highlights the importance of considering the common associations of asplenia syndrome and the possible complications related to visceral malposition, in order to diagnose and manage the patients promptly.

CASE REPORT

A 10-month-old baby girl with a history of complex congenital cyanotic heart disease presented

with a history of repeated episodes of vomiting for 6 days duration. The vomiting was pronounced after meals and with time, the frequency and the volume increased. During this time, the child developed progressive abdominal distension. She was diagnosed of having complex cyanotic heart disease with tricuspid atresia, obstructed infra cardiac total anomalous pulmonary venous return (TAPVR), Patent Foramen Ovale (PFO), large muscular Ventricular Septal Defect (VSD), and a small Patent Ductus Arteriosus (PDA). She also has had oesophageal atresia associated with tracheoesophageal fistula and imperforated anus and both were repaired surgically.

Imaging findings: Supine X-ray abdomen showed a gasless abdomen (Figure 1).

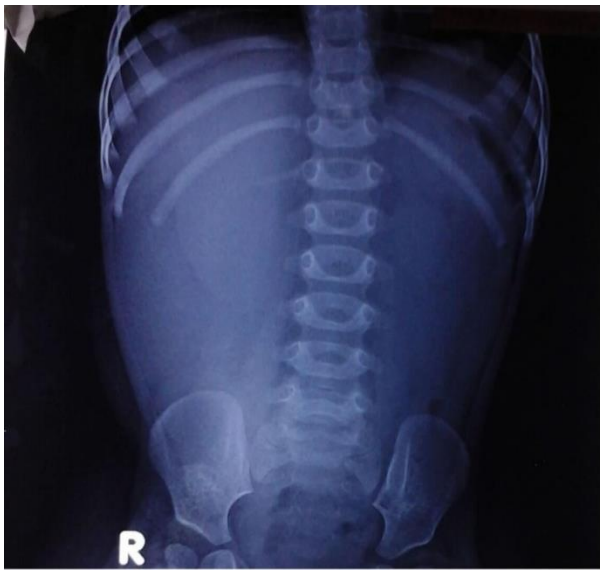

Figure 1: Supine X-ray abdomen showed a gas-less abdomen

There was no free air in the abdomen. A gastric air bubble was not identified in the left hypochondriac region.

Chest X-ray showed levocardia with mild cardiomegaly. Bronchial situs was difficult to assess on the plain radiograph.

The child was referred for an ultrasound scan of the abdomen. The ultrasound scan of the abdomen showed fluid fluid-filled distended structure in the upper abdomen. The liver was seen in the centre of the abdomen. The spleen and the gall bladder were not identified. Both kidneys were in normal position.

An upper gastrointestinal contrast study was performed to identify the exact site and possible cause for the proximal bowel obstruction. During the contrast study, a right-sided stomach with mesentero-axial volvulus was identified. The antrum was seen above the level of the gastroesophageal junction. There was gastric outlet obstruction. No contrast was seen entering the duodenum (Figure 2).

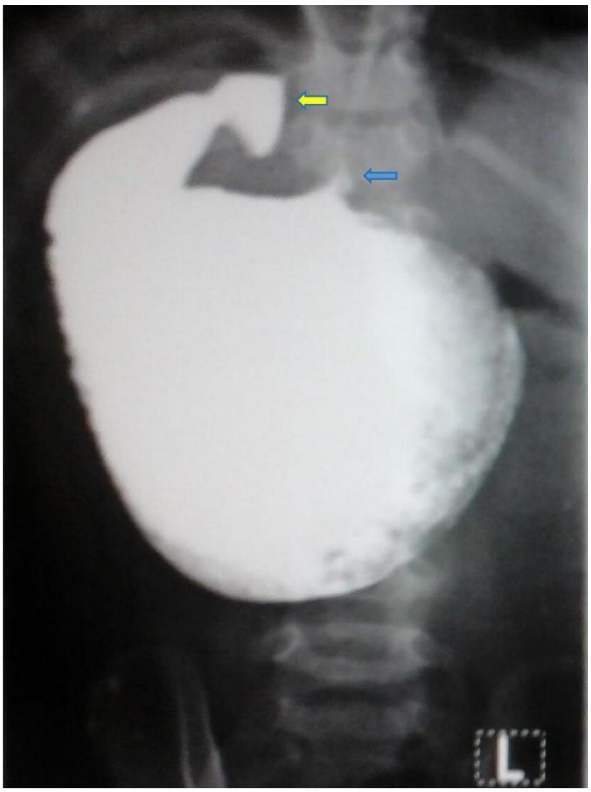

Figure 2: Upper Gastrointestinal Contrast Study showing mesenteroaxial volvulus. Gastro-oesophageal junction (blue arrow), Pylorus (yellow arrow)

The child was referred to a specialised pediatric surgery unit. During the surgery, the stomach was identified in the right side. The gastric volvulus was complicated with a small perforation at the site of the obstruction. The spleen and the gall bladder were not identified during the surgery. Seven days following the surgery, unfortunately, the child passed away due to complications.

DISCUSSION

Situs ambiguus, also known as heterotaxy syndrome, is a condition characterized by abnormal arrangement of organs in the chest and abdomen. Unlike the typical organ arrangement found in situs solitus or the mirror-image arrangement of situs inversus, situs ambiguus presents a disordered pattern. This condition is further divided into two subtypes: asplenia syndrome and polysplenia syndrome. In asplenia syndrome, individuals have both lungs with three lobes and eparterial bronchi, a midline liver, an absent spleen, and congenital heart defects. Conversely, in polysplenia syndrome, the lungs have two lobes and hyperarterial bronchi, midline liver, and multiple spleens are present (8).

The presence of complex cardiac anomalies along with asplenia and situs ambiguus led to the diagnosis of asplenia syndrome in our patient. Anomalous pulmonary venous return, common atrium, common atrioventricular canal, double outlet ventricle, and pulmonary stenosis are common combinations of complex cardiac defects associated with asplenia syndrome (1). The presence of complex cardiac anomalies and abnormal immune status due to asplenia have affected the overall survival of patients with asplenia syndrome (1). However, the advancement of cardiac surgeries and the use of prophylactic antibiotics have improved the survival of these patients (7).

Thus, while there is a reduction in the overall morbidity and mortality related to cardiac anomalies in asplenia syndrome, extra-cardiac complications, especially those related to intestinal anomalies are emerging as factors affecting the long-term outcome of patients with asplenia syndrome.

Among the reported gastrointestinal abnormalities associated with asplenia syndrome, also known as Ivemark syndrome, anomalies related to gut rotation, malposition and malfixation are common (2). Acute gastric volvulus is uncommon in the pediatric age group, yet, it carries a high mortality rate due to complications including gastric ischaemia and perforation (6).

Acute gastric volvulus is a rare presentation in asplenia syndrome and only a few cases are reported in the literature (1-6,9).

During the extensive literature search, eleven cases of acute gastric volvulus in asplenia syndrome were identified and the details of each case are given in Table 1.

The youngest age of presentation was at 1 month and the oldest child who had acute gastric volvulus was 3 years. One patient had recurrent episodes of gastric volvulus, which subsided without surgical intervention and later the child underwent an elective repair (3).

Due to the absence of the spleen, the stomach is malfixed and malpositioned in patients with asplenia syndrome. Further, a case series published in 1986 has reported a few intra-abdominal anomalies in these patients (3). These anomalies include the absence of gastrosplenic and gastrophrenic ligaments, deficient fixation of the pylorus and proximity of the pylorus to the cardia. Malfixed stomach along with the associated ligament anomalies are thought be the predisposing factors for the development of acute gastric volvulus in asplenic patients (1). Both intestinal malrotation and gastric malfixation appear to be contributing to the acute gastric volvlulus in asplenia (2). Thus, it is important that the relevant clinical management teams be vigilant of this potential serious extra-cardiac complication in asplenic patients.

The management strategy for intestinal malrotation in asplenia is still debatable. Some studies have recommended screening studies to identify intestinal malrotation and prophylactic gastropexy for gastric malfixation in asplenia syndrome (4). However, some studies have found that asymptomatic patients with asplenia

Table 1: Literature review of acute gastric volvulus in asplenia syndrome

Reference	Age and Sex	Presentation	Associated anomalies	Type of volvulus	Imaging findings	Management
Present case	10 months; Female	Acute onset vomiting and abdominal distention	PDA, TAPVR, PFO, Large muscular VSD, Oesophageal atresia with tracheoesophageal fistula, Imperforated anus, Asplenia	MA	USS: Fluid-filled distended stomach, Absent spleen and gallbladder, Symmetric liver. Upper GI series: GV	Gastropexy
Yang, Chin-Ying, et al[1]	11 months; Female	Frequent, vigorous non-bilious vomiting and abdominal distension	Right atrial isomerism, Common atrioventricular canal, the double outlet of the right ventricle, Pulmonary atresia, TAPVR, Asplenia	OA	Plain X-ray abdomen: Huge, cyst-like lesion over the upper abdomen Upper GI contrast study: Centrally located, dilated stomach with double air-fluid levels	Gastropexy Gastrostomy
Nakada, Koonosuke, et al[2]	10 months; Male	Abdominal distension	Single ventricle, Common atrium, TGA, Asplenia, PDPV, Malrotation of the intestine	GV	Plain X-Ray: Large single gastric air bubble in the middle of the abdomen	Gastropexy and Ladd procedure
Nakada, Koonosuke, et al[2]	36 months; Male	Non-bilious vomiting and abdominal distension	Single ventricle, Common atrium, Asplenia, Malrotation of the intestine	GV	Upper GI series: GV	Emergency laparotomy
Nakada, Koonosuke, et al[2]	1 month; Male	Frequent vomiting for 3 days	Asplenia, TGA, Pulmonary stenosis	GV		No surgical intervention
Aoyama, Koji, and Kazuma Tateishi[3]	17 months; Female	Recurrent non-bilious vomiting	Asplenia, Malrotation, Isolated levocardia, TAPVR	GV	Barium meal: Stomach left side	Elective repair
Aoyama, Koji, and Kazuma Tateishi[3]	14 months; Girl	Acute onset vomiting	Asplenia, Isolated levocardia, TAPVR	GV	Plain X-ray: distended stomach with fluid level. Upper GI series: Gastric outlet obstruction	Emergency laparotomy
Aoyama, Koji, and Kazuma Tateishi[3]	3 months; Female	Frequent non-bilious vomiting	Asplenia, Isolated dextrocardia, TAPVR		Plain X-ray: Distended stomach with fluid level. Upper GI series: Gastric volvulus	Emergency laparotomy
Koga, Hiroyuki, et al[4]	15 months; Male	Acute onset non-bilious vomiting and severe abdominal distention	Situs inversus, Asplenia, Single-atrium, Single-ventricle, Common atrium-ventricle valve, Pulmonary atresia	OA	Plain X-ray: hugely distended Gastric volvulus with a fluid level. Upper GI contrast study: Non-rotation of the intestine and a stomach in the right upper quadrant. CT: Hepatic symmetry.	Laparoscopic gastropexy
Marhuenda, C., et al[5]	18 months; Male	Acute onset mucous vomiting, abdominal pain, epigastric mass		MA	None reported	
Hung, Wan-Yu, et al[6]	13 months; Male	Acute onset vomiting	Asplenia	MA	Upper GI series	Reduction and gastroplexy
Oh, Sarah K., et al[9]	24 months Male	Acute onset vomiting and abdominal distention	Complex congenital heart disease, Heterotaxy, Asplenia	MA	Plain X-ray: Spherical gastric distension Upper GI Series: GV CT: GV, Asplenia	Gastropexy

(CT: Computed Tomography; GV: Gastric volvulus; MA: Mesenteroaxial; OA: Organoaxial; PDA: Patent Ductus Arteriosus; PDPV: Pre-Duodenal Portal Vein; PFO: Patent Foramen Ovale; TAPVR: Total Anomalous Pulmonary Venous Return; TGA:Transposition of the Great Arteries; VSD: Ventricular Septal Defect ;UGI: Upper Gastrointestinal Series)

syndrome has a relatively low risk of developing serious complications and thus, recommend close monitoring without prophylactic intervention (7).

CONCLUSION

This case report highlights the importance of considering the extra-cardiac complications, especially those related to intestinal malrotation associated with asplenia syndrome. The clinical team should be vigilant and prompt diagnosis and management is important to minimize the morbidity and mortality associated with acute gastric volvulus.

List of Abbreviations

CT	Computed Tomography
GV	Gastric volvulus
MA	Mesenteroaxial
OA	Organoaxial
PDA	Patent Ductus Arteriosus
PDPV	Pre-Duodenal Portal Vein
PFO	Patent Foramen Ovale
TAPVR	Total Anomalous Pulmonary Venous Return
TGA	Transposition of the Great Arteries
VSD	Ventricular Septal Defect
UGI	Upper Gastrointestinal Series

Author declaration

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