Case Report

Concurrent occurrence of a wandering spleen, organoaxial gastric volvulus, pancreatic volvulus, and cholestasis – A rare cause of an acute abdomen

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ABSTRACT

The concurrence of wandering spleen, organoaxial gastric volvulus, and pancreatic volvulus is very rare. They have been associated with symptoms such as severe abdominal pain, abdominal distention, and vomiting. However, the diagnosis remains complicated and any delay can result in ischemia and necrosis of the organs involved. In this case presentation, we present a unique case involving a 14-year-old girl who presented initially with acute abdominal pain. Assessment with enhanced computed tomography scan led to the diagnosis of wandering spleen, organoaxial gastric volvulus, and pancreatic volvulus, in addition to cholestasis, making it the first study to report on the simultaneous occurrence of this triad and cholestasis.

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Introduction

Wandering spleen, first described by Jozef Dietl in 1854, is a rare medical condition in which the spleen migrates from its usual position (peritoneal attachment) and thus assumes an ever wandering state, commonly to the pelvis or lower abdomen [1,2]. It is characterized by the absence or abnormal elongation of the spleen’s ligamentous attachments to the diaphragm, colon, and retroperitoneum [3]. Although wandering spleen and gastric volvulus share a common embryology, gastric volvulus complicating wandering spleen remains a rare association, with few cases reported in literature. In general, gastric volvulus occurs when the stomach twists on itself, causing a closed-loop obstruction. Organoaxial volvulus which is the most common type occurs when the stomach...
rotates 180° on its longitudinal axis (from cardia to pylorus) [4]. Furthermore, pancreatic volvulus is itself a rare condition, and as such, torsion of the pancreas accompanying wandering spleen is extremely unusual. Pancreatic volvulus occurs when the pancreatic tissue gets twisted in the splenorenal ligament during splenic torsion [5].

The simultaneous occurrence of wandering spleen, pancreatic volvulus, and organoaxial gastric volvulus is very rare, with few cases reported so far in literature. In this case report, we report on the simultaneous occurrence of this rare triad in addition to hepatic cholestasis in a 14-year-old girl, who presented with an acute abdominal pain. To the best of our knowledge, this is the first case presentation to report on the simultaneous occurrence of hepatic cholestasis together wandering spleen, pancreatic volvulus and organoaxial gastric volvulus.

Case report

A 14-year-old girl presented with a 5-day history of nausea, vomiting, loss of appetite, fever, headache, constipation, and central abdominal pain. She had had milder episodes of abdominal pains since childhood and had been previously admitted 4 times in the past 1 year.

She looked acutely ill at presentation, was in pain, and had a temperature of 37.6°C. Her blood pressure was 110/70 mmHg, her pulse 120 beats per minute. She had dry lips and sunken eyes with evidence of weight loss. Her abdomen was flat but with tenderness around her umbilicus. She had no guarding nor rebound tenderness. Her respiratory and cardiovascular examinations were otherwise normal. She had a white cell count of 8.1 × 10^9/L (83.2% granulocytes) and hemoglobin concentration of 12.7 g/dL. She had hypokalemia of 3.2 mmol/L and significant ketonuria. Her other serum electrolytes and kidney function tests were within normal limits.

Enhanced computed tomography (CT) scan of the abdomino-pelvis was done from the domes of diaphragm to the symphysis pubis. CT findings indicated that the stomach had rotated along its long axis with the antrum rotating anterosuperiorly and the fundus rotating posteriorinferiorly. The stomach was grossly distended. The spleen was located anteriorly and the pancreas was rotated left anterior laterally. There was minimal free fluid seen in the pelvis. There was a severe intra- and extrhepatic cholestasis with a common bile duct (CBD) having a width of up to 1.4 cm and the intrahepatic ducts up to 0.8 cm dilated. The cholestasis was most likely due to a twist in the mesentery compressing on the distal part of the CBD. There was an attendant gallbladder hydrops. The liver appeared normal in size and contour. No focal lesions were identified. The gallbladder was not fully distended and the adrenals were unremarkable. The kidneys were normal in size and there was no hydronephrosis or hydroureter. The bladder was of a smooth contour and the visualized lung bases were within normal limits. Based on these findings, a diagnosis of an organoaxial gastric volvulus, with volvulus of the pancreas, a wandering spleen, and cholestasis resulting from compression of the distal CBD by the mesenteric volvulus was confirmed.

On admission, she was empirically managed for enteric fever using Ceftriaxone and Sulbactam (1500 mg), intravenous rehydration with Ringer’s lactate solution and Sodium chloride. Hypokalemia was corrected with intravenous potassium chloride. Over the next 48 hours she had become drowsy, her vomiting became persistent, projectile, and bilious. Her abdominal pain became severe and colicky. She subsequently underwent surgery after the CT scan. By the fifth
postoperative day, she had a recurrence of vomiting, abdominal distension with tenderness, increased bowel sounds.

A repeat enhanced CT scan after postlaparotomy for an organoaxial gastric volvulus, pancreatic volvulus, wandering spleen and cholestasis, showed that the stomach pancreas and spleen had been repositioned in the normal locations. The cholestasis had resolved completely. The liver appeared normal in size and contour. The gallbladder was not fully distended and there was no evidence of intra- or extrahepatic ductal dilatation. The spleen, pancreas, and adrenals were unremarkable. There was moderate distention of the colon. The main pathology was a distal ileal intussusception with a severe dilatation of the prestenotic small bowel up to 4.9 cm. Based on these findings, a diagnosis of acute small bowel obstruction secondary to an ileo-ileal intussusception was made. Subsequently, the patient underwent a second surgery. Her recovery was uneventful and she was finally discharged on the sixteenth postoperative day.

Written informed consent was obtained from the parent to have the case details and accompanying images published.

Discussion

Wandering spleen has been characterized by the abnormal location of the spleen. This is due to the fact that the spleen under this condition lacks a fixed ligamentous attachment and is therefore prone to changing positions within the abdomen [6].
In the current case report, findings from the CT scan showed that the stomach was grossly distended and the spleen was located anteriorly (Figs. 1 and 2A). Majority of the cases are seen in women in their reproductive age (70%-80%), whiles one third of the cases reported are found in children, mostly older than 10 years (70%), as recorded in our case presentation [7]. The preferred imaging modality for the condition is CT, and its presence is identified on CT imaging as the absence of the spleen in the left upper quadrant or comma-shaped or ovoid abdominal mass [8,9]. The true incidence of wandering spleen is difficult to ascertain as a result of the rarity of the condition, but it has been estimated to be around 0.2%-0.5% [9,10]. The clinical presentation of wandering spleen is variable, however, patients normally present with acute abdominal pain due to acute torsion of the splenic pedicle with splenic infarction or ischemia. It may also be diagnosed incidentally on a routine abdominal screening [8,11–13]. The patient in our case report presented with acute abdominal pain, nausea, vomiting, and fever. Considering the fact that it was congenital, it may be due to developmental abnormalities of the dorsal mesogastrium.

Several studies have proven that wandering spleen and gastric volvulus share a common etiopathogenesis (laxity of the intraperitoneal visceral ligaments), and as such, even though they are single rare malformations, they are frequently associated [14–16]. A study reported that, out of 19 patients who were surgically treated for gastric volvulus, they recorded a concurrent wandering spleen in 16 of the cases [17]. Gastric volvulus as an entity is potentially life threatening, and it is recognized as an abnormal rotation of the stomach around one of its axis [18]. It is classified into 3 types based on the rotation of the axis: mesenteroaxial, organoaxial, and combined type. The organoaxial type, which was recorded in our case presentation is the most common, and it is when the stomach rotates around its longitudinal axis with an anterior rotation of greater curvature, which moves from bottom to top and from the left to the right [16,19]. Clinical presentation varies between recurrent volvulus and acute abdominal emergency, as recorded in our case report [20]. In determining organoaxial gastric volvulus in the present report, findings from the enhanced CT showed that the stomach had rotated along its longitudinal axis with the antrum rotating anterosuperiorly and the fundus rotating posteriorinferiorly (Fig. 2B). CT scans incorporating a radio-opaque contrast offer a very sensitive diagnostic option and is particularly useful in detecting asso-

Fig. 3 – Sixth (6th) postoperative day repeat enhanced CT scan of the abdomen postsurgery with coronal and 3D reformatting showing repositioning of the pancreas and the spleen and reversal of the organoaxial rotation.

Fig. 4 – Further postsurgery findings for the same enhanced CT scan showing a newly occurring bowel obstruction secondary to ileo-ileal intussusception.
associated abnormalities or clinical unclear cases. Primary gastric volvulus may be caused by congenital absence of the gastric ligaments or abnormal laxity, while secondary gastric volvulus, which is similar to our case study, is associated with other defects such as wandering spleen (50%-75% of cases), evantion of the diaphragm, gastric malposition or nonfixing, intestinal malrotation, and diaphragmatic hernia [18,19].

Another unusual entity that very rarely occurs concurrently with wandering spleen and gastric volvulus is pancreatic volvulus, with only a few cases reported in literature [5,14,21,22]. In our case presentation, the pancreas was found to be rotated left anterior laterally (Fig. 2C). This may be due to the twisting of the pancreatic tissue in the splenorenal ligament during splenic torsion, as reported in a similar case presentation [5].

The concurrence of this triad (wandering spleen, gastric volvulus, and pancreatic volvulus) with cholestasis is the uniqueness of this case presentation. Cholestasis was identified with the advanced CT scan as a severe intra- and extrapancreatic cholestasis with a CBD, and having the intrahepatic ducts dilated (Fig. 2B). To the best of our knowledge, this is the first case presentation to report on such association.

The patient in our case presentation was initially managed for enteric fever. The first surgery was indicated after the CT findings. The repeat CT scan after the surgery showed that the stomach, pancreas, and spleen had been repositioned in the normal location, and there was no evidence of intra- or extrapancreatic ductal dilatation (Fig. 3). The main pathology that necessitated a second surgery was a distal ileal intussusception with a severe dilatation of the prestenotic small bowel (Fig. 4). The patient was finally discharged on the sixteenth day of postoperation day after an uneventful recovery.

**Conclusion**

This paper reports on a very rare concurrence of a wandering spleen, organoaxial gastric volvulus, pancreatic volvulus, and cholestasis in a 14-year-old girl. The diagnosis of such cases is complicated and mostly requires the use of an enhanced CT scan to prevent any delay in starting appropriate management, which can result in ischemia and necrosis of the organs involved.

**Conflict of interest**

None.

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**Supplementary materials**

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2019.05.018.

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