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Atypical duodenal volvulus in an adult with heterotaxy syndrome and complete common mesentery

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Abstract

Intestinal malrotation is a rare congenital disability defined as a malposition of intestine in the abdomen. It can be an isolated condition or a part of a heterotaxy syndrome. Some physicians perform a routine prophylactic Ladd procedure to asymptomatic patient with intestinal malrotation to avoid midgut volvulus. We report an unusual case of 24 years-old woman suffering from chronic vomiting since childhood, admitted to the surgical unit for occlusion syndrome and dehydration. CT performed on emergency showed levocardia, heterotaxy visceral syndrome with polysplenia, interruption of inferior vena cave and complete mesenteric malrotation. The diagnosis of small bowel volvulus was made and patient underwent surgery. Laparotomy confirmed the volvulus by peritoneal band. Post-operative period was uneventful. There was no sign of recurrence after 18 months follow up.

Keywords

heterotaxy; intestinal malrotation; duodenal obstruction; volvulus

Introduction

Heterotaxy syndrome, also known as isomerism, is a rare situation characterized by partial organ inversion with random arrangement of the abdominal organs [1]. It affects approximately 1 in 10000 live births [2].

Malrotation of the intestine is an example of random distribution of an abdominal organ. In the absence of severe cardiac defect, heterotaxy and intestinal malrotation usually manifest by non-specific symptoms. For that reason, the diagnosis may be delayed until adolescence or adulthood as an incidental imaging finding or as the cause of acute intestinal obstruction [3-4]. Due to the high risk of midgut volvulus and despite controversies, surgical correction by Ladd's procedure is the criterion standard for all operative candidates with malrotation.

We report a case of an unusual small bowel volvulus due to peritoneal band in young woman with incidental imaging diagnosis of heterotaxy syndrome and complete commun mesentery.

Case Presentation

A 24-year-old woman presented to the hospital with a 12 hour history of sudden onset, sharp epigastric pain than began postprandially associated with nausea, biliary vomiting. She also reported generalized abdominal discomfort.

Past medical history revealed several episodes of biliary vomiting with non-specific intermittent abdominal pain since 7 years old. Last one was three months ago and complicated with severe dehydration requiring intensive care admission. She has no prior surgery or imaging.

On physical exam, she was afebrile and hemodynamically stable. Her abdominal examination revealed a depressible abdomen with epigastric tenderness and right upper-quadrant pain upon palpation, as well as involuntary guarding.

Laboratory work-up was remarkable for elevated white blood cell count of 13,4 x 109/L, hyperlipasemia (3xN).

Computerized tomography scan of the abdomen demonstrated evidence of heterotaxy syndrome included levocardia, polysplenia, left-sided liver, dextrogastria, interruption of the inferior vena cava, and intestinal malrotation (Figure 1). Even though there was a complete commun mesentery, CT showed small bowel obstruction with volvulus (Figure 2).

There was no evidence of ischemia, pneumatosis intestinalis or free air in the abdomen.

Her initial management consisted of a nasogastric tube, intraveinous fluids and antibiotics.

Patient was consented for an exploratory laparotomy after initial management failure.

Surgical exploration revealed abdominal situs invesus, polysplenia and numerous adhesions at multiple locations along the small bowel; one of them is causing extrinsic obstruction with volvulus (Figure 3). There was no sign of intestinal ischemia. Since there was a complete commun mesentery (SMV on the right and SMA on the left, small bowel on the right side of the peritoneal cavity and colon on the left side), intervention consisted of division of peritoneal bands and reduction of volvulus. The affected small bowel segment was deemed viable, avoiding resection. An appendectomy was performed due to its aberrant location. The patient's abdomen was closed primarily and she was transferred on surgical service.

Postoperative course was uneventful. Follow up at 1, 3, 6 and 18 months showed regression of clinical symptoms and no sign of recurrence.

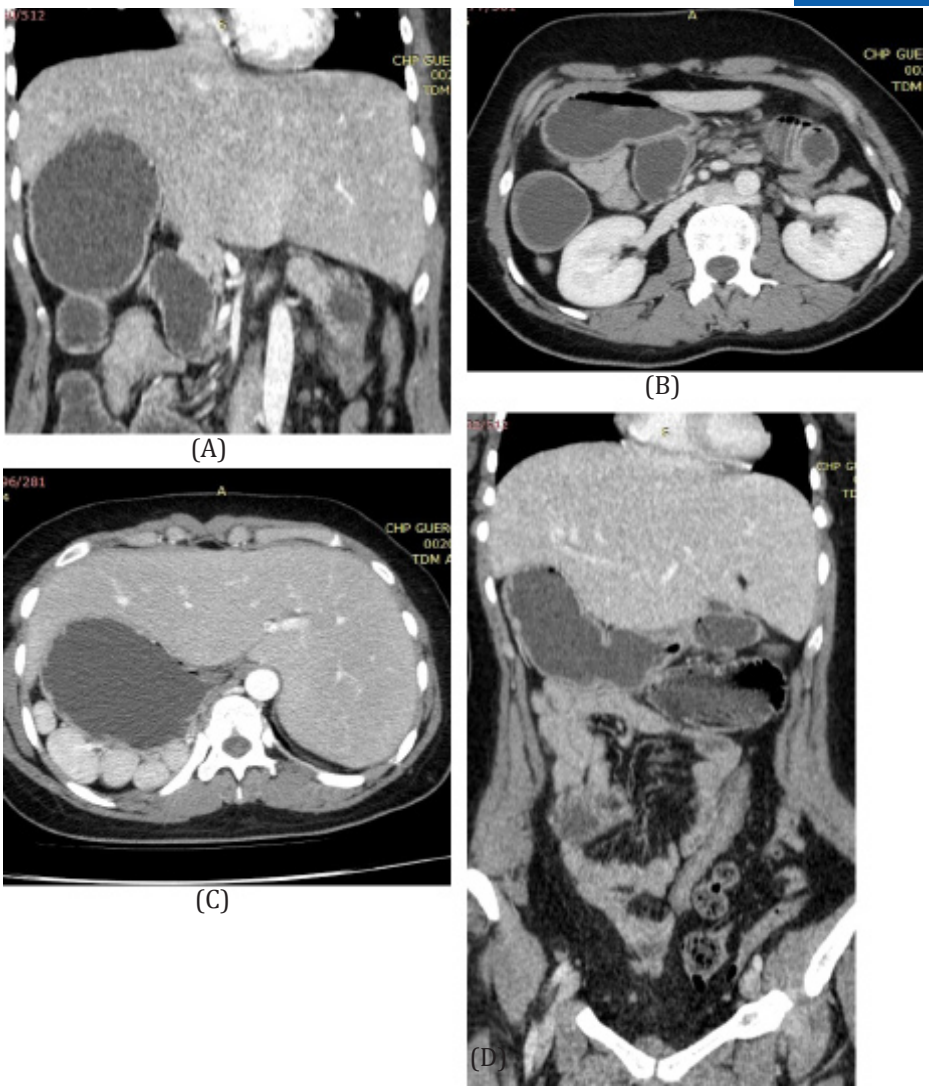

Figure 1: axial (B, C) and coronal (A, D) contrast- enhanced abdomino-pelvic CT demonstrated an interruption of inferior vena cava (A), abdominal situs inversus with polysplenia (B) and intestinal malrotation (C: IMV in the left of IMA, D: Small bowel in the right side and colon in the left side).

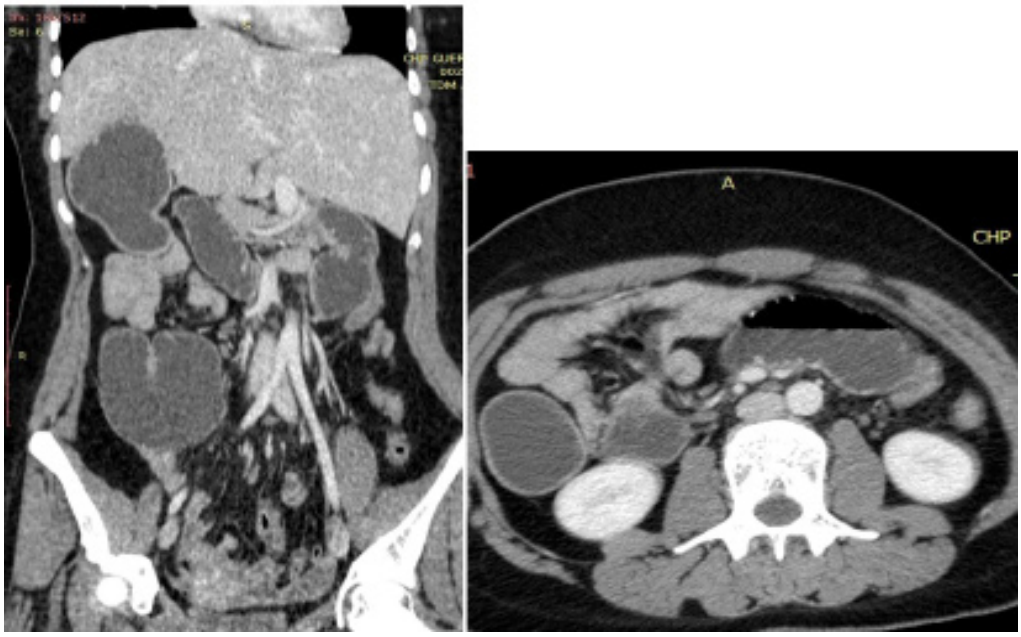

Figure 2: Coronal (A) and axial (B) images of an IV contrast-enhanced abdominopelvic CT show duodenal obstruction with volvulus (whirl sign).

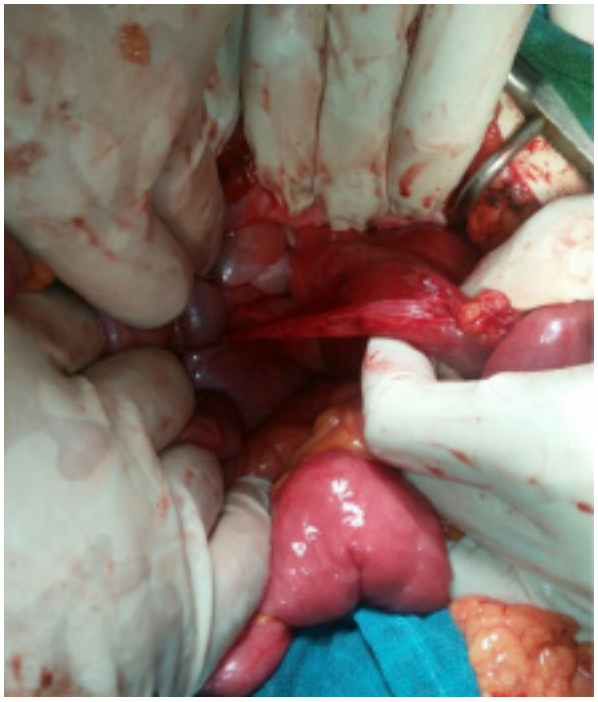

Figure 3: Per operative view demonstrated small bowel obstruction due to a peritoneal adhesion.

Discussion & Conclusions

Heterotaxy is incomplete lateralization disorder that results in any arrangement of the viscera across the left-right axis that differs from its normal position. Multiple variations were reported in literature. Burton el al. classified patients according to whether they had asplenia or polysplenia and at least 1 other known developmental association with heterotaxy and/or polysplenia syndrome [5]. According to literature, only 55.6% of patient presented with total anomalous systemic venous return [5].

Our patient presented levocadia, polysplenia, abdominal situs inversus, intestinal malrotation and interruption of inferior vena cava, so this syndrome was retained.

Malrotation of intestine is an example of random distribution of an abdominal organ. It's a congenital anomalous rotation of the gut around the superior mesenteric artery axis during embryogenesis. Diagnosis can be done early in life when patients are symptomatic or when they had imagine for other reason. Some individuals (0,1- 0,5%) progress to adulthood and remain asymptomatic or with non-specific symptom [6]. Our patient had a long history of abdominal pain and vomiting managed without further investigations, due to a low socio-economic level.

Hill et al. first described the risk of malrotation, defined at their study as a narrow mesentery and the presence of a Ladd's bands, as a factor of isomerism subtype: patients with left atrial isomerism were at significantly lower risk of malrotation compared to patients with right atrial isomerism [7]. Even if our patient fit in the spectrum of low risk of malrotation [8], characterized by her left isomerism and a complete common mesentery, she developed a small bowel volvulus due to a peritoneal band between Treitz angle and posterior peritoneum.

The Ladd's procedure remains the mainstay in surgical management. The procedure involves five steps: assessment of volvulus with counter-clockwise detorsion, Ladd's band (adhesion running from caecum to the right lateral abdominal wall) division, inter-mesenteric band division, appendectomy (prevention of future confusion) and finally placement of bowel in the corrected anatomic position.

Operative intervention versus conservative management remains controversial. Indeed, a recent study suggests higher morbidity and mortality at subsequent admission in operated patients [9,10].

In our case, the indication of the operation was guided by age, chronicity of symptoms leading several hospitalizations and the presence of acute small bowel volvulus. Patient was also at a higher lifetime risk on developing recurrent volvulus (1.8-8%) [11].

In conclusion, we present a rare case of an adult presenting heterotaxy syndrome with polysplenia, abdominal situs inversus, interruption inferior vena cava and malrotation characterized by a complete common mesentery complicated by a small bowel volvulus. Despite radiological imagine advances, the diagnosis of these entity remains a challenge in the adult population, radiologist must be able to recognize them to facilitate management and avoid complications.

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