CASE REPORT

An Imaging Pitfall of Atypical Malrotation

Tan Seu Kean[1,2], Noor Khairiah A. Karim[3], Ang Lee Nah[2], Tammy Teoh Han Qi[4]

[1] Department of Radiology, School of Medical Sciences, Health Campus, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan, Malaysia.
[2] Department of Radiology, Hospital Pulau Pinang, 10990 George Town, Pulau Pinang, Malaysia.
[3] Imaging Unit, Advanced Medical and Dental Institute, Universiti Sains Malaysia, 13200 Kepala Batas, Pulau Pinang, Malaysia.
[4] Department of Paediatric Surgery, Hospital Pulau Pinang, 10990 George Town, Pulau Pinang, Malaysia.

ABSTRACT

Atypical malrotation describes the situation where the ligament of Treitz is located to the left of the vertebral body midline but below the gastric outlet. It is usually asymptomatic and rarely complicated with midgut volvulus. A 2-years-old child who presented with bilious vomiting showed a normal superior mesenteric vein (SMV) and superior mesenteric artery (SMA) relationship with retroperitoneal position of the third part of the duodenum (D3) on ultrasound. The upper gastrointestinal (GI) contrast study demonstrated atypical malrotation, and supplementary plain abdominal computed tomography (CT) revealed midgut volvulus at proximal jejunum, consistent with operative findings. Although ultrasound is a helpful tool in diagnosing intestinal malrotation, a normal SMV and SMA relationship with retroperitoneal D3 does not exclude intestinal malrotation. An additional upper GI contrast study should be performed to evaluate the aetiology of intestinal obstruction if clinically suspicious. CT is reserved for doubtful cases if the former studies are inconclusive.

Keywords: Atypical malrotation, Pitfall, Ultrasound, Upper GI contrast study, Duodenojejunal flexure

Corresponding Author:
Noor Khairiah A. Karim, MRad
Email: drkhairiah@usm.my
Tel: +604-562 2516

INTRODUCTION

Intestinal malrotation is a generic term used to describe any anomalies in rotation and fixation of the small and large bowels during embryological development (1). It is classified into non-rotation, incomplete rotation, and reverse rotation (1). It also can be categorised into typical malrotation and atypical malrotation (malrotation variant) according to the location of the ligament of Treitz (1).

The ligament of Treitz is normally located to the left of the midline defined by the vertebral body, and at the level of the gastric outlet (1,2). Typical malrotation is described as the ligament of Treitz is absent or located to the right of midline; atypical malrotation is when the ligament is located to the left of midline but below the gastric outlet or pylorus (1,2).

Intestinal malrotation occurs in about 1 in 500 live births (3). The precise incidence of malrotation is challenging to determine as asymptomatic malrotation may be undetected throughout their lifetime (3). Atypical malrotation is not uncommon, seen in 46% (127 of 275) among the patients operated for malrotation (3). However, they are mostly asymptomatic and only 0.8% of them are complicated with volvulus and 7% has small bowel obstruction (3).

Intestinal malrotation can be diagnosed in ultrasound by abnormal relationship of the SMV and SMA with positive predictive value of 56% and negative predictive value of 87.8% (4). Ultrasound is also proposed to confirm the retroperitoneal position of the third part of the duodenum (D3) (5). On the upper gastrointestinal (GI) study, the normal position of the duodenojejunal (DJ) flexure or junction is located at the left of the left vertebral pedicle, at or above the inferior margin of the duodenal bulb on frontal views, and retroperitoneal (posterior) on lateral views (1).

CASE REPORT

A 2-years-old boy, who was an ex-premature 32-weeker with no other co-morbid conditions, presented with bilious vomiting for four days. His condition was complicated with acute kidney injury (AKI) secondary to dehydration and generalised seizure secondary to severe hyponatremia. Laboratory tests showed raised

serum creatinine (40μmol/L) and hyponatremia with serum sodium of 130mmol/L. The inflammatory markers were within the normal limit (white blood count was 12.5 x 10^3/μL, C-reactive protein was 3.2mg/L and serum lactate was 1.3mmol/L).

The abdominal radiograph (Fig. 1) showed distended stomach and duodenum with the presence of distal bowel gas. Ultrasound of the abdomen (Fig. 2) showed a normal superior mesenteric vein (SMV) and superior mesenteric artery (SMA) relationship. Clear fluid was administered via pre-existing nasogastric tube and demonstrated that the D3 was anterior to the vertebral body, crossing from right to the left between the abdominal aorta and the SMA.

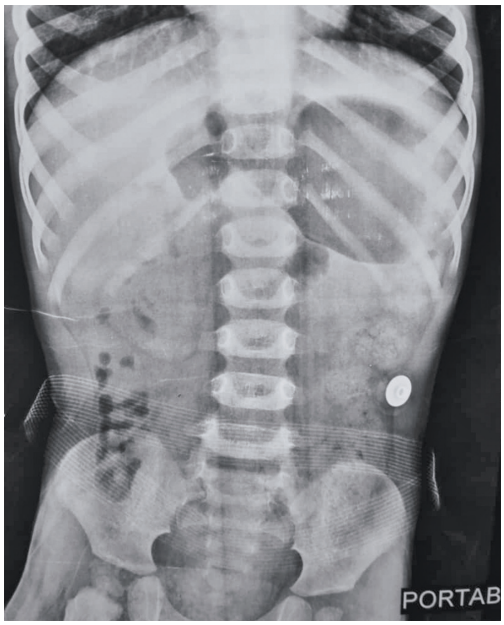

Figure 1: Abdominal radiograph. The stomach and the duodenum are distended with the presence of distal bowel gas.

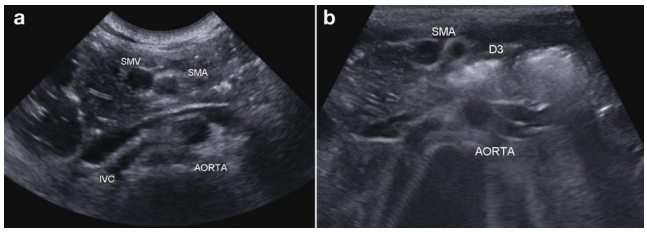

Figure 2: Abdominal ultrasound. (a) The SMV and the SMA are normal in relationship. (b) The D3 crosses from right to left between the abdominal aorta and the SMA when clear fluid was administered.

In view of inconclusive ultrasound findings, an upper GI contrast study was performed by selective duodenography method. There was back flow of contrast into the stomach due to contrast held up at the proximal jejunum, and absence of contrast flow distally despite after 15 minutes, suggestive of intestinal obstruction (Fig. 3). The second part (D2) and the third part (D3) of the duodenum were located in the retroperitoneum and superimposed on the lateral view. The DJ flexure was located anterior to the left pedicle of the L2 vertebral body, but inferior to the level of the gastric outlet. No corkscrew appearance was demonstrated.

A supplementary plain computed tomography (CT) abdomen (Fig. 4) was performed as both ultrasound and upper GI studies are inconclusive. Findings of CT were consistent with ultrasound and upper GI study. Additionally, the proximal jejunum swirled clockwise around the SMA, demonstrating the whirlpool sign. It is posterior to the distended gaseous stomach. There was minimal contrast opacification of the large bowel distally. The overall features are in favour of partial intestinal obstruction secondary to midgut volvulus. The child proceeded to emergency exploratory laparotomy.

Intraoperatively, the proximal jejunum at about 2cm distal to the DJ junction was adhered to a congenital band, causing the midgut to twist clockwise for about 270 degrees. The jejunum proximal to the adhesion band was dilated and all the distal bowels were collapsed.

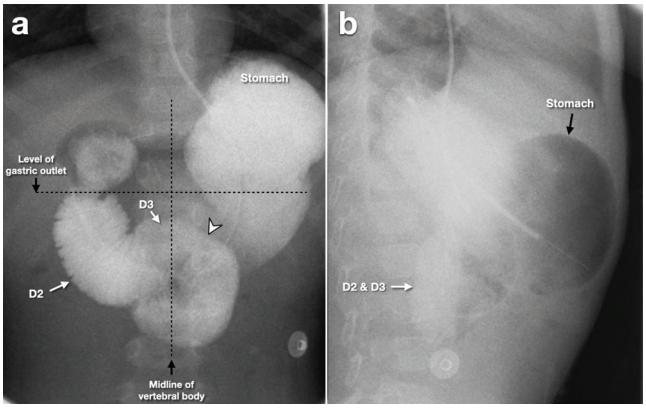

Figure 3: Upper GI contrast study in (a) anteroposterior view and (b) lateral view. (a) Classification of malrotation as typical and atypical malrotation based on the location of the DJ flexure in relation to the midline and level of the gastric outlet (1). The DJ flexure (arrowhead) is located to the left of the midline but below the gastric outlet indicates atypical malrotation. (b) The D2 and D3 (white arrow) are superimposed on each other and located at the retroperitoneum on the lateral view.

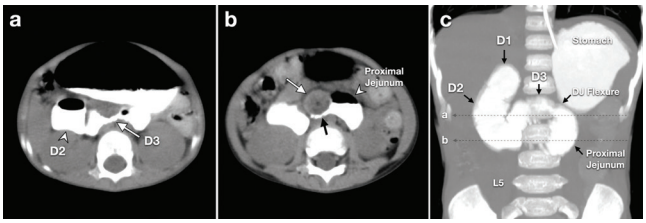

Figure 4: Supplementary plain CT abdomen in axial view (a) at the level of D3, (b) at the level of proximal jejunum (b) and (c) coronal view with maximum intensity projection (MIP). (a) The D2 (arrowhead) and D3 (white arrow) are located at the retroperitoneum. (b) Whirlpool sign (white arrow) with 'beaking' appearance of the bowel (black arrow) at proximal jejunum (arrowhead). (c) Dotted lines show the level of axial view in (a) and (b), respectively. The first part of the duodenum (D1), D2, D3, DJ flexure and proximal jejunum are identified and labelled with black arrows.

After derotating the bowel and releasing the adhesions, the duodenojejunal junction was to the right of the spine. Therefore, Ladd's procedure was performed. The ascending colon was in the right retroperitoneal region, and the caecum was in the right iliac fossa. The final diagnosis of jejunal obstruction secondary to congenital adhesion and volvulus with atypical malrotation was made.

The child recovered well with resolved AKI and electrolyte imbalance postoperatively. He was able to bowel open after feeding and discharged home well four days postoperatively.

DISCUSSION

Sudden onset of bilious vomiting in a previously healthy child is the hallmark presentation of intestinal malrotation with midgut volvulus (3). Although abdominal radiograph is often not specific, it is helpful to give us a clue on the level of obstruction by looking at the number of air-filled bowel loops. Gastric and duodenal distension with distal bowel gas indicates intermittent or partial proximal jejunal obstruction, except in a tight volvulus, it may show double bubble sign, mimicking duodenal atresia (1).

Atypical malrotation happens when the intestinal rotation has been interrupted between 180 and 270° during embryological development of the intestine (2). Consequently, the ligament of Treitz fails to reach its normal position at the level of pylorus on the left of the midline (2). Therefore, it will still have a retroperitoneal D3 with normal relationship of the SMV and SMA. This feature will cause false-negative or pitfall when excluding atypical malrotation by using ultrasound alone. In Taylor's study, he found that a child diagnosed with intestinal malrotation (2.6% of 38 patients) has retroperitoneal D3 on CT (4). Hence, using the retroperitoneum location of D3 to exclude malrotation will still have a low false-negative rate. This child is likely to have incomplete malrotation as the small bowels were located in the right abdomen and the colon in the left abdomen intraoperatively (4). In contrast to our case, the ascending colon and the caecum were in the normal right retroperitoneal region and right iliac fossa, respectively.

Nevertheless, ultrasound is recommended in diagnosing intestinal malrotation as it is readily available, non-ionising, low cost and provides a rapid diagnosis. We must be cautious that a normal relationship of the SMV and SMA with retroperitoneal position of the D3 does not exclude intestinal malrotation such as atypical or incomplete malrotation. Overall, the sonographic findings are less sensitive (67-100%) and specific (75-83%) in the detection of intestinal malrotation (5). Another advantage of ultrasound is detecting whirlpool sign which showing the SMV spirals clockwise around the SMA on colour Doppler study, indicating midgut volvulus(1,5).

Upper GI contrast study is the modality of choice in diagnosing intestinal malrotation with 93-100% sensitivity but low sensitivity (54%) in detecting the intestinal volvulus (2,5). A selective duodenography is recommended by controlling the first-pass bolus of contrast to better visualise the duodenal C-loop in frontal and lateral view. Upper GI contrast study can still diagnose atypical malrotation by demonstrating the abnormal location of DJ flexure at the left of midline but below the gastric outlet or pylorus, as shown in Fig. 3a (1). Gastric overdistension that may displace the normal DJ flexure and mimic abnormality is our concern in this case. Although there is absence of a typical corkscrew appearance of volvulus in our upper GI contrast study, proximal jejunal obstruction with 'beaking' appearance raises the suspicion of volvulus. This is best seen on axial view of CT in Fig. 4b (black arrow).

CT scan is not routine for diagnosing intestinal malrotation in children but is reserved for doubtful cases, to confirm the diagnosis, and look for complications(5). In our case, CT confirms the retroperitoneal D3 and atypical malrotation. The additional information is the proximal jejunal obstruction secondary to volvulus. CT is quick and minimally invasive, but it will subject the child to a higher dose of radiation and contrast media administration (5).

Surgical intervention for asymptomatic atypical malrotation remains controversial unless symptomatic or complicated with midgut volvulus (1,3). Although the risk of volvulus is low (estimated at less than 20%), there is a 15% risk of postoperative adhesive bowel obstruction, as well as a possibility of persistent pre-operative symptoms (3).

CONCLUSION

Atypical malrotation is often asymptomatic and remains undiagnosed unless complicated with midgut volvulus. Ultrasound is helpful in the early diagnosis of intestinal malrotation and midgut volvulus without ionising radiation. An abnormal relationship of the SMV and SMA or the absence of a retroperitoneal duodenum are solid indicators for the diagnosis of intestinal malrotation. However, we must be cautious that a normal relationship of the SMV and SMA with retroperitoneal position of the D3 does not exclude intestinal malrotation. An additional upper GI contrast study is warranted to exclude intestinal malrotation if no sonographic evidence of intestinal malrotation yet clinically suspicious. CT is reserved for doubtful cases when both ultrasound and upper GI contrast studies are inconclusive.

REFERENCES

1. Dassinger MS, Smith SD. Disorders of intestinal rotation and fixation. In: Pediatric surgery. Elsevier; 2012. p. 1111–25.
2. Mentessidou A, Saxena AK. Disorders of intestinal rotation and fixation. In: Pediatric Digestive Surgery. Springer; 2017. p. 245–54.
3. McVay MR, Kokoska ER, Jackson RJ, Smith SD. The changing spectrum of intestinal malrotation: diagnosis and management. Am J Surg. 2007;194(6):712–9.
4. Taylor GA. CT appearance of the duodenum and mesenteric vessels in children with normal and abnormal bowel rotation. Pediatr Radiol. 2011;41(11):1378–83.
5. Gymiz EB, Ayats AT, Feliubadaly CD, Martнnez CM, Tarragy AC. Intestinal malrotation–volvulus: Imaging findings. Radiol (English Ed. 2015;57(1):9–21.