CASE REPORT

Abdominal heterotaxy syndrome with annular pancreas, adhesive bands, and internal herniation: A rare case managed at a tertiary hospital in Lilongwe, Malawi

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Abstract

Heterotaxy syndrome is a disorder of embryonic development resulting in anomalous thoracoabdominal organ determination of the left–right axis. It presents with cardiac and extracardiac abnormalities associated with significant morbidity and mortality. We managed a 3-year-and-5-month-old boy with heterotaxy, who presented with bilious emesis, abdominal pain, distension, and obstipation. After resuscitation and nasogastric tube placement, an upright abdominal x-ray revealed a distended, right-sided stomach with an air–fluid level. Exploratory laparotomy confirmed this finding and revealed a midline liver, left-sided gallbladder, shortened large bowel, and internal herniation. Initial surgery reduced the small bowel and pexyed the large bowel to the right. The patient presented with obstructive symptoms 14 days after discharge, requiring a duodenojejunostomy to bypass an annular pancreas. Heterotaxy is a rare condition with variable anatomical presentations that may cause obstructive symptoms, often requiring surgical intervention. These cases can be complex, especially due to the potential for multiple coexisting sources of obstruction.

Keywords: abdominal heterotaxy, duodenojejunostomy, intestinal obstruction, paediatric surgery, Malawi

Introduction

Heterotaxy syndrome is a disorder of embryonic development resulting in anomalous thoracoabdominal organ determination of the left-right axis.[1] The clinical manifestations of heterotaxy syndrome include cardiac and extracardiac abnormalities.[2] Much of the literature on heterotaxy syndrome focuses on the cardiac sequelae, given the potentially life-threatening nature of the cardiac defects and the potential need for immediate, complex medical and surgical care.[3],[4] Extracardiac manifestations contribute significantly to morbidity and mortality and occur in a considerable percentage of heterotaxy syndrome patients.[5] Recent publications concerning the extracardiac features of heterotaxy syndrome have attempted to further characterize the syndrome and prepare physicians for its treatment or surgical management.

A subset of the literature focuses on the intra-abdominal extracardiac manifestations of heterotaxy syndrome. This literature has described asplenia, polysplenia, midline liver, extrahepatic biliary atresia and hypoplasia, pancreatic abnormalities, oesophageal atresia, hiatal and diaphragmatic hernias, right-sided stomach, bowel atresia, and malrotation of the intestine.[1],[6]-[9] These abdominal abnormalities can be asymptomatic or present with obstructive symptoms, oral intolerance, or abdominal discomfort. These symptoms depend, in part, on the degree and type of anatomic abnormality. There is an active effort to characterize the appropriate approaches for screening, operating, and managing the abdominal manifestations of heterotaxy syndrome.[10]-[12].

We report a case of heterotaxy syndrome that consisted exclusively of abdominal manifestations presenting as obstruction and requiring complex surgical care. This article reviews the presentation, management, and surgical considerations of abdominal heterotaxy syndrome in the context of intestinal obstruction.

Case presentation

Initial encounter

A 3-year-and-5-month-old Malawian boy was referred to our tertiary care centre from a district hospital after 3 days of bilious, projectile emesis (approximately 6 episodes per day after oral intake), 4 days of obstipation, generalized abdominal pain, and abdominal distension. The patient was born via uncomplicated, spontaneous vaginal delivery to a 25-year-old mother who attended 5 antenatal care visits and took antenatal supplements and medications. He cried

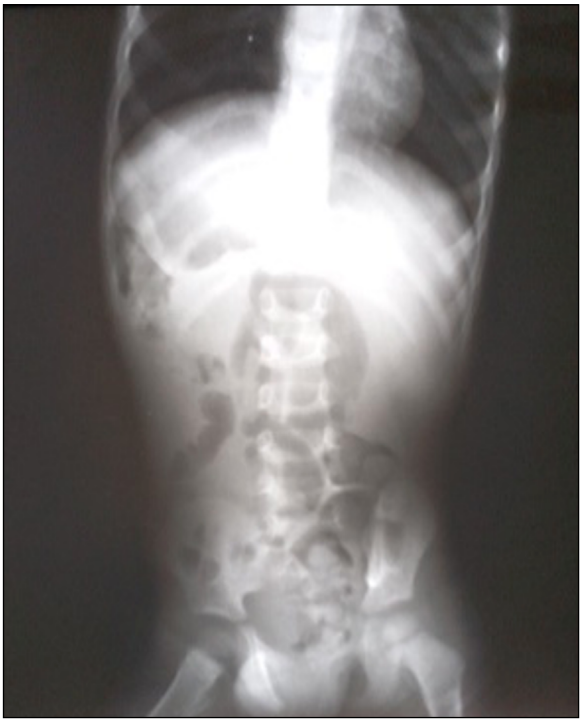

Figure 1. An erect x-ray revealed a right-sided gastric bubble, which was confirmed by nasogastric intubation.

upon delivery, was able to breastfeed shortly after delivery, and exhibited no birth defects. He went on to achieve normal developmental milestones and receive all immunizations on time. At 2 years and 8 months of age, he was admitted for tonic-clonic seizures in the context of a severe malaria infection. The child had not previously undergone any surgical procedures.

The patient's family history was significant only for asthma and hypertension.

When the patient presented to this tertiary care centre, there was no history of fever or jaundice. The patient was afebrile, tachycardic, and normotensive. On examination, he was lethargic with muscle wasting (weight of 14 kg, 25th percentile). His abdomen was distended with tenderness across all 4 quadrants. There was no rebound tenderness or guarding, bowel sounds were normal, and the rectal vault was empty.

Initial management

After resuscitation and nasogastric tube placement, he underwent an upright abdominal x-ray that revealed a distended, right-sided stomach with an air–fluid level (Figure 1). There were no echocardiographic abnormalities. The decision was made to proceed with an exploratory laparotomy. There were several pertinent intraoperative findings. These included an obstruction at the duodenojejunal junction secondary to adhesive bands, a proximal jejunal haematoma, an internal herniation of the small bowel behind the duodenum, a right-sided stomach, midline liver, and left-sided gallbladder (Figure 2). Extensive adhesiolysis was performed to release the bands, the small bowel was reduced, and the

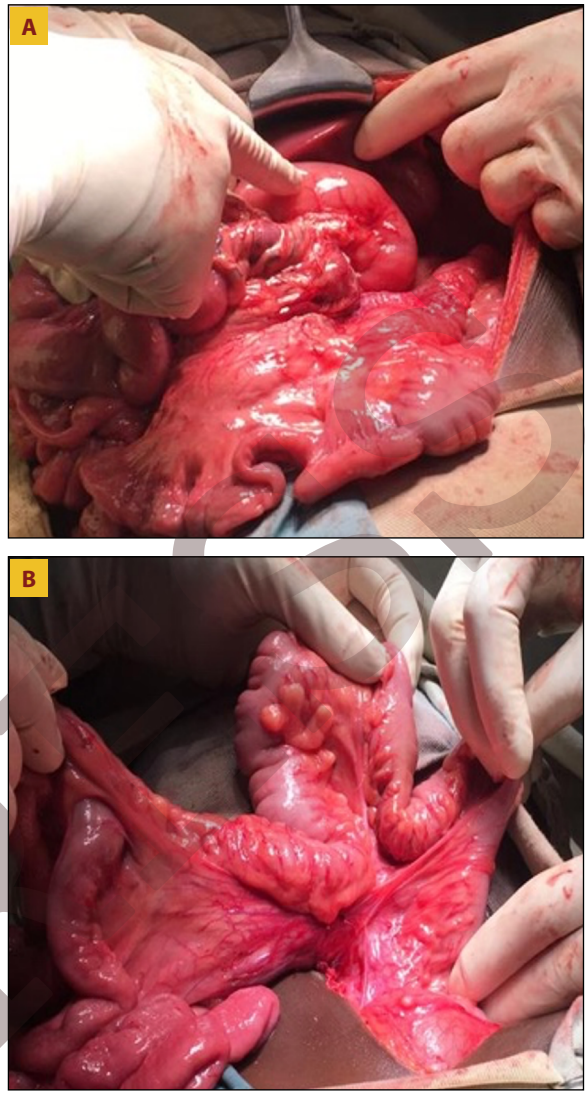

Figure 2. (A) A right-sided stomach and (B) adhesive bands were observed intraoperatively.

large bowel was pexyed to the right abdominal wall to restore its correct anatomic position. The patient's postoperative course was unremarkable, and he progressed appropriately, tolerating a diet after 24 hours and passing flatus on postoperative day 4. Following this uneventful recovery, the patient was discharged.

Second encounter

After 14 days, the patient returned to the emergency department with projectile, bilious emesis. The patient had been vomiting for 3 days (2-4 times per day after ingestion of food). The emesis was accompanied by severe left lower quadrant abdominal pain that was relieved by squatting. The patient complained of constipation, averaging 1 stool every 3 days.

On examination, he was alert and afebrile, with a normal heart rate. However, his weight had dropped from 14 kg to 12.5 kg. His abdomen was slightly distended, soft, and nontender. The previous incision site had healed. His full blood count and basic metabolic panel were notable for

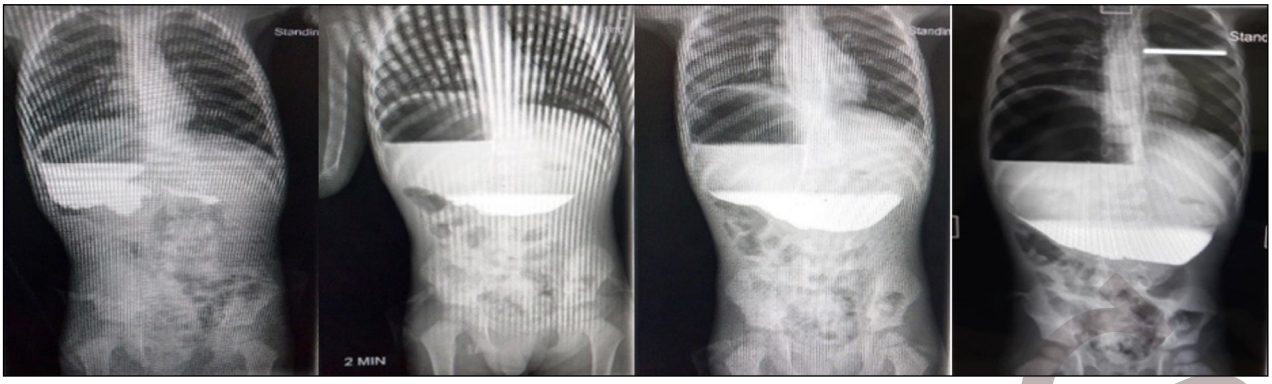

Figure 3. An abdominal series demonstrated significant gastric distension.

The results at 0, 2, 5, and 10 minutes are pictured without any passage of contrast to the small bowel, suggesting a proximal obstruction.

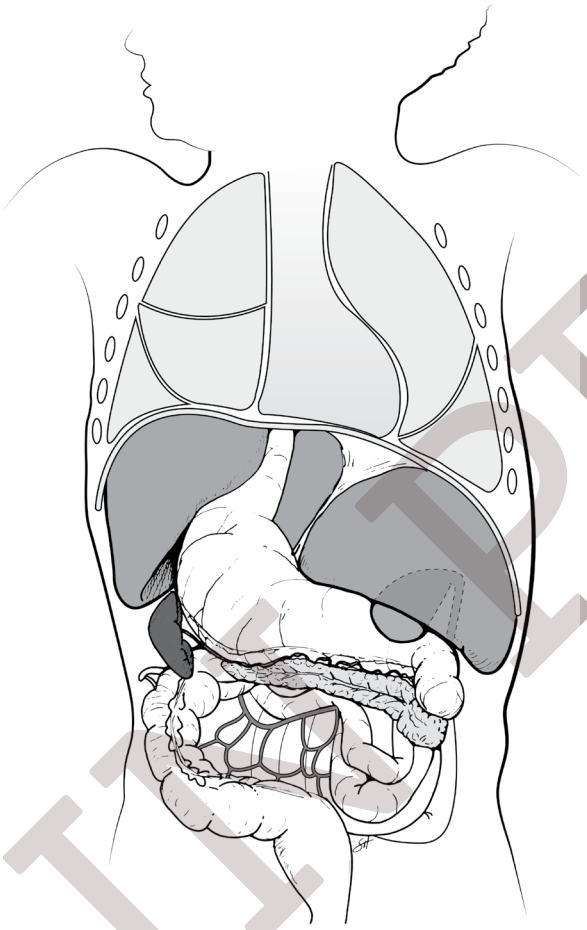

Figure 4. The patient's anatomic anomalies included a midline liver, right-sided stomach, left-sided gallbladder, diminutive spleen, annular pancreas, adhesive bands, widened mesentery, and short colon.

anaemia (haemoglobin, 10.9 g/dL), hyponatraemia (sodium, 125 mmol/L), hypokalaemia (potassium, 2.6 mmol/L), and hypochloraemia (chloride 76 mmol/L). An abdominal series showed an enlarged right-sided gastric bubble and air–fluid levels in the stomach (Figure 3). A contrast study raised concerns for a proximal small bowel obstruction, as the contrast medium did not progress beyond the stomach after 10 minutes.

Secondary intervention

The patient underwent a second exploratory laparotomy, where the previously noted anatomical variants were noted (Figure 2), including an enlarged right-sided stomach, midline liver, left-sided gallbladder, and a left-sided duodenojejunal junction with a broad mesentery. Additionally, a shortened large bowel, a small right spleen, and a right-positioned pancreatic tail were observed. A significant finding during this operation was the presence of an annular pancreas encircling and possibly obstructing the duodenum (Figure 4). To address these issues, adhesions were lysed, and the shortened large bowel was repositioned to the right side of the abdomen, with the caecum situated in the right upper quadrant. The procedure was completed with the construction of a single-layer duodenojejunostomy using 4-0 PDS (polydioxanone sutures).

Outcomes

Postoperatively, the patient was admitted to the paediatric high-dependency unit. His immediate postoperative course was characterized by poor urine output (<0.5 mL/kg/h) and suboptimal oxygen saturation levels (approximately 80%). After 72 hours, oxygen saturation and urine output normalized, and the child was transferred to the general ward. The patient received maintenance intravenous fluids for the first 4 days, then started sips of oral rehydration solution. He progressed to clear liquids and eventually a full diet. The child was discharged with scheduled follow-up in the paediatric surgery clinic. At the 2-month visit, the patient was doing well and had gained weight.

Discussion

Increasingly, physicians are attempting to diagnose heterotaxy syndrome in the prenatal setting via ultrasonographic detection and characterization of cardiac abnormalities.[13] Early detection, in turn, assists in the appropriate provision of prenatal counselling and timely medical care during the neonatal period. However, there has been evidence that heterotaxy syndrome may occur with exclusively abdominal

manifestations.[14] In this particular case, the patient had no stigmata of cardiac abnormalities on physical examination, x-ray, or echocardiogram. He exhibited only abdominal manifestations of the syndrome, including an enlarged right-sided stomach, midline liver, left-sided gallbladder, left-sided duodenum, left-sided duodenojejunal junction, broad small bowel mesentery, shortened large bowel, small spleen, and small pancreas. Abdominal heterotaxy syndrome, therefore, requires the paediatrician and the paediatric surgeon to be mindful of certain practice-based considerations.

These considerations may include basic changes, such as the inclusion of heterotaxy syndrome in the differential diagnosis when a patient presents with bowel obstruction, regardless of the age of presentation. However, more profound implications exist. For example, exclusively abdominal manifestations of heterotaxy syndrome in a resource-limited setting may influence the time to diagnosis and early performance of surgical treatment. Antenatal ultrasonography may be used as a screening tool to examine the abdominal organ arrangement.[15] However, there is much debate around the necessity of screening patients for abdominal manifestations of heterotaxy and the subsequent decision to operate if any abnormalities are found.[12],[16]-[18] These choices in clinical practice are particularly important because, as illustrated by this case, heterotaxy syndrome can lead to multiple sources of intra-abdominal bowel obstruction, including Ladd's bands, annular pancreas, malrotation, and herniation of the bowel.

Conclusions

Heterotaxy syndrome is a rare condition with a spectrum of presentations and anatomical variations. It is important for the paediatrician and the paediatric surgeon to be mindful of the abdominal manifestations of heterotaxy syndrome as they will guide their screening, surgical, and overall management practices. These practice-based considerations are particularly important given the multiple possible causes of bowel obstruction and the potential for recurrent presentations. Reoperation on these patients can be challenging, especially for surgeons not experienced with congenital bowel malposition.

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