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Polysplenia Syndrome, Heterotaxy with Jejunal Atresia: A Case Report

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Abstract. Heterotaxy is defined as an abnormality where the internal thoracoabdominal organs show abnormal arrangements across the left-right axis of the body. It might affect the stomach alone, *i.e.* dextrogastria. This anomaly can be combination of dextrogastria with functional asplenia (asplenic or hyposplenic) and other visceral heterotaxy with possible malrotation. Heterotaxy is rare condition and its association with intestinal atresia is almost very rarely reported in the literature. Presented is a case of heterotaxy with dextrogastria, multiple small splenules and associated with Type 2 jejunal atresia with brief review of the literature.

Keywords: Polysplenia syndrome, Heterotaxy, Dextrogastria, Situs ambiguous, Jejunal atresia, Intestinal atresia, Asplenia.

Introduction

Heterotaxy is defined as an abnormality where the internal thoracoabdominal organs show abnormal arrangements across the left-right axis of the body. It is a broad term that includes variety of congenital anomalies which could involve almost every organ in the body[1]. The association of this condition with duodenal Artesia is very rare, but reported in 20 patients[2]. However, in the recent literature the association of heterotaxy with jejunal Artesia was reported once by Rasool *et al.*[3]. Abdur-Rahman *et al.* reported a case of asplenia in patient of situs inversus abdominis with reverse rotation of intestine[4].

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Presented is a case report of polysplenia syndrome associated with heterotaxy, where the stomach was found on the right side of the patient (dextrogastria). Additionally, 3 splenules was found along the greater curvature of the stomach, and Type 2 jejunal artesia 10 cm distal to duodenojejunal junction.

Case Report

A preterm baby girl born at 33 weeks of gestation with birth weight of 1.285 kg, as a product of an emergency caesarean section for an un-booked mother. APGAR (Activity, Pulse, Grimace, Appearance, Respiration) score was 9 at one min and 10 at 5 min. On the first day, the baby had greenish naso-gastric tube (NGT) output. The abdominal X-ray showed the NGT is passing clearly to the right side of the upper abdomen with apparently normally-situated cardiac and hepatic shadows. Moreover, there were significantly dilated proximal small bowel loops (Fig. 1).

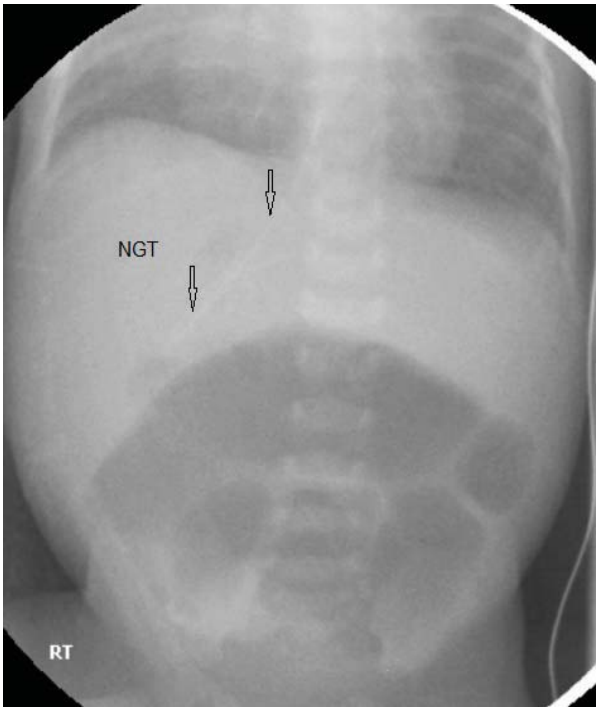

Fig. 1. Abdominal X-ray showed that the NGT is passing to the right side with significantly dilated proximal small bowel loops and normal cardiac and hepatic shadows.

Gastrograffin enema showed microcolon (disused) while the upper gastrointestinal (GI) study confirmed the dextrogastria with proximal small bowel neonatal intestinal obstruction (Fig. 2).

Echocardiogram study showed complex cardiac lesion (complete A-V canal with mild aortic and pulmonary insufficiency and patent ductus arteriosus (PDA) with left to right shunt).

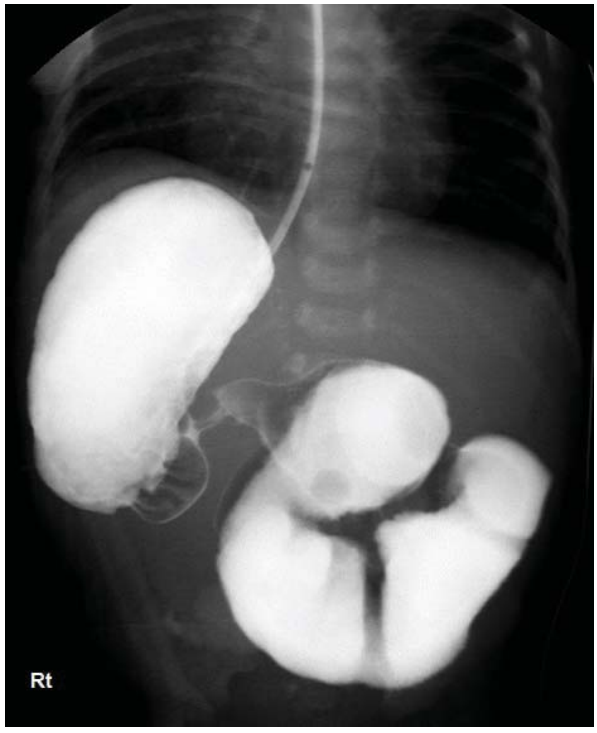

Fig. 2. Gastrograffin upper GI study confirmed the presence of proximal small bowel neonatal intestinal obstruction with dextrogastria.

Pre-operatively, patient was started on intravenous ampicillin, Genamycine and Flagyl. Through an upper right transverse incision, exploratory laparotomy was done at age of 4 days, which revealed dextrogastria (Fig. 3) with 3 splenules (polysplenia) along the greater curvature (Fig. 4), with 5 cm atretic segment of proximal jejunum (Type 2 Artesia). Resection and end to end anastomosis was done with tapering of the hugely dilated proximal loop by using a linear stapler and reinforced with second layer of interrupted 5/0 VICRYL™ (Fig. 5, 6, 7).

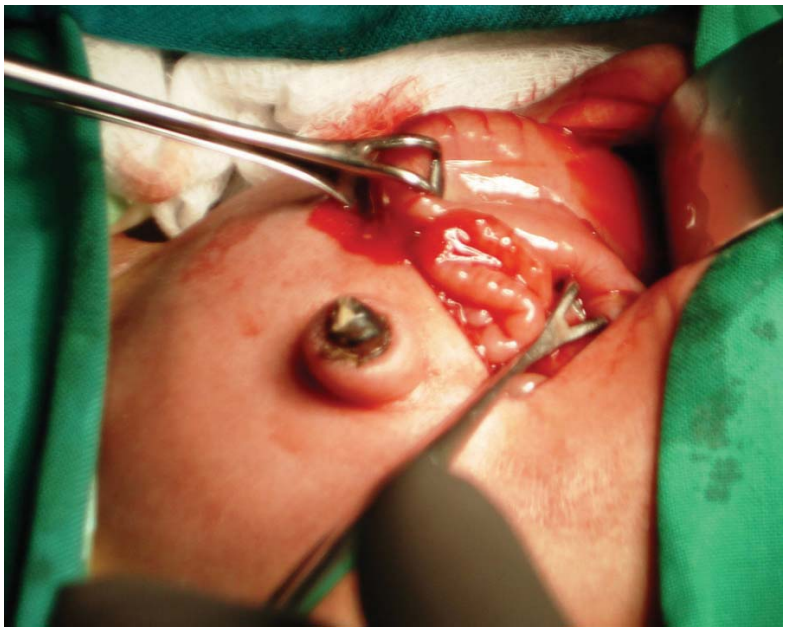

Fig. 3. The stomach located in the RT side abdomen.

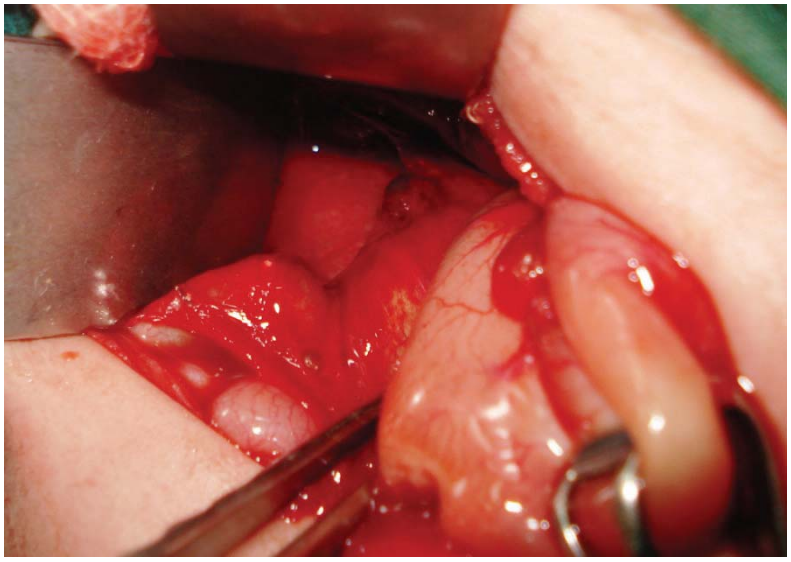

Fig. 4. One of the 3 splenules.

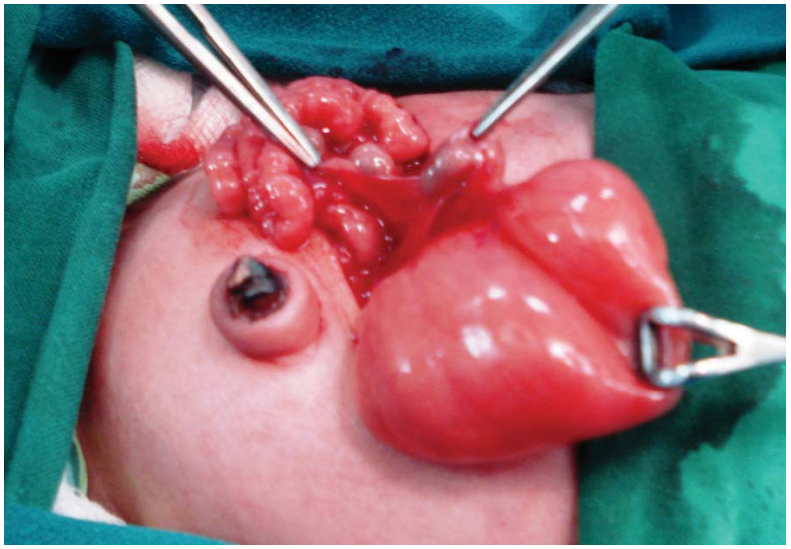

Fig. 5. The atretic jejunal segment 10 cm distal to the DJ junction.

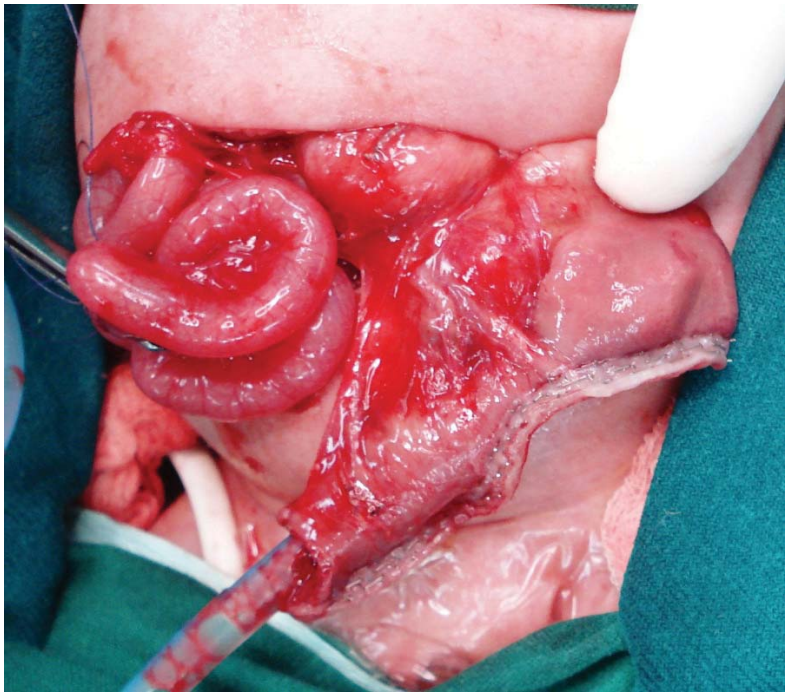

Fig. 6. Tapering the dilated proximal jejunual end.

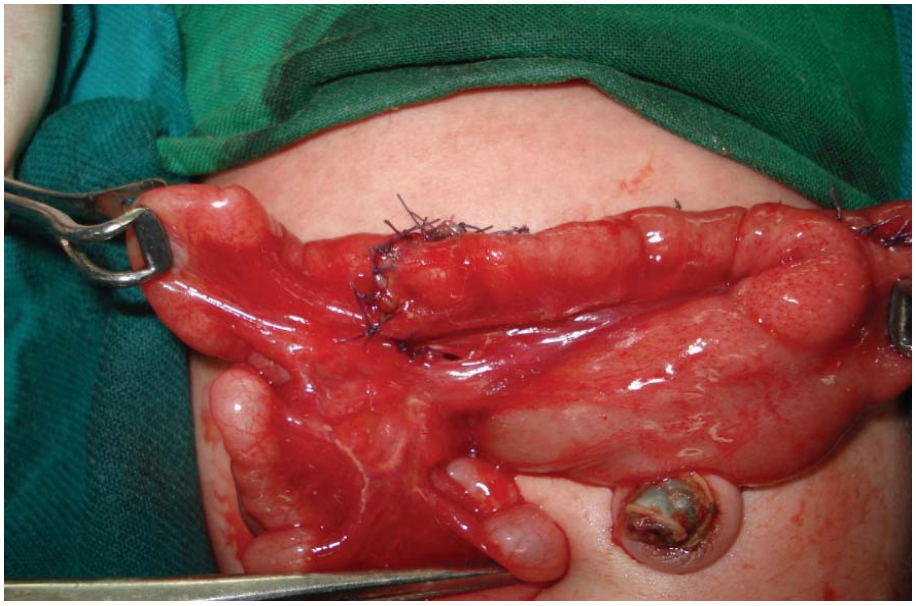

Fig. 7. Completed anastomosis of the jejunum after resection of the atretic segment and tapering the proximal dilated jejunal end.

Post operatively, several trails of gradual increase of oral feeding failed, but the last successful trail was 2 months post-operatively. During that period, gastrograffin follow-through was needed to rule out significant anastomotic stricture, but it showed the free flow of the contrast through the anastomotic area and the rest of the bowel. Patient was surgically discharged at the age of 70 days, weighing 2.5 kg, but continues his care with the NICU team. She suffered from 5 episodes of sepsis, with repeated positive blood and respiratory culture of *Enterobacter cloacae* and *Klebsiella oxytoca* at 2nd and 6th week and coagulase –ve staph at 8 week and coagulase –ve staph and *Pseudomonas aeruginosa* at 12th-16th week, post operatively for which she received the needed treatment by the neonatologist. The patient also suffered from grade one intra-ventricular hemorrhage which was treated conservatively.

At surgical discharge, the patient was well; tolerating full feeds orally on Captopril, amoxicillin and two diuretics in the form of spironolactone and furosemide, plus he is being prepared to undergo cardiac corrective surgery.

Discussion

Heterotaxy is defined as an abnormality where the internal thoracoabdominal organs demonstrate abnormal arrangement across left-right axis of the body[1].

On the other side, several terms have been used to describe similar other anomalies, like situs inversus which describes a condition where organs are mirrored either totally or partially[2]. Situs ambiguous is defined as an abnormality in which there are components of situs solitus and situs inversus in the same person; thus, heterotaxy and situs ambiguous are opposite faces of the same coin[1].

Jejunoileal atresia is an uncommon disease that affects 1 in 330 to 1 in 1500 live births[3]. The association of this anomaly with heterotaxy is very rare. Literature review up to date, revealed few case reports of jejunal atresia with situs inversus[4-7].

Left-sided liver, asplenia or right sided spleen and malrotation are the most common abdominal anomalies associated with heterotaxy. Less commonly known are the gastroschisis, biliary atresia and diaphragmatic hernia. Cardiac anomalies are the commonest extra-abdominal association and usually their symptoms are the first to be detected[8].

Asplenic individuals are known to be at an elevated risk for infection. Similarly, the patient of heterotaxy with asplenia or minimal insufficient splenic tissue mass (hyposplenia), usually at high risk of recurrent sepsis[9] as seen in our patients.

Brown *et al.* (2009), on his literature review found 20 reported cases of heterotaxy with duodenal atresia included all types of duodenal atresia[8].

In 2007, Abdur-Rahman *et al*.[5] reported a case of dextrogastria associated with jejunal atresia. However, they described the presence of the liver on the left side which makes the case more towards being a case of situs inversus rather than heterotaxi[5]. But our case had dextrogastria, polysplenia (situated on the right side of the patient) with a normally situated liver (Fig. 1 and 2) that's mentions our case was of polysplenia syndrome with heterotaxy. The association of such condition with jejunal atresia is reported once in the recent literature[4]. The associated atresia in our case led to early detection of such abdominal organ derangement,

while in hetrotaxy without atresia patient presents with recurrent sepsis or symptoms of associated complex cardiac anomaly. Our priorities in our case were to restore the intestinal continuity reach full oral feeding and prevent the sepsis, then to involve the cardiac surgeon to manage the cardiac abnormality.

Conclusion

This report presented a unique case of heterotaxia with jejunal atresia which has not been reported before. The associated atresia led to detection of the heterotaxia, thus made the treatment priorities were the surgery of the atresia to accomplish full oral feeding with the prevention of sepsis.

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تقرير حالة متلازمة الطحيليات المتعددة مع التفاف الأحشاء وانسداد خلقي للأمعاء

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المستخلص. هيتيروتاكسي تعرف بالتفاف عكسي للأحشاء الداخلية على المحور الطولي للجسم من الجهة اليسرى إلى الجهة اليمنى وهذا الالتفاف قد يشمل المعدة فقط ويسمى بالتفاف أو انحراف المعدة يميناً وقد يتصاحب مع ذلك ضعف في وظيفة الطحال لعدم تكونه أو لقلة أنسجته وأيضاً قد يتصاحب مع ذلك التفاف الأعضاء الأخرى الداخلية أو التواء الأمعاء. ويعتبر الهيتيروتاكسي من العيوب الخلقية النادرة ومصاحبتها للانسداد الخلقي الكامل للأمعاء شديد الندرة. وهنا نقدم دراسة حالة من الهيتيروتاكسي تشتمل على التفاف المعدة للجهة اليمنى ونقص في أنسجة الطحال (طحيليات صغرى متعددة) ويصاحبها أيضا انسداد خلقي للأمعاء من النوع الثاني.