Case 13602

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A rare type of Jejunal atresia (Apple peel small bowel)

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Section: Paediatric radiology

Area of Interest: Paediatric

Procedure: Barium enema

Procedure: Diagnostic procedure

Procedure: Diagnostic procedure

Procedure: Barium meal

Imaging Technique: Fluoroscopy

Imaging Technique: Conventional radiography
Special Focus: Pathology Case Type: Clinical Cases
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Patient: 6 days, female

Clinical History:

A female neonate was referred for an upper GI study. She presented with bilious vomiting and abdominal distension. **Imaging Findings:**

Plain film of the abdomen showed dilated proximal bowel loops without evidence of bowel gas in the lower abdomen (Fig. 1). An upper GI study through a naso-gastric tube demonstrated a dilated stomach, duodenum, and proximal jejunum, with an abrupt cut off at the proximal part of jejunum (Fig. 2). In order to rule out distal atresia a lower GI study was performed which showed a small caliber colon containing central filling defects representing plugs of meconium (Fig. 3). Soon after contrast injection, meconium plugs were evacuated and a microcolon was demonstrateed positioned mostly in the left side of abdomen (Fig. 4). Surgery confirmed atresia in the proximal jejunum with at least three more distal atretic areas associated with a short mesentery and spiral shaped distal bowel loops (apple peel appearance) (Fig. 5).

Discussion:

Jejunal atresia is a congenital anomaly which is characterized by obstruction of the jejunum. The closure can be anywhere within the lumen of jejunum from the ligamentum of Treitz to the jejuno-ileal junction. The atresia can be solitary or multiple. This anomaly is caused by ischemic injury during embryonic development [1]. Children with proximal jejunal atresia usually present with abdominal distension and bilious vomiting starting within hours of birth. In cases of distal jejunal obstruction, vomiting may take longer to appear [2]. Abdominal plain radiography and GI fluoroscopic studies are usually performed to diagnosis the disease.

Abdominal radiography shows proximal dilated bowel loops with the usual appearance of a three-bubble sign, which can be distinguished from the double bubble sign of duodenal atresia. In ileal atresia, dilated bowel loops are usually multiple. All types demonstrate an absence of distal bowel loop gas [1]. Although plain radiography can demonstrate proximal dilated bowel loops, it cannot exactly locate the abnormality, therefore an upper GI study can be performed. Isolated Jejunal atresia usually demonstrates a normal sized colon because the distal bowel loops produce sufficient secretions to make a normal caliber colon. When a more distal obstruction or multiple atretic

segments are present microcolon will be produced [3-5]. Barium enema is used to exclude microcolon and differentiate between an upper and lower bowel atresia as well as Hirschsprung's disease and meconium plug syndrome [2].

The present study shows a rare type of jejunal atresia, called Apple Peel small bowel (APSB). APSB is a rare familial anomaly which is believed to be an autosomal recessive genetic disorder. It usually accounts for about 10-15% of small bowel atresia [6, 7]. APSB is characterized by proximal jejunal atresia, and absence of distal mesentery and the distal superior mesenteric artery [3-4]. The short distal bowel twists around its vessels and produces an apple peel appearance [4].

For treatment of jejunal atresia different surgical procedures exist. For instance, serial transverse enteroplasty, tapering enteroplasty, and stenting of multiple bowel segments [2]. In the present case the atretic segments were resected and when the patiency of the entire length of bowel was recognized, side to side anastomoses were performed. Generally the patients have a good prognosis after surgery; however the possibility of recurrent obstruction is high at the anastomotic site during the first year of age [2, 7].

Differential Diagnosis List: Jejunal atresia, Apple peel small bowel type, Malrotation with midgut volvulus, Total colon Hirschsprung disease, Meconium ileus

Final Diagnosis: Jejunal atresia, Apple peel small bowel type

atresia. Journal of pediatric surgery 32(9): 1307-1309.

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Description: Dilated air filled stomach and proximal bowel loops with no evidence of air in the distal loops, nasogastric tube is seen inside the stomach **Origin:** Radiology Department of French Medical Institute, Kabul, Afghanistan



Description: Upper GI study demonstrated contrast material within the stomach and dilated duodenum and proximal jejunum with an abrupt cut off at the level of the proximal jejunum (arrow) **Origin:** Radiology Department of French Medical Institute (FMIC), Kabul, Afghanistan

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Description: Lower GI study shows small caliber colons containing filling defects (meconium plugs) **Origin:** Radiology department of French Medical Institute for Children (FMIC), Kabul, Afghanistan

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Description: Malrotated micro colon post meconium plugs evacuation **Origin:** Radiology department of French Medical Institute for Children (FMIC), Kabul, Afghanistan

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Description: Atresia at the level of proximal jejunum (large arrow) with proximal dilatation (small arrow) associated with twisted distal bowel loops (apple peel appearance) **Origin:** Radiology department of French Medical Institute for Children (FMIC), Kabul, Afghanistan