

Apple-peel Atresia with a Twist: A Novel Case Report

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Abstract

Jejunoileal atresia is one of the more common causes of neonatal intestinal obstruction. Its incidence is up to 1 in 3000–5000 live births. The apple-peel deformity, Type IIIb according to Grosfeld's classification, forms about 5% of all jejunoileal atresia. It consists of a proximal jejunal bowel which ends blindly into a dilated segment; and a distal jejunoileum which exhibits a helical distribution around a central blood supply which usually arises from the ileocolic artery. We are presenting a novel case of jejunoileal atresia where the proximal bowel ends in an apple-peel formation, and the distal small bowel is supplied by a branch of the ileocolic artery. The baby was a full term, good weight, and otherwise well female baby with no major associated malformations.

Key words: Apple-peel atresia, Jejunoileal atresia, Neonatal intestinal obstruction

INTRODUCTION

Jejunoileal atresia is one of the more common causes of neonatal intestinal obstruction which requires early, if not immediate, surgical management, and barring which it is incompatible with life. The incidence is up to 1 in 3000–5000 live births with a demographic variation.^[1,2] It is antenatally diagnosed by fetal sonography which may show polyhydramnios, dilated small bowel loops and in some, fetal ascites if present. If antenatal bowel perforation occurs, it can be picked up, later in gestation, as calcification in the peritoneal cavity. Jejunoileal atresia has been classified by Grosfeld as follows, Type I has the presence of a mucosal web, Type II has a fibrous cord extending from the dilated proximal to the distal segment of gut, Type IIIa has a V-shaped mesenteric gap between the proximal dilated and distal micro gut, Type IIIb is the typical “apple-peel” deformity, and Type IV is the presence of multiple atresias, referred to as a “string of sausages.”^[1,2]

The apple-peel deformity, Type IIIb, forms about 5% of all jejunoileal atresia,^[3,4] consists of a proximal small bowel which ends in a typically blind and dilated jejunal or ileal segment which is supplied by the superior mesenteric artery (SMA) and the distal unused jejunoileum which is usually supplied by a branch from the ileocolic artery or sometimes from a branch of the right colic artery.^[1,2] The name “apple-peel” comes from the fact that the distal small bowel is rotating around its arterial supply as the peel of an apple and the mesentery is not fixed to the posterior abdominal wall. The arterial supply enters the spiral of the small bowel from the distal end.^[1,2] This deformity has the highest mortality out of all types of jejunoileal atresia followed by multiple atresia Type IV.^[5] We are reporting a novel case of jejunoileal atresia with apple-peel deformity, where the apple-peel configuration is seen in the distal end of the proximal bowel.

CASE REPORT

A full-term female baby weighing 3 kg with an antenatal diagnosis of polyhydramnios and dilated bowel loops had an abdominal X-ray [Figure 1a] suggestive of jejunoileal atresia. A gastrografin enema was given and the dye study observed under fluoroscope. Micro-colon was observed and the caecum was lying in subhepatic position [Figure 1b] with few loops of unused distal ileum seen bunched up around the caecum. The baby was kept nil by mouth, given

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intravenous fluids, nasogastric tube was placed, aspirated and kept on the drain, antibiotics started, Vitamin K injection given and prepared for surgery.

On laparotomy dusky loops of small bowel [Figure 2a] were seen at the start. When an attempt was made to deliver these loops, they were seen to be twisted [Figure 2b] and an effort to untwist the bowel led us to the large dilated blind end which was densely adherent to the under-surface of the liver and the superior surface of the bladder. This dusky small bowel was twisted around a leash of mesentery which was free and artery forceps could be passed under it [Figure 2a] freely. While attempting to untwist this loop, it was seen that the vessels in this mesentery were dusky and blackish in color. We decided to resect this gut as it did not improve on applying the warm sponge to the bowel and giving 100% oxygen in the respiratory circuit.

Proximally the bowel was resected about 1–2 cm away from the unhealthy margin but distally the grossly dilated blind end, seen as the large circular loop of gas-filled bowel in Figure 1a, was densely adherent to the liver and gall bladder superiorly and the urinary bladder inferiorly, indicating an antenatal perforation with fibrous adhesion. All attempts to separate, this adhesion was causing damage to the liver and the urinary bladder; hence, the parts which were adherent were left behind and the mucosa was peeled off. The resected specimen when held up showed the apple-peel configuration [Figure 3]. The proximal small bowel left behind was measured and was approximately 48–50 cm in length starting from the duodenojejunal junction. The distal small bowel was now seen more clearly as it had been lying jumbled up around the caecum, posterior to the dilated end of the proximal bowel [Figure 1b]. After injecting it with saline and confirming the distal patency, it was seen that the proximal 5–6 cm was a fibrous cord, which was excised. The remaining distal ileum was approximately 30 cm long. End to back anastomosis was done, mesenteric defect approximated.

Postoperatively the baby had metabolic acidosis, was kept ventilated, and needed bicarbonate correction. She showed signs of sepsis with fall in platelets on the 3rd post-operative day. Aggressive management was continued. The baby passed a small amount of meconium per rectum. On the 5th post-operative day, there was a bilious leak in the surgical site and we decided to re-explore expecting a suture line leak which was the case. The bowel appeared pink and viable and except for a small leak, the rest of the repair was holding well. The distal gut had improved in caliber and contained meconium. We repaired the leak, inserted a central line, and kept the baby ventilated. On the 2nd post-operative day, the baby came off the ventilator, but at the same time, the leak reoccurred. We kept the baby nil by mouth, on total parenteral nutrition (TPN) and started a very low dose of octreotide,

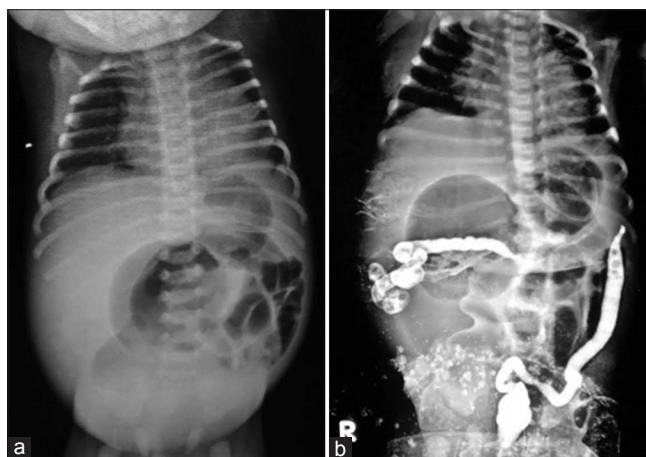


Figure 1: (a) Infantogram showing small bowel loops with dilated blind end and abrupt cut-off of bowel gas. The large bowel is not seen, gas in the stomach and a few loops of the small bowel are suggestive of jejunoileal atresia. (b) Gastrograffin enema showing unused “micro-colon” with caecum in sub-hepatic position. Dye is seen in loops of unused ileum which is bunched up in sub-hepatic region (there are artifacts in the lower region of the fluoroscope screen picture)

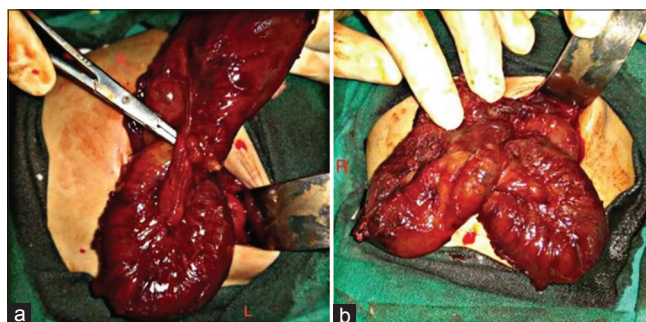


Figure 2: Dilated loops of small bowel proximal to the atresia, seen to loop around the mesenteric vessel forming an apple-peel configuration. The bowel is dusky with black patches all over. (a) An artery forceps eliciting a free mesentery around which the small bowel is rotating. (b) shows the apple-peel configuration of the proximal dilated dusky small bowel. Gangrenous patches are visible in the dusky loops

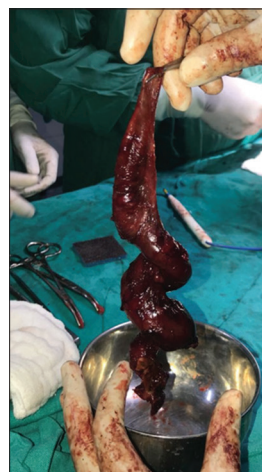


Figure 3: Apple-peel configuration of the resected proximal gangrenous small bowel

intravenously. Within 2 weeks the entero-cutaneous fistula had reduced in size and was draining a maximum of about 5–8 ml in 24 h, and the baby started passing small quantities of meconium about twice a day through anus. Postoperatively the weight had come down to 2.5 kg, and following TPN it had come up to 2.75 kg. We started oral feeds and gradually increased the quantity while reducing the TPN. The baby was now improving and was 30 days old. As we were gradually going up on feeds and had finally stopped the TPN, the parents, who were actively involved at this stage, insisted on taking the baby home, confident of being able to manage.

Within 6 days, the baby was brought back, severely dehydrated, in septicemia, with bradycardia, and in gasping condition. Although we resuscitated her and put her on life support measures, she succumbed to sepsis after 3 days.

DISCUSSION

We are reporting a case of jejunoileal atresia with an apple-peel deformity at the end of the proximal gut. This is completely different from the Type IIIb jejunoileal atresia which has been described by Grosfeld.^[1] The blood supply to this apple-peel segment was a distal jejunoileal branch of the SMA which ran through the center of the apple-peel configuration, entering it from the proximal end. The distal segment of ileum was supplied by arterial branches which appeared to be arising from the ileocolic branch of the SMA. The ascending colon was deficient and the caecum was in a sub-hepatic position. Alnosair *et al.*^[4] have reported five cases, including their own, of a variant of apple-peel atresia where there is duodenal atresia, absence of 3rd and 4th part of duodenum, absence of SMA and the entire jejunoileum is in the form of an apple-peel deformity with a marginal arterial supply arising from the inferior mesenteric artery. To the best of our knowledge, we have not come across an anomaly similar to the one we are reporting, in the available literature.

Low birth weight, prematurity, and associated anomalies, mainly cardiac, are known to be the cause of death in the post-operative period.^[1,2,4,5] None of these were present in our patient. The reason for an anastomotic leak, which is known to be a common post-operative complication,^[1,2,6] could not be explained as the distal gut had moved a few times and the anastomosis itself was pink and patent showing no signs whatsoever of any vascular compromise on re-exploration. We can safely presume a functional obstruction of the distal ileal segment.^[1,2,6,7]

The mortality and morbidity are higher with antenatal perforation of the bowel and vascular compromise. In

our patient, on exploration there were definite signs of an antenatal perforation and the blind end of the proximal bowel was densely adherent to the under-surface of the liver and the superior surface of the urinary bladder. There was no indication of an antenatal perforation on abdominal X-ray nor was there any indication of the gangrenous gut in the general condition or blood biochemistry of the baby. She appeared good weight, full term, active and well maintained, not giving us any warning of the gangrenous gut in the abdomen. Although we did not see it coming the baby was taken up for surgery at 24 h of age, so the delay was not the reason. We do feel that had the baby stayed in the hospital, we would have had a better chance to gradually shift her from parenteral to enteral feeds and attempted an early closure of the entero-cutaneous fistula. The major problems that we faced were the inability to maintain TPN for long due to the high cost and a high rate of hospital-acquired infections.

CONCLUSION

We have reported a novel case of jejunoileal, apple-peel atresia and described the anatomy as we found it. It is a matter of further speculation and research as to what could be the developmental incidents which have led to this, as far as we know as yet, unreported form of intestinal atresia.

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