A Case of Duodenal Atresia with Apple Peel Appearance: Challenging the Current Embryology

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ABSTRACT
Classically, embryology of duodenal atresia has been linked to defect in recanalization process, while apple-peal atresia of small bowel has been considered as due to vascular accident during embryonic life. We present a 33 week preterm neonate with duodenal atresia with the apple-peel appearance of proximal jejunum for which resection of the jejunum with apple-peel configuration, plication of the duodenum, and duodeno-jejunal anastomosis was done. Thus, this rare case of ours questions the embryology of duodenal atresia with the apple-peel appearance suggesting it to be due to a vascular disruption phenomenon during embryonic life.

Key words:
Apple-peel atresia, duodenal atresia, embryology

INTRODUCTION
Duodenal atresia is one of the most common sites of neonatal intestinal obstruction. The incidence of duodenal atresia has been estimated at 1 in 6000-1 in 10,000 live births.[1] Duodenal atresia is often discovered antenatally on routine sonogram during pregnancy. Maternal polyhydramnios and classic “double-bubble” sign on fetal ultrasonography suggests the diagnosis.[2] Duodenal atresia is thought to result from problems during embryo's development, in which the duodenum does not normally change from a solid to a tube-like structure.[3,4] We present here a case, which queries the embryological basis of duodenal atresia possibly due to vascular accident.

CASE REPORT
A 2-day-old preterm (33 weeks), weighing 1.3 kg neonate was referred to us from neonatal intensive care unit as it was prenataly diagnosed as a case of duodenal obstruction on ultrasonographic findings of polyhydraminos with the double-bubble appearance. On examination, the baby was alert, active, and had no obvious external morphological congenital anomalies. There was upper abdominal fullness and nasogastric tube drained bilious aspirate. External genitalia and anal opening was normal. Further evaluation by X-ray flat plate abdomen showed the classic “double-bubble appearance” of duodenal atresia with total absence of distal bowel gas [Figure 1]. Echocardiography didn't reveal any congenital heart anomaly. Ophthalmological examination was done to rule out strumme syndrome. Ophthalmological examination was normal. Based on this information, baby was taken-up for surgery. On exploratory laparotomy, it was revealed that duodenum was atretic and there was marked dilation of proximal duodenum. The upper 15 cm of jejunum had apple-peel configuration [Figure 2]. This part of bowel was spiraling around a retrograde vascular arcade, rest of the small and large bowel was normal. As only 15 cm of jejunum was having the apple-peel configuration with precarious blood-supply, we decided to resect this part of the bowel. End to oblique duodeno-jejunal anastomosis was done along with the plication of grossly dilated proximal duodenum [Figure 3]. Postoperatively baby was kept on ventilator support for 1 day. Gradually increasing nasogastric feed was started from postoperative day 6. The baby was on full breast feed by postoperative day 10 and discharged from hospital.

DISCUSSION
Intestinal atresia most frequently affects the duodenum, followed by the jejunum. This defect is usually diagnosed prenatally via ultrasound or shortly after delivery by the presence of vomiting (mostly bilious) after each
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feed. Approximately 50% of infants with duodenal atresia/stenosis have another anomaly, including cardiac, genitourinary, or anorectal defects, and annular pancreas. Although no specific genetic abnormality is known to cause duodenal atresia, the number of reports of the anomaly occurring among siblings and among several generations of a family, as well as its frequent association with trisomy 21, suggests one may be present. A severe form of intestinal atresia/stenosis is described as “apple-peel deformity.” This name is derived from the appearance of the intestine as it spirals around the blood-supply and resembles an apple-peel. The mortality of apple-peel atresia remains high though it has come down greatly in most of the reported series in last two decades.

The embryological etiology of the duodenal atresia differs from that of small bowel atresia. In the 4th week of development the endoderm gives rise to the gut tube. Rapid proliferation of the gut epithelium in the 6th week of development results in obliteration of the intestinal lumen. Recanalization of the intestine occurs over the next several weeks of development.

An error in this recanalization process is considered to be the embryological basis of duodenal atresia and stenosis. This differs from jejuno-ileal atresia, which is assumed to result from vascular disruption phenomenon during the later phases of gestation. Apple-peel atresia is a rare variant of intestinal atresia in which the distal bowel is precariously supplied by a retrograde vascular arcade from the ileocolic, right colic or inferior mesenteric arteries. Our patient with duodenal atresia had apple-peel configuration of upper 15 cm of jejunum. This association of duodenal atresia with apple-peel small bowel atresia is extremely rare. Our patient did not have other associated congenital anomalies commonly seen with duodenal atresia. The presence of apple-peel atresia of upper jejunum being precariously supplied by retrograde vascular arcade and absent associated congenital anomalies suggest that duodenal atresia in our case resulted from vascular disruption phenomenon in late gestation.

Some literature in the past reportedly favored vascular accidents over recanalization defects in the cases of duodenal atresia. Likewise, our case also with findings described channelizes our thoughts to an embryological variant duodenal atresia.

CONCLUSION

Duodenal atresia with the apple-peel appearance is a rare anomaly with limited cases and literature in the past.

Nevertheless, though the current understanding favors recanalization theory, but rarely vascular disruption phenomenon can cause duodenal atresia with the apple-peel appearance which may indicate insult occurring in late gestation.
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