Duodenal atresia with 'apple-peel configuration' of the ileum and absent superior mesenteric artery: A rare presentation

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ABSTRACT

According to the current understanding, duodenal atresia is considered to be a primary malformation resulting from the errors in recanalisation in early gestation. We report a rare case of duodenal atresia with apple-peel configuration of remaining small bowel with absent superior mesenteric artery in a preterm child, which indicates that in rare circumstances vascular accidents may be the underlying cause for duodenal atresia, and jejuno-ileal atresia; only second such case to be reported in the English literature.

Key words: Apple-peel configuration of the ileum, duodenal atresia, vascular accident

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INTRODUCTION

The duodenum is the most common site of neonatal intestinal obstruction, accounting for nearly half of all cases.[1] Incidence of duodenal atresia has been estimated at 1 in 6,000 to 1 in 10,000 births.[2] It is considered to result from errors in recanalisation of embryonic foregut in early gestation.[3] This aetiology differs from that of intestinal atresias in other parts of the bowel, which are assumed to result from vascular accidents during later phases of gestation.[4] Due to the significantly different aetiology, the association of duodenal atresia with apple-peel, small bowel atresia is very rare. We present one such rare association, which raises questions on the aetiology of duodenal atresia. To the best of our knowledge, ours is the second such case to be reported in the English literature.[5]

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CASE REPORT

An 8-day-old boy, 1.2 kg preterm (34 weeks) neonate, was referred from neonatal intensive care unit (NICU) to us with complaints of persistent high bilious aspirates. The child had passed stools in scanty quantities. Antenatal ultrasonography (USG) showed maternal polyhydromnios. On examination, the baby was alert, active, and had no other external congenital anomalies. Abdominal examination revealed soft, nontender abdomen, with fullness of the epigasrium [Figure 1]. There was normally situated anal opening. Plain radiograph of the abdomen showed the classic "double-bubble" sign [Figure 2] of duodenal atresia with total absence of distal bowel gas. After optimisation of general condition, the patient had exploratory laparotomy, which revealed loss of third and fourth part of the duodenum with an apple-peel configuration of the remaining small bowel with absence of superior mesenteric artery [Figure 3a and b]. The precarious blood supply of apple- peel small bowel depended entirely on the marginal artery that was fed retrogradely through the left colic artery. Duodenum was anastomosed end-to-end to the hypoplastic apple-peel jejunum. Postoperatively, patient was kept on ventilator support. However, he succumbed on second day due to very low birth weight with prematurity.

DISCUSSION

The cause of duodenal and small bowel atresias may vary according to the site of the lesion. Embryologically, the lumen of the gut in a five-week-old embryo is well-defined throughout its length. After the formation of a primary intestinal canal, the developing embryonic foregut goes through a complete solid core stage that subsequently vacuolises.[5] Duodenal and upper jejunal atresias are believed to be related to failure of recanalisation of the solid stage of developing embryonic foregut, as proposed by Tandler in 1900.[6] Atresia of rest of the small bowel is considered to result from interruption of vascular supply to the atretic bowel segment.[7]



Figure 1: Clinical photograph showing upper abdominal fullness



Figure 2: Plain radiograph of the abdomen showing the classic "double-bubble" sign of duodenal atresia

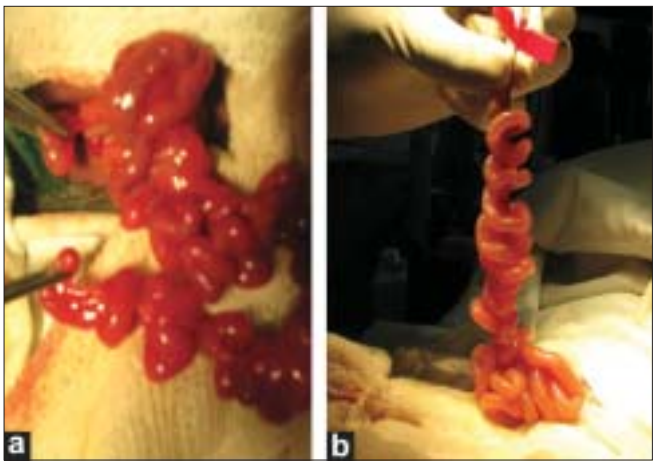


Figure 3: (a) Intraoperative photograph showing the atretic duodenum (b) Intraoperative photograph showing the apple-peel small bowel

In 1961, Santulli and Blanc used the term apple-peel atresia for the first time.[8] Apple-peel atresia consists of a high jejunal atresia with a discontinuity of the small bowel and a wide gap in the mesentery. The distal segment of ileum is shortened and assumes a helical configuration around a retrograde perfusing vessel, which compensates for the partially absent superior mesenteric artery. An intrauterine vascular accident in late gestation has been accepted as the cause of apple-peel atresia and presents with a spectrum of occlusions of one or more branches of the superior mesenteric artery.[9]

The present patient had an atretic third and fourth part of duodenum and proximal jejunum with typical apple-peel configuration of the ileum with total absence of superior mesenteric artery (SMA). The totally missing SMA probably caused duodeno-jejunal atresia due to interruption of pancreatico-duodenal arcade, in addition to apple-peel atresia. Although, an unrelated occurrence of duodenal atresia and apple-peel atresia in this child cannot be ruled out; it seems to be very unlikely.

The current concept favours the recanalisation theory to explain the duodenal atresia. Our case suggests, however, that in rare circumstances vascular accidents may be the underlying cause of duodenal atresia.

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