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Case Report
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A Very Rare Case of Intestinal Atresia, the Apple Peel Syndrome

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Abstract

Small bowel atresia is among the rarest congenital gastro-intestinal anomalies. Apple-Peel syndrome is a very uncommon pattern of small intestine atresia: <5%. The present paper records the case of a neonate, who presented an Apple peel syndrome. The clinical presentation featured an occlusive syndrome and was explored with abdominal X-ray and ultrasound. The preoperative exploration found a small intestine atresia with a snail-like aspect. We performed an end-to-end jejunal single-stage anastomosis. The aim of our work is to make a focus on this uncommon neonatal condition as well as the benefits and risks of its treatment by a single-stage approach without undergoing an enterostomy.

Keywords: Atresia; Apple Peel syndrome; Abdominal X-ray

Introduction

Described for the first time in 1683 [1] jejuno-ileal atresia is among the most uncommon congenital anomalies of the gastrointestinal tract (3/10000 alive birth) [2,3]. Apple peel syndrome is a very rare pattern of small intestine atresia (<5) [4]. Also known as "pigtail-like syndrome", it constitutes a rare entity with a high interruption of the superior mesenteric artery and a precarious revascularization of the distal small bowel portion by the ileal recurrent artery [4]. The aim of this study is to make a focus on this uncommon neonatal condition as well as the benefits and risks of its treatment by a single-stage approach without undergoing an enterostomy.

Case Presentation

It was about a 4-day-old male newborn who was admitted for a neonatal bowel obstruction. He was born through normal delivery, after 36 weeks gestational period, followed with two prenatal cares and two unremarkable ultrasonography. The birth weight was at 2800 g with an Apgar score at 9/10. The questioning found a failure to pass meconium and the occurrence of bilious vomiting 3 days after birth. Clinical examination revealed a flat abdomen and a negative probe test. Abdominal X-Ray (Figure 1) showed air-fluid levels with central dilated loops of the small intestine signing a proximal obstruction. The rest of the abdomen was gasless. Abdominal ultrasound showed jejunal enlargement with caliber disparity and jejunal interruption. Surgical investigations by transverse subumbilical mini-laparotomy approach pointed out a jejunal atresia, approximately distant to 30 cm to the angle of Treitz, with distal small intestine twisted on its axis giving a pigtail-like aspect (Figures 2 and 3), without necrosis. About 50 cm was resected to the jejunum. An end-to-end jejunal anastomosis was performed with proximal jejunal loop modeling associated to appendectomy. The left portion of the intestinal loop was estimated at 190 cm. The tissue sample was sent to anatomopathology. The neonate was referred to a neonatal resuscitation unit for post-operative suites. During post-operative suites, malformative assessment was performed and unremarkable. It included: cardiac Doppler ultrasound, transfontanellar ultrasound and complete spine X-Ray.

Discussion

Apple peel syndrome is a rare form of small intestine atresia due to superior mesenteric artery obstruction, giving the aspect of proximal atresia with a pigtail-like form of the twisted distal segment around its vascular supply [5]. The atresia seats close to the angle of Treitz, with an approximate distance of 30 cm in our case. The downstream intestine was foreshortened and supplied by a branch of the ileo-coecoappendiculo-colic artery around which it spirals. There was no mesenteric supply

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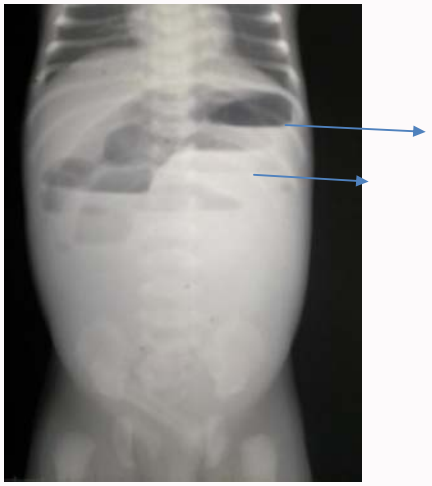

Figure 1: Abdominal X-ray: central air-fluid levels (small intestine), blue arrow.

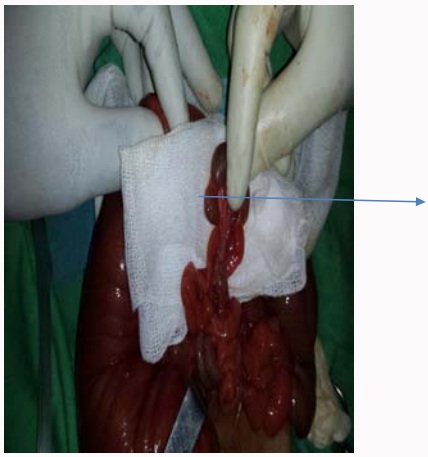

Figure 2: Per-operative aspect showing a pigtail-like aspect of small intestine atresia (blue arrow).

[6]. Apple-peel syndrome accounts for 5% of intestinal atresias [4].

The prevalence of Jejuno-ileal atresias is rated at 1 case per 1500 to 2000 births [4]. Jejuno-ileal atresias are anatomically classified in 5 types according to Martin and Zerella [7] (Figure 4):

- Type 1 : complete membranous obstruction of the lumen with no intestinal defect
- Type 2 : intestinal discontinuity connected with a fibrous band
- Type 3a : disconnected mesenteric interruption
- Type 3b: jejunal atresia with a lack of superior mesenteric artery distal, the distal portion is wrapped around a recurrent vascular arcarde giving the appearance of apple peel with a shortened loop.
- Type 4: multiple atretic segments that resemble a string of sausages.

Sex prevalence is proportional, but most often they are associated to down syndrome in cases [9]. In our case, malformative assessment was unremarkable. Antennal diagnosis is possible with a high ultrasound quality allowing to see the dilatation of the proximal bowel

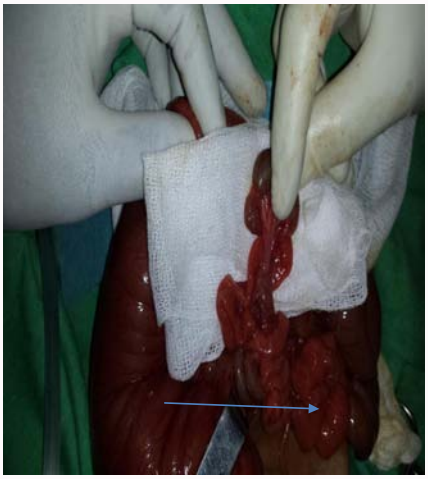

Figure 3: Jejunal enlargement downstream the atresia (blue arrow).

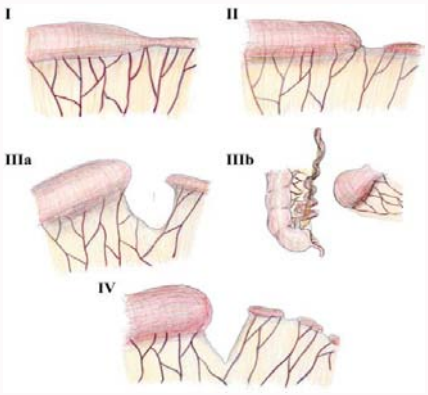

Figure 4 : Martin and Zerella classification [8].

portion indicative of small intestine atresia [2,10]. Unfortunately, in our patient, antenatal ultrasounds didn't indicate small intestine atresia. If antenatal diagnosis is not made, the diagnosis will be suspected only after birth with a neonatal obstruction condition [4]. In our case, the diagnostic was tardinessly made after birth with the occurrence of an obstructive syndrome.

Once the diagnosis is confirmed and the neonate well resuscitated, surgical approach will be able to be performed. The first step consists of the exploration of the abdominal cavity to the confirm the diagnosis; verify the absence of other atresias or gastrointestinal anomalies (intestinal malrotation and duplication). Whatever the strategy could be, the main aim will be to maintain the greatest length possible to prevent short bowel syndrome [11]. Mostly, a resection and primary anastomosis of the atretic portion will be performed. This anastomosis is sometimes difficult because of a disparity of diameter between the two segments leading to some techniques range from modeling the upstream plicature to the realization of a split line downstream [11].

For our patient, surgical explorations didn't find intestinal malrotation, neither duplication nor associated atresia. In relation of a normal vascularization without necrosis and a good visualization of the loops after performing jejunal resection, we chose a single-stage anastomosis after modeling the jejunal proximal portion. In developing countries especially in Africa, the diagnosis is often lately made after birth with the presence of occlusive syndrome. The

prognosis of these neonates is worsened by the diagnostic tardiness and the lack of suitable neonatal resuscitation unit, leading to a high mortality rate. Recently, the prognosis of this condition has become good by a well managed complete surgery with single-stage approach [6]. Mortality remains high, mostly related to short bowel syndrome or hydro-electrolytic loss in case of stormy. In Festen et al. [12] recent series, mortality accounted for 20% in 15 patients presented Apple peel syndrome. Post-operative suites were unremarkable in our case. An enough length of the loop and the single-stage approach without stormy allowed to prevent short bowel syndrome which represents a major mortality factor.

Conclusion

Apple-peel syndrome is a rare condition of small intestine atresia. The diagnosis is often made in perioperative period. In some cases, the treatment of Apple peel syndrome can be performed with single-stage surgery when underlying vessels allow it. This helps preventing to undergo a second stage.

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