

Triple Atresia: A Rarest Variety - Case Report.

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Abstract: *Intestinal Atresia is a common cause of neonatal intestinal obstruction. Triple atresias involving jejunum, ileum and colon is rare entity. We had a case of Triple atresia with imperforate anus in a baby with Down's syndrome. This rarity and mode of presentation make it an interesting case to be reported to know such entity and to deal with them in possible best way.*

Keywords: *Anus, Imperforate; Cesarean Section; Colon; Down Syndrome; Female; Gestational Age; Intestinal Atresia; Jejunum; Laparotomy*

I. Introduction

An atresia is defined as 'a congenital absence of a body opening or body passage [1]. Intestinal Atresia is a common cause of neonatal intestinal obstruction[2]. Lack of revacuolization is probable cause for most cases of duodenal atresia. Jejunum-ileal atresias occur as a result of intestinal volvulus, intussusception, internal hernia or strangulation in a tight gastroschisis or omphalocele defect.

Our diligent search could not find any such study with triple atresias involving jejunum, ileum and colon, with imperforate anus in a female newborn with Down's syndrome. This rarity and mode of presentation make it an interesting case to be reported.

Case report :

Female patient of age 34, G2P1L1 with 39 weeks gestation with three previous lower segment caesarean sections (LSCS) delivered a full term female baby weighing 2.75 kgs. Baby cried immediately after birth. Umbilical cord clamped and cut under aseptic precaution wherein 2 arteries and 1 vein was found. APGAR score was 7 at 1 min & 8 at 5 min, naso-gastric tube was passed till stomach and tracheo oesophageal fistula was ruled out.

Heart rate was 142/min and respiration rate was 42/min. There was abdominal distension and imperforate anus at birth. Central nervous examination was according to gestational age. Antenatal ultrasonography (USG) revealed dilated stomach, duodenum & proximal small bowel loops. Post natal USG revealed right Kidney measuring 3.6cmx1.6 cm with evidence of extra renal pelvis with mild hydronephrosis. Rest USG finding was within normal limits. Echocardiography showed small Patent ductus arteriosus with left to right shunt, small atrial septal defect. Karyotyping revealed trisomy of chromosome 21.

Baby was taken for emergency laparotomy which revealed very rare combinations of anomalies. Patient had jejunum-ileo- colonic atresia [Figure 1,2,3]. This is a one of the rarest combination of anomalies. It is usually seen in a patient with Down's syndrome with anorectal malformation. Urethral and vaginal opening were distinctly present. An abnormal fistulous tract could not be demonstrated in the perineum. Appendix was found to be inflamed and hence appendectomy was done.

Histopathological report of atretic segment of jejunum, ileum and colon showed no specific cellular pathology. Post-operatively patient recovered well from general anaesthesia and was administered on routine antibiotics and other medication. However, patient did not survive and died on post operative day 2 as a result of cardio respiratory arrest due to pulmonary haemorrhage.

II. Discussion

Two major theories regarding etiology of intestinal atresia are Tandler's concept of lack of revacuolization of solid cord stage of intestinal development and classic study of Louw and Barnard suggesting a late intrauterine mesenteric vascular accident is the cause of most jejunum-ileal and colonic atresias[3,4]. Familial instances of jejunum-ileal and colonic atresias have also been observed suggesting that genetics plays a part in these cases [2].

The first colostomy was performed in 1783 [5]. In a Retrospective study by Laura K et al. of 277 neonates with intestinal atresia & stenosis treated from July 1 1972 to April 30 1997, operative mortality for neonates with duodenal atresia was 4%, with jejunum-ileal atresia it was 0.8% and with colonic atresia it was 0%. [2] In this study, 5% cases had imperforate anus associated with duodenal atresia, 28% cases had malrotation

associated with duodenal atresia and duodenal obstruction was observed in 16% of neonates with duodenal atresia .

Jejuno- ileal atresia with duodenal atresia was found 3.6% of cases[2]. Colonic atresia was found 1.5% of cases with duodenal atresia. Ladd Procedure with appendectomy was performed in 28% of cases with malrotation of the gut. [5] In our case, newborn female did not survive after under going ileao-duodenal and jejunal resection and anastomosis with appendectomy with colostomy inspite of efforts in line with the set protocols. However, the rarity of this case and mode of presentation make it an interesting case to be reported.

III. Conclusion

Triple atresias is a very uncommon scenario. Jejuno-ileo-colonic atresia with imperforate anus with Down's syndrome is a real rarest variety of case to be seen and treat. Such atypical presenatations need to be reported to know such entities and deal with them.

References

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Figures

