



A RARE CASE OF HIGH ANORECTAL MALFORMATION ASSOCIATED WITH MECKEL'S DIVERTICULUM AND MALROTATION OF MIDGUT

Paediatric Surgery

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ABSTRACT

Anorectal malformations are among the most common congenital anomalies encountered in paediatric surgery, with an approximated incidence ranging between 1 in 2000 to 1 in 5000 live births. Their association with Meckel's diverticulum and Malrotation of midgut is very rare and to the best of our knowledge has not been reported previously in the literature. Here we report a case of neonate with High Anorectal malformation with an incidental findings of Malrotation of midgut and Meckel's diverticulum intraoperatively.

KEYWORDS

Anorectal Malformation, Meckel's Diverticulum, Malrotation

INTRODUCTION:

Anorectal malformations (ARM) are among the most common congenital anomalies encountered in paediatric surgery, with an approximated incidence ranging between 1 in 2000 to 1 in 5000 live births (1). ARM may present with a wide spectrum of defect ranging from relatively low type of malformation to very complex high type of malformation. Meckel's diverticulum (MD), an omphalomesenteric (vitelline) duct remnant present on the antimesenteric border of the distal ileum, has a prevalence of 1.5 -2% (2). Malrotation of the intestines, conjointly cited as intestinal rotation abnormality, happens in 0.5% of the population and may result in devastating consequences like volvulus, ischemic bowel, short bowel syndrome, or maybe death (3). The condition results from abnormal rotation of the intestines during embryological development and is most often diagnosed in the first year of life (3). The true incidence of malrotation is not fully known; however, it is reported as approximately 1 in 500 live births (4).

The association between ARM with MD, ARM with Malrotation and also MD with Malrotation have been reported earlier. However, an association of ARM, MD with malrotation together in a same patient is very rare and to the best of our knowledge has not been reported previously in the literature.

Case history:

The present case came to department of Surgery at R.C.S.M. Government medical college and hospital, Kolhapur. It was a 1-day old male baby delivered to a 28-year-old woman by spontaneous vaginal delivery. Child cried immediately after birth and birth weight of child was 1.9 kg and term baby. Mother had no comorbidities and she was P2L2A0. Clinical examinations revealed absence of anal opening and anal area showed pigmentation with presence of median raphe with anal dimple making the diagnosis of Imperforate anus (Figure No. 1). On further evaluation, it was found that baby had meconuria too. Invertogram done on 2nd day of life showed high type anorectal malformation (Figure No.2). Prenatal scanning was reported as normal. Vitals were normal. The abdomen was distended with no organomegaly.

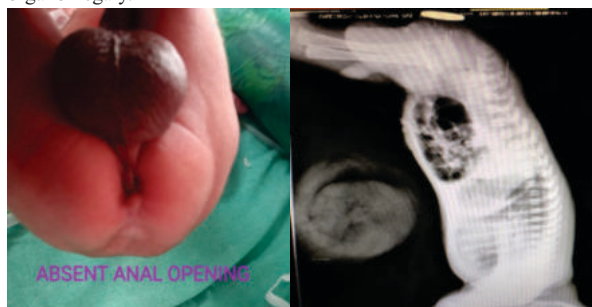


Figure No.1- Absent anal opening

Figure No.2- Invertogram showing ARM

Child posted for surgery on day 2 of life with ventilator and NICU back up. Surgery plan was transverse colostomy. But intraoperatively there was incidental findings of caecum at the epigastric region and DJ Flexure over the right side of abdomen (Figure No.3) suggestive of midgut Malrotation and presence of Meckel's diverticulum about 30 cm away from I-C junction (Figure No.4) and evidence of distended colon. Malrotation corrected & Transverse colostomy done. Meckel's diverticulum left in situ as it is, as neck of diverticulum was wide and also there was no signs of inflammation over the ileum. Patient shifted to NICU with ventilator support. On postoperative day 4, patient lost due to septicemia and low birth weight.

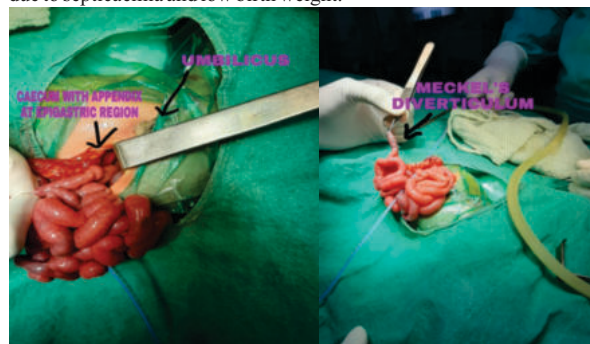


Figure No.3- Caecum with Appendix at epigastric Region with umbilicus

Figure No. 4- Meckel's Diverticulum

DISCUSSION:

In high malformations, the rectum terminates in the urinary tract through a recto-urinary fistula (5). In low malformations, the rectum terminates within about 1 cm from the skin, there is no recto-urinary communication and often the meconium shows up in the perineum through a small opening (perineal fistula) either at the site of normal anus or along the median raphe in the scrotum (ano-cutaneous fistula) (5). 15 % have VACTERL Association, which comprises vertebral anomalies, anal atresia, cardiac malformations, trachea-oesophageal fistula, renal anomalies and limb abnormalities.

MD is usually asymptomatic but may presents with complications like inflammation, gastrointestinal bleeding, intestinal obstruction and intussusception. In 1598, Wilhelm Fabricius Hildanus, a German doctor, first delineated the presence of a diverticulum of the small bowel occurring within the distal part of the ileum, that soon referred to as by the last name of the German anatomist, Johann Friedrich Meckel who first delineated its embryological origin in the year 1809 (6).

Malrotation can cause a large varieties of clinical symptoms. Infants with malrotation that has progressed to volvulus present with bilious vomiting and symptoms of acute bowel obstruction, including obstipation and abdominal distension. These cases most commonly present in the first month of life and 90% present within the first year.

Malrotation may also manifest as recurrent episodes of bilious or nonbilious vomiting, failure to thrive, solid food intolerance, malabsorption, diarrhoea, bloating, and abdominal pain (7). Malrotation describes a congenital anomaly of bowel development in which failure of the normal intestinal rotation at 10–11 weeks' gestation results in an abnormally positioned midgut (8). In few cases, fibrous bands cross the duodenum and cause early high ileus, often presenting within the primary 3 weeks of life.

The association between ARM with MD, ARM with Malrotation and also MD with Malrotation have been reported earlier. However, an association of ARM, MD with malrotation together in a same patient is very rare and to the best of our knowledge has not been reported previously in the literature.

ARM and intestinal malrotation rarely occur in the same patient. Some sporadic reports exist in the literature (9). The association of Imperforate Anus, Malrotation and Hirschsprung's Disease which is very rare has also been mentioned in the literature (10). Germana Casaccia et al. reported 2 cases of ARM associated with Meckel's diverticulum, Tracheoesophageal atresia (type III), duodenal atresia (type II) (11). Few cases of concurrent Meckel's diverticulum and malrotation together have been described in the literature to date (2,12-13).

CONCLUSIONS:

The combination of Anorectal Malformation, Meckel's Diverticulum with Malrotation is an extremely rare event and surgical intervention is needed. Our experience demonstrates that abdominal exploration may be needed for some neonate with Anorectal malformations, when there is suspicious of other bowel anomalies.

Declaration of patient consent The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest:

There are no conflicts of interest.

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