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Case Report

Anorectal malformation with apple peel atresia: case report of a rare association

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ABSTRACT

Anorectal malformations (ARM) are common congenital malformations. They are mostly associated with other congenital anomalies including but not limited to vertebral anomalies, cardiac malformations, Tracheoesophageal fistula, esophageal atresia, Renal anomalies, limb anomalies and aneuploidy (Trisomy 21). Apple-peel atresia or 'Christmas-tree deformity' consists of a high jejunal atresia with discontinuity of the small bowel and a wide gap in the mesentery. Association of Apple peel atresia with ARM has been reported by many authors in past and in any neonate presenting with ARM proximal atresia needs to be ruled out before surgical intervention is undertaken and newborns with ARM and presenting with drooling of saliva, bilious or non-bilious vomiting in presence of non-distended abdomen must give rise to a suspicion of associated atresia. We are reporting this case of high anal atresia with apple peel atresia of terminal ileum who was treated by double barrel ileostom to emphasize the importance of knowing this association.

Keywords: Anorectal malformations, Apple peel atresia, Associated anomalies, Double barrel ileostomy

INTRODUCTION

Anorectal malformations (ARM) are amongst the common congenital malformations with an incidence of approximately 1 in 1862 in eastern India.[1] Many types of gastrointestinal malformations have been described with ARM. Oesophageal atresia is the commonest gastro-intestinal association, found in approximately 10% of the patients. Duodenal atresia occurs in 2-3% of ARM patients.

Jejuno-ileal and colonic atresias have previously been described in various case reports.[2] Etiopathogenesis of atresia points towards vascular accidents much later in the embryological time-frame and Prematurity has found to have probably some relationship to immaturity of blood vessels leading to vascular insult to the developing gut, giving rise to atresias. Apple-peel atresia or 'Christmas-tree deformity' consists of a high jejunal atresia with discontinuity of the small bowel and a wide gap in the mesentery. The distal segment of the 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, which presents with a spectrum of occlusions of one or more branches of the superior mesenteric artery.[3]

Newborns with ARM and presenting with drooling of saliva, bilious or non-bilious vomiting in presence of non-distended abdomen must give rise to a suspicion of associated atresia in the mind of the attending surgeon. Since anorectal malformations themselves are cause of a

lot of concern and morbidity sometimes associated atresias are missed unless due importance is given to associated symptoms which are not explained solely by anorectal malformations. Since it is not uncommon that an internal atresia is overshadowed by more visible anorectal malformations a careful history, thorough clinical examination and appropriate imaging is necessary so as to not miss the proximal atresia in these neonates3. We here report a case of ARM associated with jejunoilleal apple peel atresia who was treated by double barrel ileostomy.

CASE REPORT

A 4-day-old male neonate, very low birth weight (1.4Kg), premature (32 weeks), first born child of non-consanguineous parents presented to our pediatric emergency with complaint of absence of anus. Upon history the antenatal history was uneventful. Mother received 2 tetanus toxoid injections. There was no history of fever or any other significant illness during antenatal period and mother did not take any medications other than iron and folic acid tablets. The delivery was normal vaginal delivery and the baby reportedly cried immediately after birth. On day 3 of life since the baby didn't pass stool mother checked and found that there was no anal opening for which he was brought to our department. On admission the neonate was lethargic and dehydrated and there was presence of respiratory distress in the form of intercostal and subcostal retractions. Abdominal distension was also present. On examination anus was found to be absent with poorly developed gluteal muscle suggesting high ARM. Spine and sacrum appeared normal. Straight x-ray and Invertogram showed few gas levels in upper abdomen without any visible gas shadow in the lower abdomen. The distance between gas shadow and the coin on Invertogram was found to be more than 3 cm suggestive of high anorectal malformation [Figure 1].

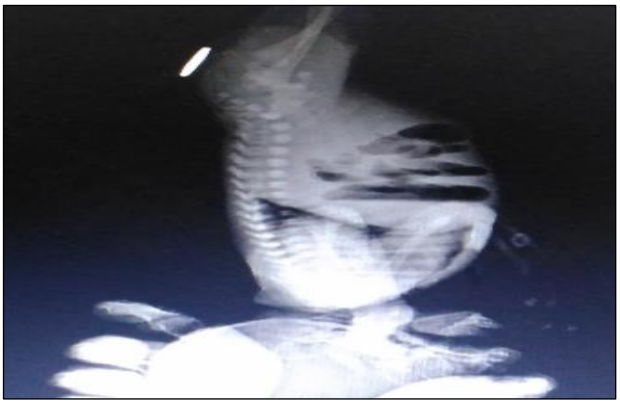

Figure 1: Invertogram showing absence of gas shadow in lower abdomen. Note the distance between anal verge and gas shadow s/o high arm.

Initially the preference was given to stabilization of neonate. A nasogastric tube was put, and aspiration was done every 2 hourlies. Baby was kept nil by mouth and appropriate intravenous fluid and antibiotics were started. Electrolyte imbalance and metabolic acidosis was corrected. Oxygen inhalation was also started. After initial stabilization and appropriate preoperative investigations, the baby was taken for surgery. Initially transverse colostomy and transverse colon was delivered out of the incision upon examination of the colon it was found to be microcolon hence a decision was taken for laparotomy. Upon laparotomy apple peel atresia of terminal ileum was found [Figure 2].

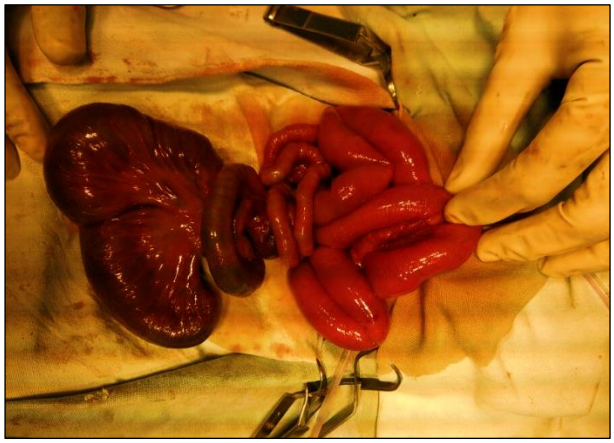

Figure 2: Intraoperative photograph showing apple peel type of intestinal atresia.

The apple peel atretic part with no mesentery was resected. Distal patency of the 10 cm terminal ileum was confirmed, and double barrel ileostomy was done. During operative period the baby was thermodynamically stable. Later the baby succumbed to complication on postoperative day 4.

DISCUSSION

Anorectal malformations including anal atresia are one of the common congenital malformations which manifests in neonatal period particularly immediately after birth. One of the important aspects of managing these neonates is to remember that these anorectal malformations are commonly associated with many other malformations which are described popularly by acronym such as VACTERL (vertebral anomalies, anal atresia, cardiac malformations, tracheoesophageal fistula, esophageal atresia, renal anomalies, radial aplasia and limb anomalies).

It is important that all such associated congenital malformations must be ruled out in any patients presenting with Anorectal malformations. Moreover, its association with various chromosomal anomalies particularly trisomy 21 has also been reported by many authors.[4] The incidence of isolated anal atresia is very low (around 20%) and majority of the patients (80%) of the neonates having anal atresia will have some or the

other associated congenital anomalies or malformations. Brantberg A et al in their study of 69 cases with imperforate anus found that additional anomalies, most of them diagnosed prenatally, were present in 59/69 (85.5%) of the cases.[5] The most frequent additional anomalies were found to be urogenital (53.6%).

The karyotype was abnormal in nine cases (13.0%). A retrospective evaluation of available videotapes of 22 cases of imperforate anus that were not diagnosed prenatally revealed that it was possible to suspect the diagnosis in 11/22 (50%) cases. Sixteen infants were born with imperforate anus without prenatal diagnosis of any abnormality.

Twenty-four (34.8%) infants survived, including all 10 with isolated imperforate anus and seven of eight cases with only one additional anomaly. The authors concluded that examiners should intensify their search for typical findings of imperforate anus especially when other anomalies that frequently accompany this condition are present. Moreover, this study also underlines the fact that isolated imperforate, though uncommon, is associated with a better prognosis as compared to those neonates in whom additional anomalies are present.

In 1961, Santulli and Blanc used the term 'apple-peel atresia' for the first time. Helen Blyth and Dickson JA reported cases with apple peel small bowel.[6-8] In these case reports the authors described characteristic 'Christmas tree', 'maypole' or 'apple peel' intestinal atresias in infants.

Zwiren GT et al were the first authors to stress upon the agenesis of dorsal mesentery in cases with apple peel artresia and Leonidas JC described the imaging feature in which there was Prenatal occlusion of the superior mesenteric artery resulting in a distinct type of distal duodenal or proximal jejunal atresia in which the dorsal mesentery is absent and the distal small bowel assumes a spiral configuration around its vascular stalk, strongly resembling an apple peel.[9,10]

The authors reported that in some instances the condition is transmitted genetically as an autosomal recessive disorder. In their study the authors found the common cause of mortality in these patients to be necrotizing enterocolitis which once again points towards the vascular insufficiency seen in this anomaly.

Since its initial descriptions there are various case reports describing the association of Anorectal malformations with that of apple peel atresia emphasizing the importance of ruling out this atresia in any patient presenting with ARM since missing it would result in revision surgeries and unacceptably high morbidity and mortality.

CONCLUSION

Association of ARM with apple peel atresia, though uncommon, is known. Treating pediatric surgeons must be aware of this entity and in any neonate presenting with ARM intestinal atresia must be ruled out by appropriate imaging techniques before surgical interventions are undertake.

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