CASE REPORT

Hereditary Multiple Intestinal Atresia association with Pyloric Atresia, Duodenal Atresia and Choledochal Cyst – A Rare Association

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Abstract:

Choledochal cyst is a rare congenital dilation of bile ducts. Its association with multiple intestinal atresia like pyloric atresia, duodenal atresia, and whole bowel atresia is an extremely rare condition. We report here a case of hereditary multiple intestinal atresia with choledochal cyst due to its rarity.

Key words – Choledochal Cyst, Duodenal Atresia, Hereditary, Intestinal Atresia, Pyloric Atresia.

How to cite this article: Dinesh Kumar Barolia, Pradeep Kumar Gupta, Pramila Sharma, Arvind Kumar Shukla Hereditary Multiple Intestinal Atresia association with Pyloric Atresia, Duodenal Atresia and Choledochal Cyst – A Rare Association. Walawalkar International Medical Journal 2018; 5(2):53-57. http://www.wimjournal.com

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Received date: 05/11/2018 Revised date: 17/11/2018 Accepted date: 22/11/2018

Introduction:

Hereditary multiple intestinal atresia is a rare disease of neonates. Intestinal atresia may occur from stomach to rectum. Guttman et.al.reported that hereditary multiple intestinal atresia is an autosomal recessive disease in 1973[1].Bilodeau et al reported that it is a universally fatal disease [2]. Lumen of small intestine and large bowel is atretic in whole length with calcification of epithelium of lumen. Fernandez et.al. reported that mutations of the tetratricopeptide repeat domain–7A (TTC7A) gene lead to hereditary multiple intestinal atresias. Congenital pyloric atresia is an extremely rare anomaly, first described by Calder in 1749 [3,4].Isolated congenital atresia have good prognosis but associated with other congenital anomalies like hereditary multiple intestinal atresia have fatal outcome.

Case report:

A two days old male child weighing 1.9 kilogram born full term normal vaginal delivery admitted here with presenting complaint of non-bilious vomiting after each feed. Clinical examination of baby showed flat abdomen with epigastric fullness, well developed perineum with anus. Baby was dehydrated. X-ray abdomen showed single bubble sign. Upper GI contrast study showed dye not passed beyond the pylorus of stomach. Pyloric atresia was our probable diagnosis. After admission hydration and electrolytes were corrected according to arterial blood gas analysis report. Baby was posted for laparotomy. On exploration following findings were found (1) dilated stomach with pyloric web (2) duodenal web at duodeno-jejunal junction (3) cystic dilation of common bile duct {choledochal cyst} (4) multiple small bowel atresia with meconium beads (5) multiple large bowel atresia with meconium beads.

A longitudinal incision was given over pylorus and pyloric web was excised. The longitudinal incision of pylorus was closed in transverse manner, known as heineke-mikulicz pyloroplasty. Same procedure was done at duodenalweb, longitudinal incision was given, and web excised and transversely repairs. We got few length of small bowel after heineke-mikulicz anastomosis and distal end exteriorized. Choledochal cyst was drained by tube cystostomy. Baby was taken on total parenteral nutrition post operatively. Up to the third Post-operative day's patients' general condition was good with well cry. Due to family problem father of baby took him home even after explanation of risk and prognosis.

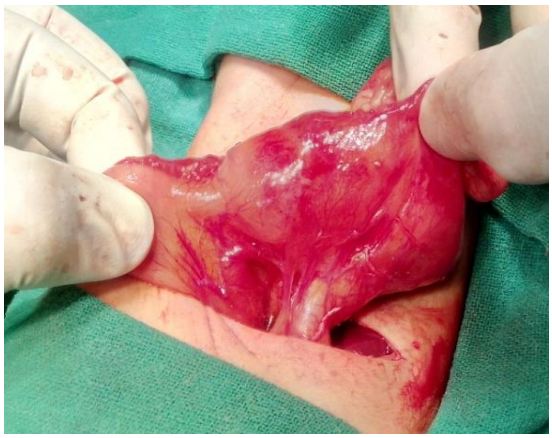

Fig-1

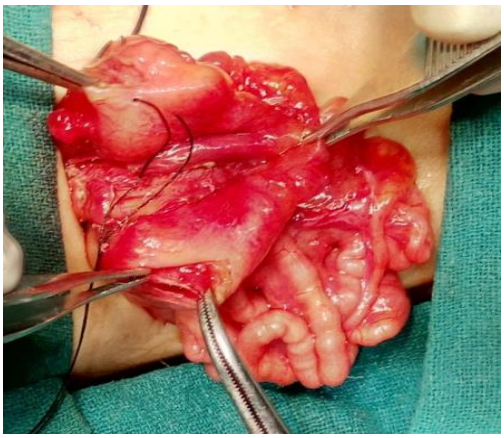

Fig-2

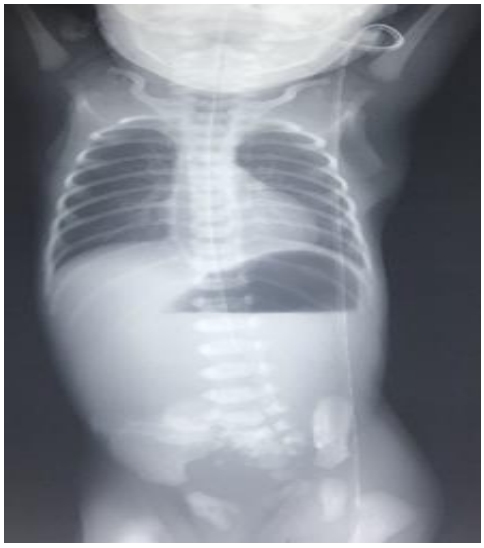

Fig –3

Fig – 1 showed pyloric atresia between distended stomach and distended duodenum and choledochal cyst.

Fig – 2 showed web at pylorus and duodenum with atretic small bowel with meconium beads visible impression.

Fig – 3 showed babygram of neonate with single bubble sign, indicate pyloric atresia.

Discussion:

Congenital choledochal cysts is a rare entity. Its incidence is 1:10,000 - 1:150,000 live births. Females are four times more affected than males. Congenital choledochal cysts have some features that made different from classical pediatric age group. (a) Cystic dilation of common bile duct, (b) lump Abdomen with jaundice and acholic stools, (3) No symptomatic association occur with acute pancreatitis and (4) level of amylase is lowinbile [5].We repot here congenital choledochal cyst in male baby, who has less incidence than female. In our case common bile duct was dilated as cystic form and during tube cystostomy showed white bile. The white bile indicates that biliary drainage was obstructed for long time, during fetal life.

Congenital pyloric atresia is an extremely rare entity, having incidence of 1 in 1, 00,000 newborns. The incidence of pyloric atresia is 1% of all types intestinal atresia [3,4]. It is classified in three types - type1 is pyloric web or membrane, type 2 is pyloric atresia with a solid cord between the two atretic ends, type 3 is pyloric atresia with a gap between the stomach and duodenum [6]. In this case we found type 1 pyloric atresia. Pyloric atresia may be associated with following anomalies like epidermolysis bullosa (EB), aplasia cutis congenita, duodenal atresia, ileal or colonic atresia, esophageal atresia with distal trachea-esophageal fistula, sensori-neural deafness and dysplastic kidney [7,8]. In our case pyloric atresia was associated with congenital choledochal cyst, duodenal atresia and hereditary multiple intestinal atresia.

Conclusion:

It is very difficult to deal case of hereditary multiple intestinal atresia because surgeons don't have any option for atretic bowel in whole length. Even after multiple resection and anastomosis may leads to Short bowel syndrome. Hereditary multiple intestinal atresia association with pyloric atresia, duodenal atresia and choledochal cyst is extremely rare association.

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