ejpmr, 2018,5(1), 265-267

SJIF Impact Factor 4.161

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Report

ISSN 2394-3211

EJPMR

CONGENITAL PYLORIC ATRESIA IN A 2 MONTHS OLD FEMALE CHILD- A RARITY

Dr. Kamal Nain Rattan[1], Dr. Rashmi Hooda*[2] and Dr. Deepali Garg[3]

[1]Sr. Professor Department Paediatric Surgery, Pt. B.D. Sharma Postgraduate Institute of Medical Sciences, Rohtak, India.

[2]Senior Resident, Department of Pediatrics, Pt. B.D. Sharma Postgraduate Institute of Medical Sciences, Rohtak, India.

[3]Junior Resident, Department of Pediatrics, Pt. B.D. Sharma Postgraduate Institute of Medical Sciences, Rohtak, India.

***Corresponding Author: Dr. Rashmi Hooda**

Senior Resident, Department of Pediatrics, Pt. B.D. Sharma Postgraduate Institute of Medical Sciences, Rohtak, India.

Article Received on 05/11/2017 **Article Revised on 26/11/2017** **Article Accepted on 17/12/2017**

ABSTRACT

Congenital pyloric atresia(CPA) is an uncommon cause of upper gastrointestinal tract obstruction. Type 1 is the most common out of the three types. This often results in incomplete obstruction leading to misdiagnosis and late presentation. The outcome is good if there is isolated CPA. We are reporting a case of CPA in a 2 month old child presenting with on and off non-bilious vomiting. The case was managed successfully by surgical exploration and Heineke-Mikulicz pyloroplasty. Because of rarity, we are prompted to report this case with a wide review of literature.

KEYWORDS: Congenital pyloric atresia, epidermolysis bullosa, multiple intestinal atresias.

INTRODUCTION

CPA is very uncommon etiology of upper gastrointestinal obstruction in neonates. This condition was first described by Calder way back in 1749.[1] it accounts for less than 1% of all upper gastrointestinal atresias with an estimated incidence of 1:100,000 live births[1,2], with an equal occurrence between males and females. This is associated with low birth weight and polyhydramnios. This can occur in isolation or may be associated with other anomalies like epidermolysis bullosa (EB), aplasia congenital cutis (ACC) or hereditary multiple intestinal atresias (HMIA)\.[3,4] The association of pyloric atresia with epidermolysis bullosa is more than just coincidental. The pathogenesis of this syndrome was first described by Carmi, so this is also called as the 'Carmi Syndrome'. The outcome is very good in cases of isolated pyloric atresias, but the mortality increases with the presence of other associated abnormalities.[5,6]

CASE REPORT

We are reporting a case of congenital pyloric atresia in a 2 months old female child. The child was a product of non-consanguinous marriage, born full term with birth weight of 3.2 kg by normal vaginal delivery, cried immediately after birth. The mother was 28 years old, booked and immunized with three antenatal scans, all of which were normal. The antenatal period was uneventful. The child was fed within 1 hour of birth and was exclusively breast fed thereafter. There was history of on and off non bilious vomiting for 1 month, for which the child was shown to some private practitioner, but since the child was not relieved, she was referred to tertiary care centre. At presentation, there was failure to thrive with presenting weight of 2.9 kg and visible gastric peristalsis. So, a provisional diagnosis of hypertrophic pyloric stenosis was kept. Ultrasound abdomen showed dilated stomach with no hypertrophic pylorus visible. Barium study was done which showed hugely dilated stomach (Fig 1). The child was admitted and started on parenteral antibiotics and intravenous fluids. After correction of hemodynamics and electrolyte abnormalities, the child was taken up for surgery. The abdomen was opened by right supraumbilical transverse incision, the stomach was massively dilated and hypertrophied, and pylorus was normal with no evidence of hypertrophic pyloric stenosis. Duodenum was not dilated. A longitudinal incision was made at pylorus and pyloric web was identified and excised, Heineke-Mikulicz pyloroplasty was done. The abdomen was closed in layers. The child was kept nil per oral for 72 hours and started orally once she had passed stools. The child was discharged after 7 days and stitch removal was done on tenth day. The child is doing well on follow up.

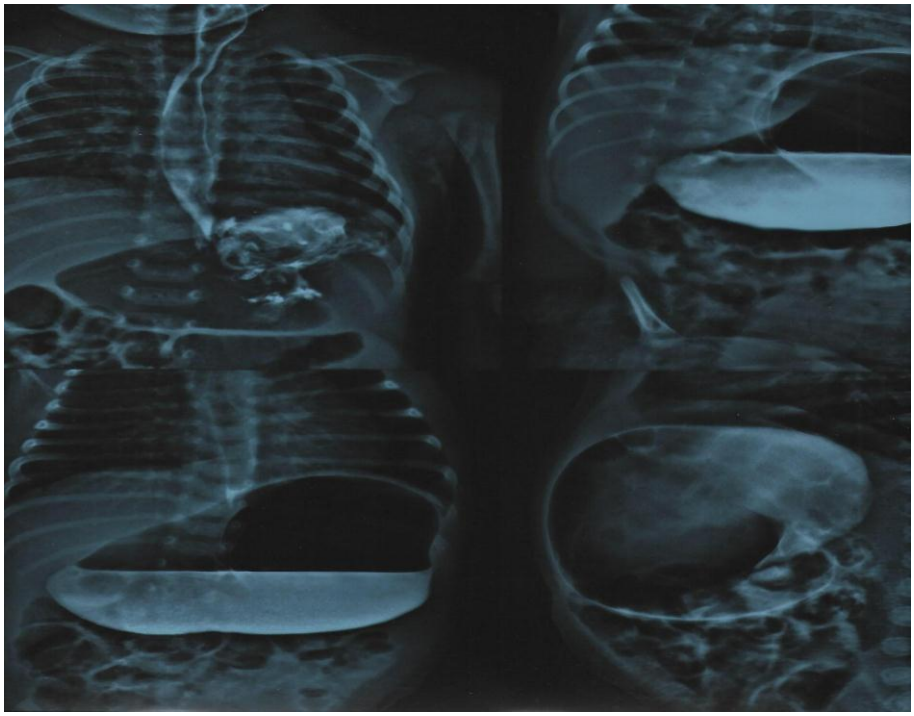

Fig 1: Barium meal follow through showing distended stomach and pylorus suggestive of pyloric atresia.

Discussion: This condition is inherited as an autosomal recessive condition.[7] The exact etiology of this condition is not very clear. It has been postulated to result due to developmental arrest between 5^{th} -12^{th} week of gestation.[8] The most widely accepted theory is that it results due to failure of canalization of pyloric canal.[9]

Anatomically, it is divided into 3 types: type A consists of pyloric web or diaphragm, type B is associated with a solid cord between the pylorus nd the duodenum whereas in type C, there is gap between the pylorus and duodenum. Type A is most common with an estimated incidence of 54%, 34% and 9% respectively of types A,B and C.[10]

The association with epidermolysis bullosa is more than just coincidental. The epidermolysis bullosa can be of three types: epidermolysis bullosa simplex, junctional epidermolysis bullosa and dystrophic epidermolysis bullosa. CPA is most frequently reported with junctional epidermolysis bullosa. This condition is known by a distinct clinical syndrome known by the name of CPA-EB syndrome. Alpha 6 integrin and its ligands have been postulated to be responsible for this distinct clinical syndrome.[11,12] CPA is also reported to be associated with aplasia cutis congenital.[13] Another very interesting association of CPA is with hereditary multiple intestinal atresias (HMIA).[2,5] The x-ray abdomen in these patients shows the presence of multiple calcifications. These can be identified by the instillation of saline distally into the bowel.

The antenatal diagnosis can be made in the second trimester by ultrasonographic findings of low birth weight and polyhydramnios. The clinical features are non-bilious vomiting and abdominal distension. The diagnosis is made by the presence of single bubble on abdominal x-ray.

In the index case, the prognosis was good, as it was an isolated CPA. However, the outcome of cases associated with epidermolysis bullosa is poor due to sepsis and electrolyte abnormalities. But the surgery for CPA can still be done as the prognosis of EB is improving in view of recent advancements in the management. The surgery can be done after correction of electrolyte abnormalities and gastric decompression by feeding tube. The type 1 and 2 atresias can be managed by excision of the membrane and Heineke-Mikulicz or Finney pyloroplasty. Pyloroduodenostomy can be done for those with a gap between the pylorus and duodenum. Gastrojejunostomy should be avoided as there are high chances of ulcer formation. It is important to explore the whole intestine to look for additional strictures.

CONCLUSION

In an infant presenting with on and off non bilious vomiting, the diagnosis of congenital pyloric atresia, although rare, should be kept in mind, and should be managed early so as to prevent morbidity and mortality.

REFERENCES

1. Moore CM. Congenital gastric outlet obstruction. J Pediatr Surg, 1989; 24: 1241-6.
2. Ilce BZ, Erdogan E, Kara C, et al: Pyloric atresia. 15-year review from a single institution. J Pediatr Surg, 2003; 38: 1581-4.
3. Tomà P, Mengozzi E, Dell'Acqua A. Pyloric atresia: report of two cases (one associated with epidermolysis bullosa and one associated with

multiple intestinal atresias). Pediatr Radiol, 2002; 32: 552-5.
4. Carmi R, Sofer S, Karphus M. Aplasia cutis congenita in two sibs discordant for pyloric atresia. Am J Med Genet, 1982; 11: 319-28.
5. Al-Salem AH: Congenital pyloric atresia and associated anomalies. Pediatric Surg Int, 2007; 23: 559-63.
6. Kajal P, Rattan KN, Bhutani N. Congenital pyloric atresia: Early and delayed presentations—A single centre experience of a rare anomaly. Indian J Gastroenterol, 2016; 35: 232.
7. Usta IM. Familial pyloric atresia: Report of a family and review of the literature. J Matern Fetal Med., 2000; 9: 190-3.
8. Snyder CL, Mancini ML, Kennedy AP, Amoury RA. Multiple gastrointestinal atresias with cystic dilatation of biliary ducts. Pediatr Surg Int, 2000; 16: 211–3.
9. Semrin M, Russo M. Anatomy, histology, embryology, and developmental anomalies of the stomach and duodenum. In: Feldman M, Friedman L, Brandt L, eds. Sleisenger and Fordtran's Gastrointestinal and Liver Disease. 9th ed. St. Louis, MO: Saunders, 2010; 773-88.
10. Al-Salem AH, Abdulla MR, Kothari MR, Naga MI. Congenital pyloric atresia, presentation, management, and outcome: a report of 20 cases. J Pediatr Surg, 2014; 49: 1078–82.
11. Varki R, Sadowski S, Pfendner E. Epidermolysis bullosa. I. Molecular genetics of the junctional and hemidesmosomal variants. J Med Genet, 2006; 43: 641-52.
12. Pfendner E, Uitto J. Plectin gene mutations can cause epidermolysis bullosa with pyloric atresia. J Invest Dermatol, 2005; 124: 111-5.
13. Achiron R, Hamiel-Pinchas O, Engelberg S, et al: Aplasia cutis congenital associated with epydermolisis bullosa and pyloric atresia: the diagnostic role of prenatal ultrasonography. Prenatal Diagn, 1992; 12: 765-71.