

Case Report

Type B congenital pyloric atresia: a case report

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ABSTRACT

This is a case of congenital pyloric atresia type B not associated with any other anomaly rare condition, seen as an isolated anomaly with excellent prognosis) of two days female child presenting with vomiting since birth. X-ray abdomen showed only stomach air bubble, sonography showed distended stomach with echoes and on barium meal no passage of contrast was seen beyond pyloric antrum even after 24 hours. Patient underwent Heineke-Mikulicz pyloroplasty and postoperative recovery was uneventful.

Keywords: Congenital pyloric atresia, Heineke-Mikulicz pyloroplasty, Neonatal intestinal obstruction

INTRODUCTION

Pyloric atresia is a rare congenital anomaly that causes partial or complete obliteration of the gastric lumen. Its actual incidence is not known, but it is thought to be forming less than 1% of all bowel atresias, giving it an estimated incidence of about 1 in 100,000 live births.^{1,2} We had a case of female neonate presented with distended abdomen non-bilious vomiting since day 2 of life. On radiological evaluation diagnosis of congenital pyloric atresia without associated anomalies was made which was confirmed on surgical intraoperative findings.

CASE REPORT

A 4-day old female neonate presented with vomiting since day 2 of life was admitted. She was born after completed 9 months by normal vaginal delivery. Antenatal records were not available. Abdominal examination revealed upper abdominal distention. Plain x-ray abdomen showed gastric dilatation with no air distal to the pylorus i.e. single bubble appearance (Figure 1). Ultrasound examination showed the dilated stomach bubble with internal echoes however no evidence of compressing masses in the antral region was seen, pyloric

region was not seen with duodenum appeared to be normal (Figure 2).



Figure 1: Frontal chest and abdomen radiograph showing single stomach bubble with gasless abdomen.



Figure 2: Sonography of upper abdomen showing distended stomach with echogenic contents.

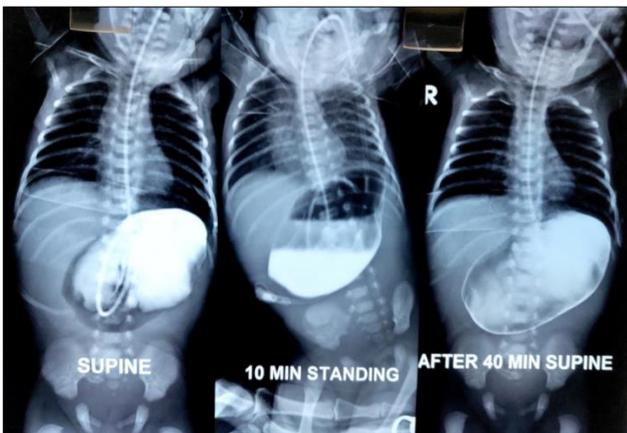


Figure 3: Upper GI gastro-graffin meal study showing non-passage of contrast distal to stomach on serial examination.

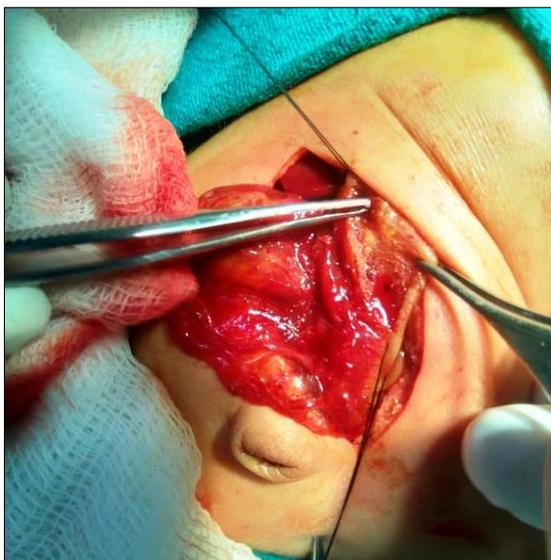


Figure 4: Intraoperative finding suggestive of cord like structure suggestive of type B pyloric atresia.

Upper contrast studies showed no passage of contrast distal to stomach suggesting diagnosis of congenital pyloric atresia (Figure 3).

On laparotomy, a solid cord connected the stomach to the duodenal bulb was seen which suggested pyloric atresia type B. Patient had undergone Heineke-Mikulicz pyloroplasty and postoperative recovery was uneventful (Figure 4).

DISCUSSION

Congenital pyloric atresia (CPA) attribute to less than 1% of all bowel atresia. With incidence of 1 in 100000 live births. The exact etio-pathogenesis of the CPA has not been recognized but is associated with factors like genetic predisposition, familial inheritance and potential intrauterine incidents such as failure of canalization or a vascular event. No sexual predilection is seen. CPA has three anatomic types, type A (membrane/web), type B (solid cord) and type C (gap between the stomach and duodenum). CPA in isolation is probably a familial autosomal recessive congenital disease. In half of the cases, it is associated with other syndromes viz autosomal recessive epidermolysis bullosa with aplasia cutis congenita. Less frequently, CPA come with association of hereditary multiple intestinal atresia syndrome which is a rare lethal autosomal recessive condition.^{3,4}

CPA can be diagnosed on level 2 antenatal scan as dilated stomach with polyhydramnios (single air-bubble). Postnatally neonate present with non-bilious vomiting and abdominal radiograph showed a single large gastric air-bubble with a gasless abdomen distally. Confirmation of diagnosis can be done on a barium meal but usually it is often not required. Ultrasound is a non-invasive examination which gives additional information by showing radiological anatomy of antro-pyloric region and ruling out other causes of gastric obstruction complimenting plain x-rays so that a specific radiological diagnosis can be achieved. Different types of pyloric atresia require different surgical procedures by paediatric surgeons.^{5,6}

Because of the rarity of CPA, the confirmation of the diagnosis often delayed. The cases can be diagnosed as proximal duodenal atresia which is more common. Therefore, a high index of suspicion is needed for diagnosis because delay in diagnosis can lead to complications like pulmonary aspiration, gastric perforation etc.⁴

CONCLUSION

The diagnosis and management congenital gastrointestinal lesion of a neonate with non-bilious vomiting of may be complex and requires high index of suspicion with reliable radiological diagnostic tools in order to achieve the most appropriate surgical management.

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