Congenital Intrinsic Duodenal Atresia (Type III) - A Rare Case Report

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INTRODUCTION

Congenital intrinsic duodenal atresia [CIDA] is one of the rare intestinal anomalies occurring in 1 in every 5000 - 10000 live births.[1] Among the various types, type III is an uncommon CIDA as compared with types I and II.[2] About two thirds of all congenital duodenal obstructions are intrinsic, characterized by onset of bilious vomiting occurring within 24 hrs. after birth.[3] Associated anomalies are frequently seen in nearly 50% of CIDA patients.[4] In this case report we describe a patient with CIDA Type 3 without any associated congenital anomalies.

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PRESENTATION OF CASE

A 2-day preterm neonate weighing 2.24 Kg was referred to the paediatric department with sudden onset of bilious vomiting and aspiration. Maternal history revealed pregnancy induced hypertension with hypothyroidism, polyhydramnios and a threatened labour. Prenatal ultrasound findings revealed duodenal atresia of the fetus (Figure 1). On examination, the neonate was alert, active, and had no obvious external morphological congenital anomalies. There was upper abdominal fullness and nasogastric tube drained bilious aspirate. External genitalia and anal opening were normal. Ophthalmological examination was normal. Further evaluation by X-ray plain abdomen showed double-bubble appearance indicating duodenal atresia with total absence of distal bowel gas (Figure 2). The neonate was delivered by breech presentation at 33 weeks of gestation and a nasogastric tube was passed immediately after birth and parenteral fluid was placed. Later, exploratory laparotomy was performed and specimen was sent for histopathological examination.

Macroscopy revealed that duodenum was atretic and there was marked dilation of proximal duodenum. Microscopy suggested abundant fibrosis with congested vessels and inflammatory cells confirming atresia. The neonate was apparently on ventilator for 23 days and on 25th day of life, baby expired due to sepsis and cardiac arrest.

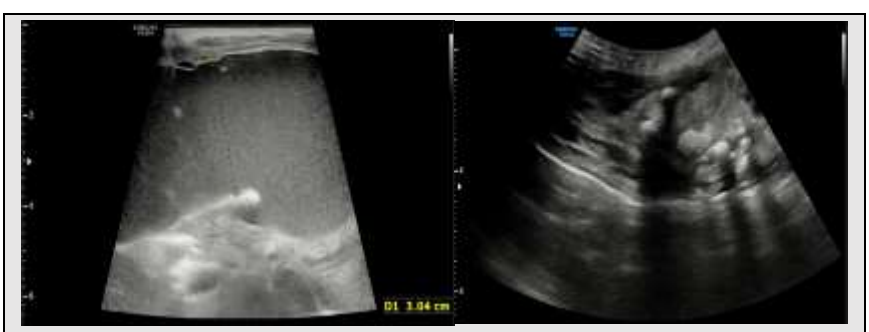

Figure 1. Prenatal Ultrasound
Revealed duodenal atresia of the fetus. Dilated stomach and duodenum in the upper abdominal quadrant

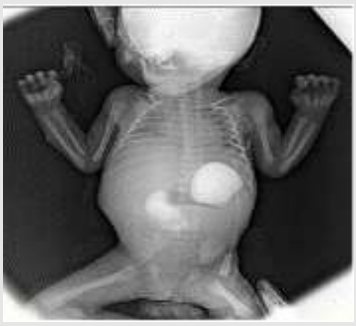

Figure 2. Postnatal Roentgenogram Demonstrated a Double-Bubble image

PATHOLOGICAL DISCUSSION

Figure 3

A segment of intestine with dilated proximal end and atretic distal end measuring 6.0 X 5.5 cms was received.

Microscopically- The proximal end of intestine consisted of mononuclear inflammatory infiltration, while the distal end consisted of abundant fibrosis with dilated and congested vessels. Muscular layer also appeared atrophic at the distal end.

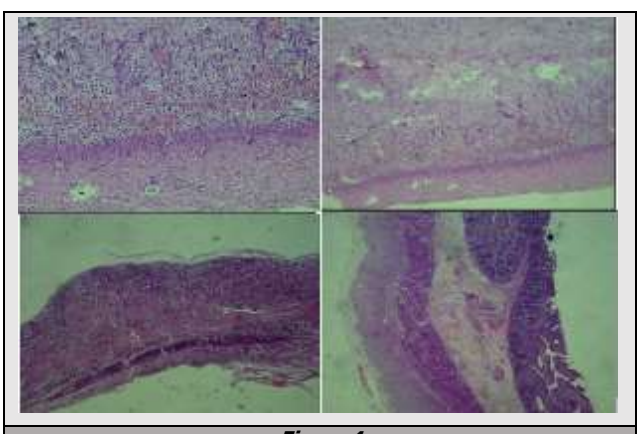

Figure 4

CIDA has been considered as a result of several embryologic defects in foregut development, canalization or rotation. Duodenal atresia is a congenital bowel obstruction usually seen in the second part of the duodenum. Both embryologically and clinically, it is a very interesting anomaly. In addition, unusual embryologic relationships between the duodenum and other close anatomic structures in the proximity can also lead to congenital duodenal obstruction the duodenal lumen can be obliterated by the rapidly growing epithelium between the fourth to sixth week of intrauterine life. Also, by the end of twelfth week recanalization of the duodenal lumen occurs. Hence, congenital duodenal atresia has been thought to result from either failure of recanalization or arrest of duodenal growth.[2,5] Unlike duodenal atresia, the atresias in other parts of the intestine usually results from an intrauterine vascular accident.[6] Congenital duodenal obstruction can be divided into intrinsic and extrinsic based on the etiopathogenesis. The causes for intrinsic obstruction include stenosis, atresia and duodenal diaphragm with or without a hole while extrinsic causes are annular pancreas, malrotation with congenital bands, duplication, and preduodenal portal vein.[7] The probable incidence of associated anomalies with congenital duodenal atresia appears variable.

Congenital heart disease and Down's syndrome still continue to be the most common anomalies associated with congenital duodenal obstruction. Other commonly associated anomalies are[2] prematurity, growth retardation, situs inversus, etc. Duodenal atresia has been classified into 3 types. In Type 1, there is an obstructing septum formed from the mucosa and submucosa with no defect in the muscularis, but the mesentery is intact. Type 2 atresia has a short fibrous cord that connects the 2 blind ends of duodenum and the mesentery is intact. Type 3 has no connection between the 2 blind ends of duodenum and there is a v shaped mesenteric defect[2]. Duodenal atresia can be diagnosed beginning from the early stages of pregnancy in line with developments in diagnostic methods. Upright plain abdominal radiograms of the newborns which demonstrate double air levels and distal bowel devoid of intestinal gas have diagnostic values[2]. A plain X-ray abdomen with a characteristic 'double-bubble' sign was observed here. Gastro duodenal dilation seen in ultrasound scan during prenatal period was diagnostic for duodenal atresia or stenosis.[2] In the present case report, ultrasonography performed at prenatal 29th week revealed dilated stomach and duodenum in the upper abdominal quadrant. The preoperative findings could not detect any associated anomaly. Although some studies have reported bilious vomiting as the most common presenting feature,[8] our patient had also bilious aspirate from the nasogastric drainage indicating that the site of obstruction was post ampullary. A prompt diagnosis of CIDA is very important to prevent various complications related to delayed diagnosis such as electrolyte imbalance, dehydration or aspiration pneumonia.[9] There was no history of vomiting in our patient as a nasogastric tube was passed immediately after birth and the neonate was placed on parenteral fluid until the diagnosis was confirmed. In cases with duodenal atresia, surgery is the treatment modality. CIDA can be fatal unless promptly diagnosed and treated surgically. Various surgical procedures like gastrostomy, trans anastomotic tube and reduction duodenoplasty have been attempted for congenital duodenal atresia, but with poor outcome. Duodeno-jejunostomy is considered a less physiological procedure when compared to duodeno-duodenostomy.[8] Weber et al suggested that diamond-shaped duodenoduodenostomy is superior to side-to-side duodenojejunostomy and side-to-side duodenoduodenostomy for repair of duodenal atresia, resulting in earlier feeding and discharge.[10] Postnatal 2nd day, the neonate was operated for duodenoduodenostomy.

The survival rate has increased to 88.6% during the last 10 years. The factors that have been crucial to this increased survival rate include: early diagnosis and extensive preoperative workup, improved surgical technique with a greater awareness for common pitfalls, expert administration of anesthesia, use of parenteral nutrition; and better intensive nursing care.[10] On 25th day of life, our patient expired due to sepsis and cardiac arrest. Mortality depends mainly upon the presence of sepsis, prematurity, very low birth weight and associated anomalies in developing countries.

FINAL DIAGNOSIS

Congenital Intrinsic Duodenal Atresia- Type 3

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