Case Report

Congenital Duodenal Diaphragm Presenting as Gastric Outlet Obstruction: A Rare Case with Review of Literature

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Abstract

Introduction: Duodenal obstruction is related to the orifice of the bile duct in 83% of the cases. 50% of cases of duodenal atresia have a true atresia, 40% cases have a duodenal diaphragm with or without central opening and 10% cases show stenosis [1]. Duodenal diaphragm develops due to failure of vacuolation of the proliferating epithelial lining of the duodenum [2]. Congenital obstruction of the duodenal accounts about one half of all the intestinal obstruction seen in the new born.

Case report: Full term female baby presented with multiple episode of vomiting, refusal of feeding and not gaining weight with severe dyselectrolytemia. Surgical intervention through excision of duodenal diaphragm with transverse closure of the duodenum (excision with duodenoplasty) was done along with post-operative balanced nutritional support.

Conclusion: Early diagnosis, surgical intervention, use of total parenteral nutrition and adequate investigations for congenital anomalies may improve the outcome.

INTRODUCTION

Congenital obstruction of the duodenum accounts about one half of all the intestinal obstruction seen in the new born. Duodenal atresia has incidence of 1 in 10,000 live births reported in literatures. Duodenal obstruction is related to the orifice of the bile duct in 83% of the cases. 50% of cases have a true duodenal atresia, 40% cases have a duodenal diaphragm with or without central opening and 10% cases show stenosis [1].

Embryologically, in the fifth week of gestation the primitive gut closes solidly with a proliferation of epithelial cells which recede by a process of vacuolization until bowel takes its final histological lining. It is believed that intrinsic malformation in development of duodenal diaphragm due to faulty recession of these cells [2].

CASE REPORT

22 year old mother gave birth to a full term female baby at 39 weeks of gestation through normal vaginal delivery at private hospital. Her birth weight is 2.6kgs. After 20 days of birth baby presented with 2 to 3 episodes of vomiting per day since 6 days, refusal of feeding since 6 days and failure to thrive. At the time of presentation her weight was 1.7kgs. Baby was in respiratory distress with oxygen saturation (SpO2) of 90% at room air, with poor tone, cry and activity including moderate effort to suck (Figure 1).

The investigations revealed severe dyselectrolytemia, altered renal parameters with moderate to severe dehydration and metabolic alkalosis (it would be nice if the actual figures of electrolytes, renal function and blood gases if possible and feasible please). On clinical grounds baby was provisionally diagnosed to have hypertrophic pyloric stenosis with pre renal acute kindney injury (AKI).

Baby was resuscitated with correction of severe dyselectrolytemia, severe dehydration and metabolic alkalosis with neutralising the pre renal azotemia. Corrected her nutritional status by transfusing rich plasma and packed RBCs.

On further evaluation, barium meal follow through study revealed distended stomach and first part of duodenum i.e., double bubble sign (Figure 2,3), further segments of duodenum was not visualised. Ultrasound scan (USS) of abdomen and pelvis suggested distended stomach and 1st part of duodenum leading to abrupt narrowing of the second part of duodenum,
immediately proximal to level of ampulla of Vater likely to be duodenal obstruction.

After stabilising, baby was taken up for operation, abdomen opened through transverse incision, distended stomach and 1st part of duodenum was obvious. Gastrostomy was carried out and no. 8F Foley’s catheter could not be negotiated through 2nd part of duodenum. Vertical incision was taken at the junction of dilated and collapsed duodenal loop. It was found to be thick duodenal diaphragm (Figure 4).

Stay sutures were taken and the web was excised circumferentially leaving the medial side to prevent injury to the ampulla and the margins were overrun with a continuous vicryl suture. The longitudinal incision of the duodenum was closed transversely by vicryl suture in two layers, here Foley’s catheter was brought through duodenum and has been used as feeding tube during post-operative period. This procedure was combined with incidental appendicectomy. Rest of the small intestine was normal. There was no evidence of malrotation and other anomalies. During post-operative period she was supplemented with partial parenteral nutrition for 3 days, transfused 1 unit of FFP, later initiated with feeding tube, baby passed stools on the 4th post-operative day (POD). Drained was removed on POD 8, gastrostomy tube was removed on POD 10. Operative wound was healthy. X-ray abdomen at the time discharge shows free flow of barium meal (Figure 5). At present baby is doing well and she is on regular follow up (Figure 6).
DISCUSSION

Duodenal diaphragm accounts only 2% of the congenital duodenal obstruction. Aetiology is unknown. In embryological development duodenum is lined by endoderm, which proliferates rapidly and obliterator the lumen between 8 to 10 weeks of gestation and duodenal branch canalised by 12th week of gestation. This period is considered crucial, any insult during this phase lead to duodenal diaphragm and atresia [3].

H. Forssner’s presents a classification which is complete and scientific. This classification can be used in all cases to aid investigation. He describes three types of duodenal obstruction

Type I- duodenal diaphragm - complete / incomplete
Type II- duodenal stenosis - complete / incomplete
Type III- duodenal atresia - blind ends connected by a band / unconnected blind ends [4].

Moreover >50% of new born with duodenal obstruction associated with other congenital anomalies like intestinal malrotation, situs inverses, congenital heart disease, tracheoesophageal fistula including trisomy 21 in 40% cases. In our case trisomy 21 is absent and not associated with other congenital anomalies [4].

In our cases baby presented 20 days after birth, with significant weight loss, severe dysselectrolytemia, metabolic alkalesis with pre renal azotemia, this explains the consequences of delayed diagnosis and few literatures quoted, death of neonates due to aspiration and respiratory arrest [5].

The duodenal diaphragm thickness ranges from few millimetres, contains mucosa fibres, epithelium on either side, located commonly at pre-ampullary position in the 2nd part of duodenum in 70% of cases. If the duodenal diaphragm is located in juxta-ampullary position than surgery and endoscopic resection is potentially dangerous [6,7].

Baby with congenital duodenal obstruction shows double bubble sign on x-ray abdomen which taken after birth, which can be identified by an antenatal ultrasound scan. However these finding may not be evident if screening USS done before 20 weeks of pregnancy, as in our case mother did not have periodic antenatal visits and relevant investigations. Polyhydromnios is the prenatal marker for diagnosis of upper gastro-intestinal obstruction in foetus during antenatal period in 50% of cases [8-10].

In our case baby was been treated with excision of duodenal diaphragm with duodenoplasty in two layers. Several reports of endoscopic incision or laser ablation also appear in the literature, but subsequent surgery may be needed because of the scar formation which results in stenosis [11]. Recently, endoscopic surgery with a high-frequency wave cutter assisted by suction apparatus or a balloon catheter pulling up the diaphragm from the distal side has been reported to be safe and effective [10]. Laparoscopic or laparoscopically assisted management also represents an alternative if the surgeon has the appropriate instruments and suturing skills [12].

CONCLUSION

Congenital duodenal obstruction needs to be diagnosed as early as possible, otherwise its consequences leads to death of neonate. It would be better all pregnant mother should have antenatal visits and scanning. If new born presented with persistent vomiting refusal of feeding the clinician should think of anatomical malformation of upper GI tract like tracheoesophageal fistula(TOF), duodenal obstruction and it’s variants like duodenal diaphragm, duodenal web in addition to correcting the metabolic disturbances.

REFERENCES

Manthal et al. (2017)
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