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Case Report

An unusual association: duodenal atresia and situs inversus incompletes-a case report and discussion

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ABSTRACT

Coming across a number of duodenal atresia in daily practice any paediatric surgeon may conclude that it's one of the most technically challenging small gut atresia with variations of pathology that has an incidence of 1 in 4000 or 5000 live births. It has marked association with a number of conditions like downs syndrome (trisomy 21) being on crest, accounting for 20-40% of cases. Other prominent associations include VACTERL (vertebral, anotectal malformation, cardiac, tracheo esophageal, renal, limb) anomalies, concurrent other small gut atresias, preduodenal portal vein. A relatively rare association we have come across recently has been situs transverus co existing with duodenal atresia that prompted us to rummage through literature for similar cases embarking us to publish our own finding as very little has been said so far in this regard.

Keywords: Atresia, Duodenum, Syndrome, Situs inversus

INTRODUCTION

Duodenal atresia is the incomplete formation or hindrance to continuity at the level of duodenum causing an obstruction at very early course of transit of contents, the incidence of this condition being 1:4000-5000 live births.[1]

The condition manifests itself frequently with a syndrome; Down's syndrome in as much as 40% of patients.[2] It remains one of the most easily picked up small gut atresia with a classic radiographic presentation of double bubble sign.

Having said that here we would like to proclaim the unusual syndicate we came across which had been reported as little as roughly 25 times in past. Situs inversus has an incidence of 1 in 10000 live births and its further connection with duodenal atresia is scarce.[3]

Situs solitus is the normal positioning of the body organs. If there is mirroring of alignment then this condition is regarded as situs inversus. Around 15% of patients with Situs inversus has compounding abdominal surgical conditions, duodenal atresia being rare entity. The commonest associations are polysplenia or jejunal atresia.

CASE REPORT

A 7 day old male baby born at full term through spontaneous vertex delivery weighing 3 kg was brought in to emergency room with complaints of bilious vomiting and no passage of meconium since birth. On physical examination abdomen was soft, non-distended and non-tender. NG in place with bilious aspirates, DRE revealed empty rectum. Workup carried out with X-ray and baselines investigations. X-ray showed reverse gastric shadow with triple bubble sign on the right side, heart retaining its left sided position on X-ray (Figure 1).

Blood parameters were normal so case was proceeded for surgery. Per-op findings revealed a right sided stomach covered on its posterior aspect by spleen (Figure 2). On left side liver was found, no polysplenia appreciated. Duodenojejunal junction wasn't fixed and ICJ on left. Mesentry was wide. There was continuity of lumen yet hold up was found in last part of duodenum depicting a windsock deformity (Figure 3). Opened from anti mesenteric aspect a redundant diaphragm discovered with a pin point central aperture. Resection of web done keeping in view the proximity of ampulla of Vater and formal duodenoduodenostomy by Kimura technique carried out.

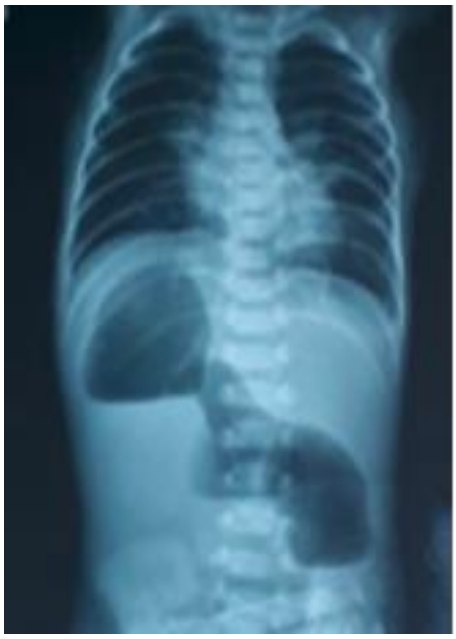

Figure 1: Reverse triple bubble.

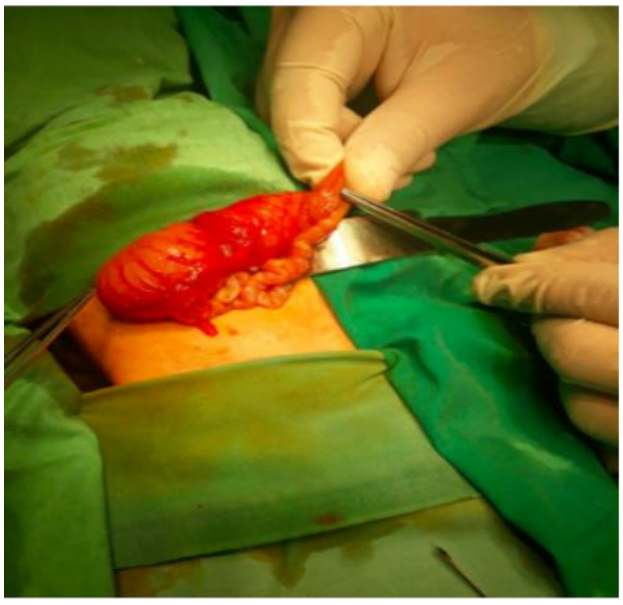

Figure 2: Right sided stomach.

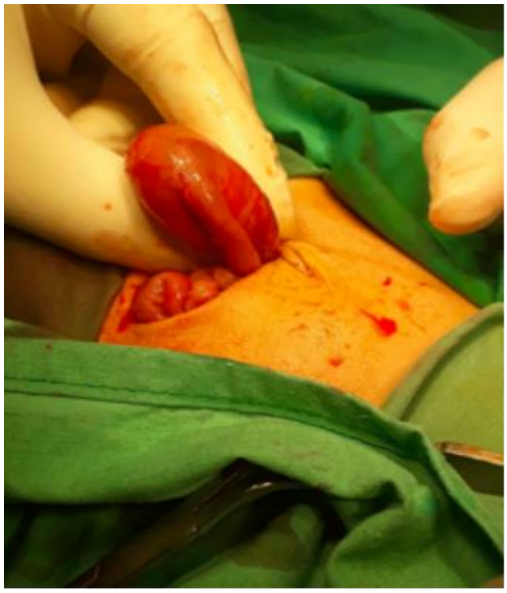

Figure 3: Duodenal obstruction.

Patient recovered smoothly from anesthesia and maintained saturation throughout. Although no ECHO available to firmly comment on cardiac status. Patient had uneventful recovery and was allowed nasogastric feed on 5th day after he passed stool on 4th day. Feed was tolerated well so orally introduced on 6th day. Due to unremarkable recovery patient was discharged on 8th day in excellent condition.

DISCUSSION

Situs inversus has a fascinating radiographic demonstration with a reverse gastric bubble, here it was coupled with reverse triple bubble sign making it even more interesting. Situs inversus has diverse presentations ranging from situs inversus totalis with major shifts of thoracoabdominal organs to opposite end, situs inversus ambiguous where midline position is retained by majority of organs or situs inversus with levocardia where heart instead of being dextrocardic maintained its usual domain in left hemithorax.

Mirroring of all major organs termed as situs inversus totalis had been the usual form of presentation of this condition ranging from 1 in 8000-25000.[4] Hetrotaxy syndrome/situs inversus ambiguus was also very rare but the most rarest form of presentation remained situs inversus with levocardia/situs inversus incompletus with incidence of as low as 20,00,000 of general population. Although situs inversus had been an incidental finding in generality being picked up only when patient had been probed for some other health issue, the common associations remain cardiac issues and polysplenia. Congenital cardiac issues were 5-10% in cases of situs inversus totalis predominant being transposition of great vessels and CHD were in up to 95% in patients with situs inversus with levocardia. The exact etiology remained unclear but certain theories alluded ciliary dyskinesia as

an instigation for malalignment during embryologic advent.[5]

Duodenal atresia evolved when the solid cord state doesn't undergo recanalization in 12th week of embryologic development, result being either a web or absolute disruption. Another cause of duodenal obstruction was annular pancreas where the circumferential encompassing occluded lumen.

The two conditions co-existed in rarity and when they did most commonly association had been found between annular pancreas or a diaphragm with central aperture.[6,7] The only plausible explanation was that the rotational anomaly led to vascular accident resulting in small gut atresias.

So far only limited cases have been reported of this association with prominent literature being given by Lee et al, Lopez et al and Nawaz et al.[3,5,7]

The recommended mode of correction would be a formal duodenoduodenostomy with the excision of the web giving meticulous care to ampulla of Vater as PBMJ with annular pancreas or windsock deformity may predispose to pancreatic duct injury.

It was advisable to conduct a thorough workup before proceeding if we are suspecting a situs inversus to rule out cardiac issues in context of anesthesia optimization also to delineate anatomical plan before surgery.[8]

CONCLUSION

Despite the complexity of the condition the prognosis is redeeming if there is no underlying cardiac issue. Consideration should be given to investigate thoroughly pre op with radiographs, ultrasound and contrast studies to know better about coexisting conditions.

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