A Neonate With Duodenal Duplication And Omphalocele With Accessory Ectopic Liver Tissue

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ABSTRACT

A neonate presenting with more than one congenital anomalies is not uncommon however two rare anomalies presenting together is unusual. We report a case of duodenal duplication along with omphalocele minor containing accessory ectopic liver tissue. Patient under went partial excision of the duplication cyst with stripping of mucosa from residual part, excision of ectopic liver tissue and repair of omphalocele. Postoperative recovery was uneventful.

Key words *Duodenal duplication, Ectopic liver tissue, Omphalocele, Congenital anomalies.*

INTRODUCTION:

Duplication cyst is a rare congenital condition that results during embryonic period of alimentary tract development.[1,2] Most of the duodenal duplication cysts are located in the second or third part of duodenum. They share muscle layers with the main part of the gut for variable extent. Congenital liver anomalies are uncommon. Accessory hepatic lobes are either in continuity with the liver or found in an ectopic location.[3] We report a neonate who had combination of different anomalies.

CASE REPORT:

A 15-day old female baby was brought for the evaluation of antenatally diagnosed intra-abdominal cyst. Birth history was unremarkable. She was tolerating feed, and passed meconium and urine normally. Abdominal examination showed a small membrane covered defect at umbilicus. Abdomen was soft and no mass palpable. Biochemical profile was reported as within normal range. Ultrasound abdomen showed a well-defined cyst of 50 mm x 41 mm size in right side of abdomen adjacent to the right kidney and liver. CT scan abdomen showed a large homogenous cyst in right side of abdomen between the liver and right kidney (Fig. I). Based upon the finding our differential diagnosis was mesenteric cyst or choledochal cyst.

Surgical exploration was planned on elective OR list. On removal of umbilical stump and shriveled membrane, a vascular, globular, pedunculated tissue noted which continued into the peritoneal cavity. On further exploration this structure which had resemblance with liver tissue, was found attached to the right lobe of liver (Fig II). It was easily excised. Another cystic structure (C) identified along the second part of the duodenum (D). Aspiration of cyst revealed clear fluid. Cyst was found attached to the 2nd part of duodenum and sharing common wall. Cyst was opened which had a well defined mucosal lining (Fig. III). It was partially excised because of difficulty in separating it from duodenal wall. Mucosectomy was done from the left over cyst wall. Umbilical defect was also repaired. Baby remained well postoperatively. Feeding was allowed on 4th postoperative day and discharged on 7th day. Biopsy report showed the duodenal duplication and liver tissue with biliary channels in accessory tissue.

DISCUSSION:

Duodenum is one of the rare locations of gastrointestinal tract duplications. It constitutes 5% - 7% of all gastrointestinal duplications.[1,2,4] A meta-analysis of reported cases between 1998 and 2009 found 47 cases of duodenal duplication cysts.[5] These are usually cystic and non-communicating. The cyst can be diagnosed on antenatal ultrasound as noted in our patient. Number of clinical manifestations are reported in literature which include abdominal pain, nausea and vomiting, however our patient had no symptoms.[6,7]

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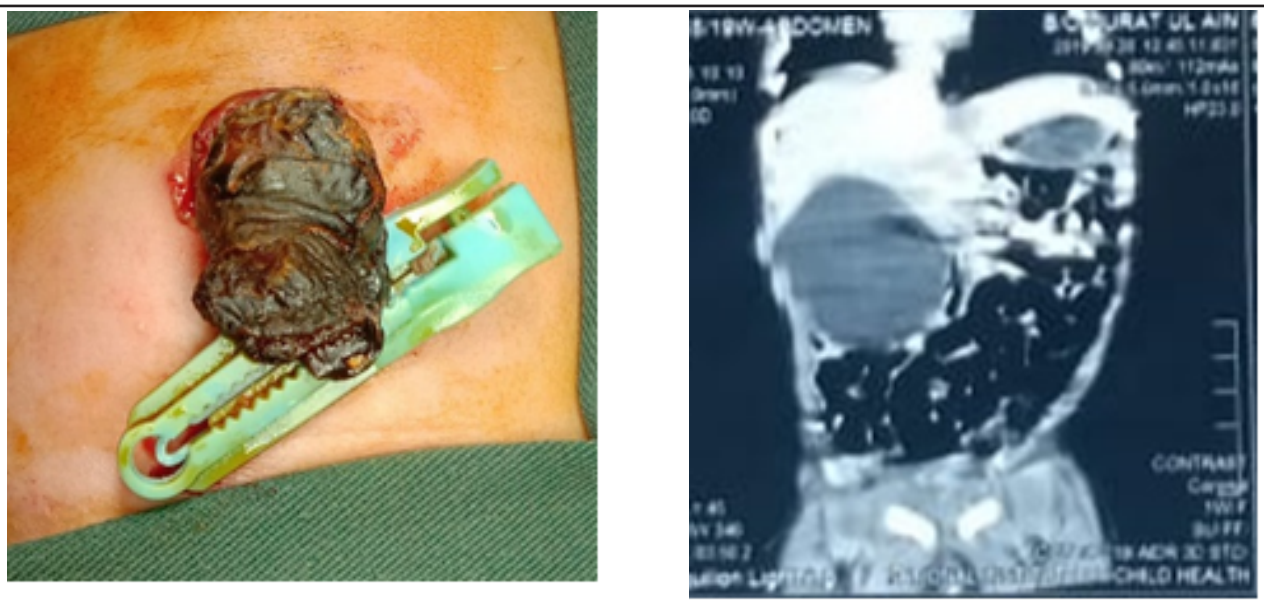

Fig: I a) Swelling in umbilical region with impression of omphalocele minor. b) CT scan showing large homogenous cyst in right upper abdomen.

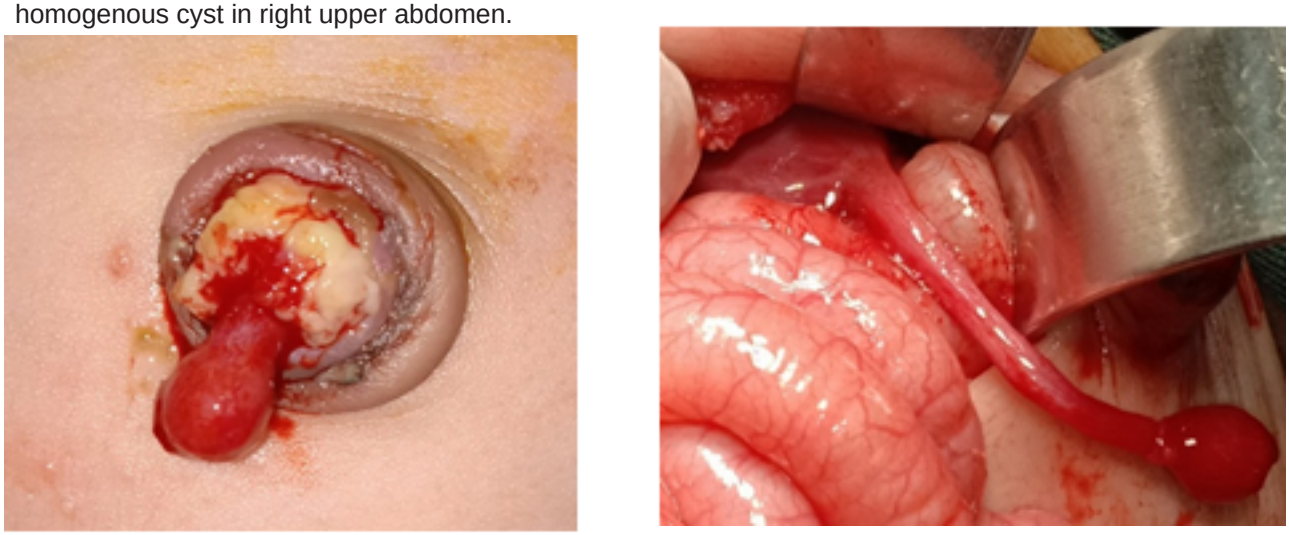

Fig II: a) Polypoid tissue at umbilicus, b) accessory pedunculated tissue attached with liver passing through umbilical stump.

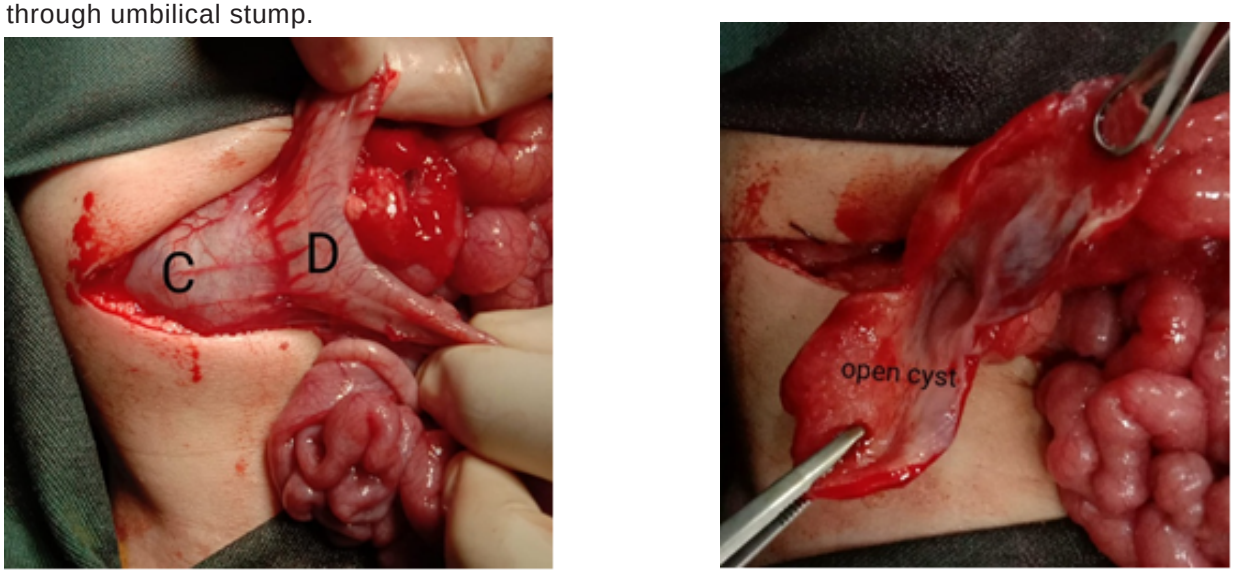

Fig III: a) Duodenum (D) and cyst (C) sharing common wall, b) Cyst opened.

Ultrasound and CT scan are useful modalities in making a diagnosis of duodenal duplication cyst.[6] Treatment of duodenal duplication cyst is usually surgical, and total excision, if possible is the procedure of choice. However, in difficult cases and critical location, partial removal and mucosctomy are effective.[7] Endoscopic treatment in which duodenocystostomy done, is also a favored and less invasive procedure. Laparoscopic exploration is also reported in which duodenotomy followed by duodenocystostomy or fenestration of the cyst is done.[2,8] In our case partial excision of cyst and stripping of mucosa was done and duodenum remained intact. Histologically, duodenal duplication cysts have all layers of intestinal wall. In few cases ectopic gastric and pancreatic mucosa may be present.[2] In our case histopathology does not show any ectopic mucosa.[2]

Accessory hepatic lobe develops secondary to defects of liver organogenesis. These may be found in falciform and gastrohepatic ligament. These may be associated with defects of anterior abdominal wall. Same was found in our patient.[9] Before surgery we presumed a communication of polypoidal mass at umbilicus with the intestine, however it was the liver tissue with stalk. This was an unusual finding. Our patient had smooth postoperative stay and discharged home in stable condition and remained well in follow up.

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