Situs Inversus Totalis with Malrotation : A Known Association in an Unknown Territory

Kandiah V[1], Aliaa M[2], Quah SY[3], Najua R[4], Vellusamy MA[5]

Paediatrics Surgery
Department of Surgery
Hospital Sultanah Aminah
Johor Bahru
Malaysia

Abstract

Situs inversus totalis with rotational anomaly is a known association but rare entity. Here we are reporting a case of neonatal SIT + Malrotation with a rare intra-operative finding of Pre-duodenal portal vein with an anterior CBD in a solitary case and our experience in managing this difficulty.

Introduction

Situs inversus totalis with rotational anomaly is a known association but rare entity.
May manifest with an early obstruction and hence followed up with a laparotomy and corrective Ladd's procedure. Rarely, the portal vein & CBD may be located anterior to the duodenum resulting in extra-luminal compression and described aptly as pre-duodenal portal vein (PDPV).

We would like to report a case of neonatal SIT + Malrotation with a rare anomaly of Pre-duodenal portal vein & CBD and our experience in managing this case.

Case

Baby L, 3.2kg, girl was born term and noted to be tachypnoeic after delivery. An X-ray done showed the cardiac shadow lie within the right hemithorax and liver shadow towards the LHC. and she was diagnosed to have situs inversus.
Naso-gastric aspirations were persistently bilious since D6 of life which raised the suspicion of a malrotation. Abdominal Radiographs revealed a right sided gastric bubble with minimal bowel gas seen distally. After a Peadiatric Surgical consult, USG & Upper GI contrast study was done and confirmed the diagnosis of a Malrotation and child was taken over immediately on D7 of life and an emergency laparotomy done.

The intra-operative findings revealed a situs inversus totalis with a malrotation with a loose volvulus. The DJ was located on the left of the vertrebrae; however there was a rare finding of the CBD and Portal vein traversing anterior to the D1 & D2 causing an extraluminal compression to the duodenum (D1-D2). Distal bowels appeared patent therefore proceeded with Ladd's Procedure and it was uneventful.

Suspecting the persistently high bilious aspirates post-operatively was due to the PDPV and CBD causing the extra-luminal compression, we then decided for a re-laparotomy.

At 2nd. operation, only the D1 and D2 (proximal to PDPV) remained dilated. With our suspicion confirmed, we decided for a bypass a gastro-jejunostomy as PDPV & CBD gave little room to perform Duodeno-duodenostomy.
Post-operatively, the child was extubated by Day 2. She was on TPN and we were able to establish feeds on D18 post surgery and discharged home at 1 month of age.

Subsequently on follow up, she was thriving with no complaints from her parents.

Discussion

Heterotaxia with malrotation has been described previously with the possible PDPV. The existence of the PDPV was first decribed by Knight in 1921.

The embryogenesis of this anomaly was described by Gray & Skandalakis. Developmentally, primitive vitelline vein that forms portal vein passes in front of duodenum and pancreas rather than passing inferior and behind the pancreas. In general, PDPV without associated anomalies may be the cause of duodenal obstruction in the first or second portion, though uncommon for obstruction to be due to extrinsic pressure on the bowel. Rather it has been due to short length of the PV or associated anomalies such as duodenal diaphragms, duodenal colic bands, malrotation, or annular pancreas. Extra-luminal compression of the duodenum is not due to the instrinsic pressure of PV or CBD but more likely from the short length of the 2 structures causing a traction towards the porta pulling the D1-D2 cranially and hence the obstruction.

To conclude that the etiology of duodenal obstruction is purely from the presence of the anterior lying PV and CBD is not true. There are incidences of intrinsic webs which warrant exploring intra-luminally.

The aim of treatment is to bypass the obstruction and avoid injury to these vital structures. Ideally, a Duodeno-duodenostomy would have been more physiologic and also less likely to develop dysplasia over time. The available length of the duodenum was not sufficient to perform a Duodeno-duodenostomy inour patient therefore the gastro-jejunostomy. .

The risk of dysplasia and development of malignancy is present and this child will require long term follow up and perhaps surveillance endoscopy when she's an adult.

Conclusion

In cases of SIT, it's important for the paediatricians and paediatric surgeons alike to be aware of such an association especially if there are signs of bowel obstruction.

Although congenital duodenal obstruction is a relative surgical emergency, the patient should not be rushed to the operating room until the baby is stable. During surgery definitive cause for the obstruction must be identified and managed accordingly. In our case we performed the Ladd procedure for the associated malrotation in the 1st surgery. Although this did not solve the obstruction, knowing that the PDPV & CBD is the definite cause of obstruction, we performed a 2nd surgery and bypass with a Gastrojejunostomy. There isn't a fixed guideline in managing this entity although adhering to principles of surgery, we performed a safe bypass. Long term follow up is necessary for this case.

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