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Case Report

Situs Inversus Abdominis in Association with Intestinal Malrotation and Ladd's Bands in a Girl of 4 Years

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Abstract

Situs inversus is a rare condition. But situs inversus in association with malrotation of the gut and Ladd's bands not reveled in new-born period is extremely rare. This report describes a girl of 4 years with persistent bile-stained emesis during two months. Her radiological investigations revealed reverse position of stomach and duodenum whereas the heart located in normal position. Surgical exploration noticed stomach and spleen in the right side and liver in the left side with gallbladder in the epigastric area. Intestinal malrotation with Ladd's band was also noticed. Two levels of obstruction were found. The duodenal obstruction was partial and due to extrinsic compression by Ladd's band. Jejunal obstruction linked to an intramesenteric "8" crossing immediately downstream the Treitz angle and was also partial. Only jejunal derotation and Ladd procedure allowed the girl healed with an eventful postoperative period. Treatment of duodenal or jejunal obstruction with or without situs inversus is the same.

Introduction

Situs inversus is a rare condition. Its frequency is about one on 10,000 of the normal population [1,2]. It can be asymptomatic and found incidentally during laparotomy for a different pathology or at autopsy; but when associated with other anomaly which precociously leads to emergency situation, it is often detected early in the new-born period [3]. The common abdominal anomalies associated to situs inversus abdominus which train to emergency situation in the new-born period are duodenal atresia, annular pancreas, intestinal volvulus due to malrotation or duodenal extrinsic compression related to Ladd's bands across the duodenum [3,4]. But situs inversus in association with malrotation of the gut and Ladd's bands is extremely rare. We report a case of situs inversus abdominus in association with malrotation and Ladd's bands, revealed like a duodenal stenosis in young girl of 4 years.

Case Report

A young girl presented at 4 years for bilious and alimentary emesis with fiver in the beginning. Despite the treatment against malaria and typhoid, and the correction of the temperature, emesis persisted and she lost 3 kg in one month with deshydration and denutrition. Abdominal ultra sonography was no conclusive. She was then referred to pediatric surgery department after two months of medical management. Her birth and development history was normal and was thriving well till the current episode. Clinically the child was sick and dehydrated. The abdomen was soft and not distended with normal bowel sounds. There was no classic double-bubble appearance on abdomen and chest x-rays but the heart was in normal position. Abdomen ultrasound showed distended stomach and duodenum, and suggested upper gastrointestinal. Upper gastrointestinal abdominal x-rays revealed distended stomach and duodenum with duodenal stenosis. The stomach was in the right upper side and the duodenum was in the left upper side whereas the heart was in normal position (figure 1). The Echocardiography was normal.

After correction of dehydration troubles, the patient then went under laparotomy. There were stomach and spleen on the right side, and liver on the left one. The gallbladder was located in the epigastric area and the duodenal loop was on the left side. Jejunum had in tramesenteric "8" crossing immediately downstream Treitz angle. There was also intestinal malrotation with icomplete common mesentery at 180 degrees and duodenal stenosis by extrinsic compression by Ladd's bands. Two levels of obstruction were found. The duodenal obstruction was partial and due to extrinsic compression by Ladd's band. Jejunal obstruction linked to an intramesenteric "8" crossing immediately downstream the Treitz angle and was also partial. Portal vein was retro duodenal. The figure 2 shows the schema of the guts position during laparotomy. In summary the patient had situs inversus abdominis associated to intestinal malrotation with Ladd's band and jejunal intramesenteric

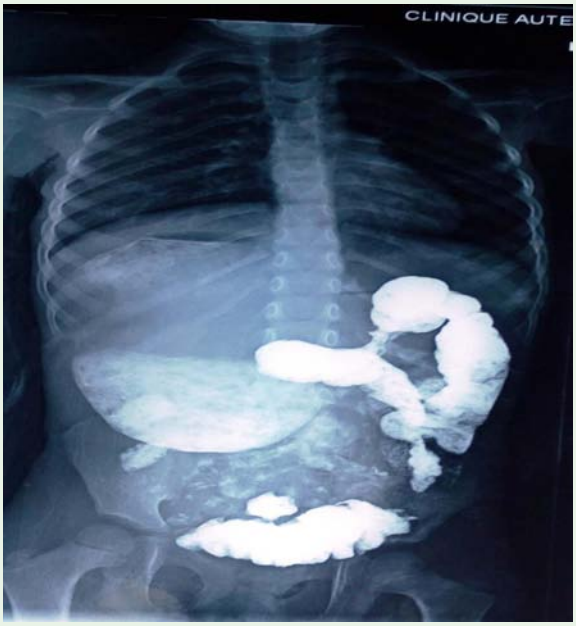

Figure 1: Upper gastrointestinal x-rays of the girl: notice distended stomach at the right upper and distended duodenum at the left upper side and the heart at normal position.

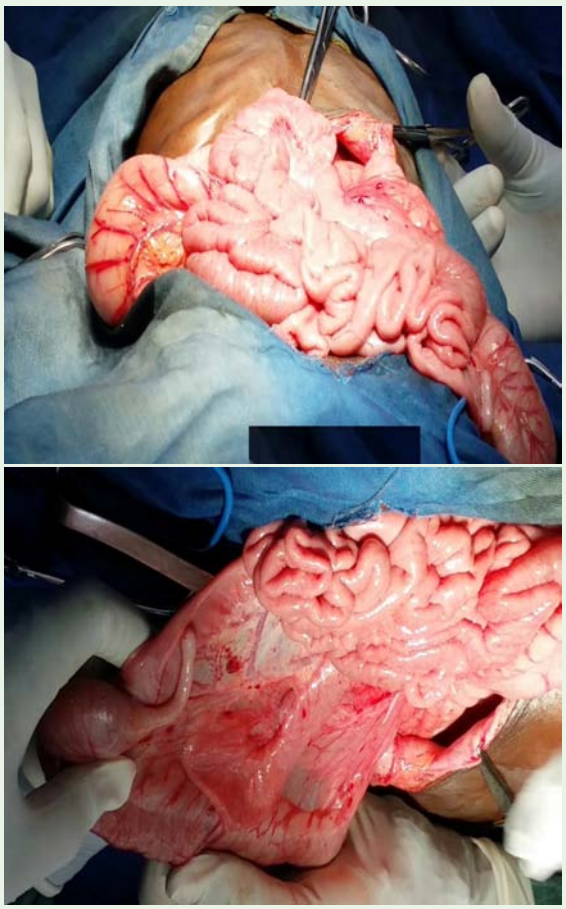

Figure 2: The schema of the guts position during laparotomy.

volvulus. Release of Ladd's band following by jejunal derotation, and replacement of intestines in complete common mesentery position (Ladd procedure) were performed. There was not any anastomosis because, after release of Ladd's band and jejunal derotation guts were sufficiently permeable. Post operative period was eventful after a follow-up of 7 months.

Discussion

Situs inversus abdominus is a rare malformation. It is characterized by inversion of the abdominal organs but with a normally located left-side heart. Its frequency ranging from one in 4,000 to one in 20,000 of the normal population [1,2]. It's commonly in association with serious cardiac and splenic malformations [5-7]. However, the association of abdominal situs inversus with duodenal obstruction is extremely rare [3,4,8,9], moreover its association with malrotation [10,11]. Situs inversus is totally asymptomatic in common condition and incidentally detected by laparotomy or at autopsy. But when associated to other malformations as in most of times, leading to emergency situations, it is revealed by the symptomatology linked to that malformation. Then its association with duodenal obstruction is detected in the new-born period by neonatal intestinal obstruction signs [4,11,12]. Duodenal obstructions has several causes; it may be duodenal complete atresia, duodenal stenosis, pancreas annular, preduodenal portal vein, duodenal extrinsic compression by Ladd's bands and volvulus due to intestinal malrotation [3,4,8] Among them, pancreas annular seems to be the most common, followed by duodenal web [12-15]. These associated malformations can be singly or jointly reported [8] and malrotation can also be reported in various degrees from classic malrotation predisposing to volvulus to nonrotation [16,17]. Four groups of intestinal rotation were determined: (1) normal rotation, (2) incomplete rotation or nonrotation, (3) reversed rotation, and (4) reversed incomplete rotation or nonrotation. The most common type found in pediatric patients is incomplete rotation predisposing to midgut volvulus, requiring emergent operative intervention [18,19]. In our case, we had an incomplete rotation stopped at 180 degrees (common incomplete mesentery at 180 degrees) but with chronic jejunal volvulus. In fact, intestinal malrotation can present as either an acute or chronic process. It can present in different clinical aspects: acute midgut volvulus, chronic midgut volvulus, acute duodenal obstruction, chronic duodenal obstruction or internal herniation [20].

In which the age of presentation is concerned, traditional teaching suggests that as many as 40% of patients with malrotation present within the first week of life, 50% in the first month, and 75% in the first year. However, more recent series have shown that malrotation is increasingly identified in adults. A series of 170 patients with intestinal malrotation diagnosed at a single institution between 1992 and 2009 found that 31% were infants, 21% were aged 1-18 years, and the remaining 48% were adults [21]. A second series found that 42% of patients with malrotation were adults [22]. Partial volvulus seems to be more frequent than the tight and acute one. Then for those who continue to think that majority of malrotation are revealed before the first birthday it is not evident to think about malrotation and its complications in a child more than one year. In chronic midgut volvulus, the most common symptoms are recurrent abdominal pain and malabsorption syndrome [23]. Other clinical features include recurrent bouts of diarrhea alternating with constipation, intolerance of solid food, and gastro oesophageal reflux [24]. Our girl of 4 years was treated for emesis and abdominal pain during two months before

the upper gastrointestinal; and it was by exploration laparotomy the diagnosis was finally done. Ultrasonography did not find the volvulus perhaps because of its intermittence. The heart is the most common affected intra-thoracic organ in situs anomaly, and in most instances cardiac symptoms are usually the first ones that lead to the detection of this anomaly. It is important to establish the anatomy of the heart with an echocardiogram as part of the pre-operative assessment of the patient before undergoing any operative procedure so as to detect the presence of an associated cardiac anomaly, which may influence the prognosis of the patient [9].

Situs inversus is really not a disease but a simple arrangement anomaly. Associated anomalies which accompanied the situs inversus are the revelation way of the malposition; and constitute mortality and morbidity factor of these patients. Then there is no treatment of situs inversus but treatment for associated anomalies. The treatment of intestinal volvulus secondary to a malrotation with Ladd's bands with or without situs inversus is the same: derotation of the gut and Ladd procedure. In case of acute volvulus with intestinal necrosis an intestinal resection with end-to-end intestinal anastomosis will be performed.

Data from recent series reveal that mortality rates in adults and children operated on for intestinal malrotation range from zero to 14%. Higher mortality are seen in cases with acute onset of midgut volvulus, delayed diagnosis, or the presence of intestinal necrosis [18,25-29]. Children with other associated anomalies also have higher overall mortality rates. A case report of 25 years experience demonstrated congenital cardiovascular disease in 27,1% of patients with intestinal malrotation; those patients had a morbidity rate of 61,1% after intestinal malrotation surgery [30]. None of higher risk factor was seen in our case and especially the echocardiography was normal.

Conclusion

Abdominal situs inversus is simply an anomaly of organs arrangement. It can be discovered either casually during an exploration for another disease or before associated anomalies manifestations. The treatment of associated diseases is the same as without it, especially for volvulus due to malrotation with duodenal obstruction by Ladd's bands. Gut derotation with Ladd procedure gives good results.

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