Situs ambiguous with duodenal atresia in a neonate: A case report with literature review

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ABSTRACT
Background: Situs ambiguous is a rare congenital anomaly characterized by an unusual and irregular distribution of the major visceral organs and vessels within the chest and abdomen. In this condition, the organs are arranged in a way that cannot be classified as either situs inversus or situs solitus. We report this case due to the rarity of this anomaly and its uncommon association with duodenal atresia in a neonate.

Case Presentation: A full-term newborn female presented with repeated bilious vomiting and failure to pass meconium for five days. An erect abdominal X-ray revealed a double-bubble sign in the left hemiabdomen, and a CT scan confirmed an abnormal relationship between the liver and stomach. Operative exploration confirmed the diagnosis of situs ambiguous with duodenal atresia, which was treated with a diamond duodenoduodenostomy.

Conclusion: Situs ambiguous is a rare variant of situs anomaly with few reported cases worldwide. It has two subtypes: left and right isomerism, based on the anatomy of the spleen. Clinically, it can present with features of intestinal obstruction when associated with duodenal atresia.

INTRODUCTION
Situs ambiguous is a rare congenital abnormality characterized by a bizarre and irregular distribution of the major visceral organs and vessels within the chest and abdomen. In this condition, the organs are arranged in a way that cannot be classified as either situs inversus or situs solitus[1]. It can be subdivided into two types based on the anatomy of the spleen: right isomerism (situs ambiguous with asplenia) and left isomerism (situs ambiguous with polysplenia)[2]. The incidence of situs ambiguous is approximately 1 per 25,000 live births, and about 25% of cases are associated with immotile cilia syndrome (including Kartagener syndrome) [3]. We report this case due to the rarity of this anomaly and its uncommon association with duodenal atresia in a neonate.

CASE REPORT
A full-term newborn female delivered by cesarean section presented with repeated bilious vomiting and failure to pass meconium for five days before being brought to the pediatric surgical center. The abdomen was non-distended except for localized right lumbar distention. Antenatal ultrasonography at 36 weeks of gestation revealed polyhydramnios. An erect abdominal X-ray showed a classic double-bubble sign. Still, in the reverse direction (right side) (Fig. 1). The echocardiographic report indicated levocardia with patent ductus arteriosus but no other major cardiac defects. A CT scan of the abdomen and chest confirmed the diagnosis of situs ambiguous with bilateral bilobed lungs.

After preparation, an exploratory laparotomy through a supraumbilical transverse incision was performed. A redundant stomach and the first part of the duodenum were identified, secondary to a duodenal web located in the second part. The stomach was on the right side of the abdomen, while the duodenum was anteriorly in the left hemiabdomen. Nonrotation was evident, with the small bowel found on the right side of the abdomen, the colon on the left, and the cecum located in the left hypochondrium. The liver was found in the left hemiabdomen, while the spleen was not visualized; four spleniculi were located on the right side near the greater curvature of the stomach (Fig. 2a).
A limited web resection with a diamond duodeno-duodenostomy was performed after confirming distal bowel patency by injecting saline solution through a nasogastric tube (Fig. 2b). A trans-anastomotic stent was left in place for feeding. An appendectomy was also performed due to the abnormal location of the appendix in the left hypochondrium.

Figure 1: Erect abdominal X-ray revealing a double-bubbled sign in the right hemiabdomen, with a liver shadow on the left side.

Figure 2: (a) Type 1 duodenal atresia (web) marked by forceps causing a dilated stomach (green arrow) located on the right hemiabdomen, with a dilated first part of the duodenum (black arrow). The appendix was in the left hypochondrium (blue arrow). One of the spleniculi that located close to greater curvature (yellow arrow). (b) Diamond duodeno-duodenostomy performed.

The patient had a smooth postoperative course, starting formula feeding through the nasogastric tube on day three and oral feeding on day five postoperatively, and was discharged home stable the following day. Postoperative follow-up visits on days 10, 30, and 60 showed normal weight gain and regular bowel movements.

DISCUSSION

The term “situs” refers to the location of the abdominal viscera and heart on one side of the midline inside the abdomen and chest [4]. Situs solitus is the normal positioning of organs. Situs inversus describes the reverse location of all organs (mirror image) [2]. Situs ambiguous is the rarest variant and represents a bizarre and irregular distribution of the major visceral organs and vessels within the chest and abdomen [1]. This variant has two subtypes: right isomerism (normal cardiac situs, situs ambiguous, asplenia, left-sided inferior vena cava, and bilateral trilobed lung) and left isomerism (situs ambiguous, normal cardiac situs, polysplenia, interruption of the inferior vena cava by the azygos or hemiazygos vein, and bilateral bilobed lung) [2].

A defect in lateralization around day 28 of gestation is thought to be the underlying cause of situs anomalies. This lateralization defect leads to deviation from the normal location of the viscera [5]. Vetrini et al. and Narasimhan et al. discovered eight genes and loci associated with situs ambiguous: NODAL, ZIC3, HTX3, CFC1, PKD1L1, ACVR2B, MMP21, and CCDC11. The mode of inheritance is heterogeneous: autosomal dominant for NODAL and CFC1, autosomal recessive for CCDC11, PKD1L1, and MMP21, and X-linked recessive for ZIC3 [6,7].

Hepatic and biliary tract malposition are also reported with polysplenia and left isomerism. Fulcher et al. discovered a mirror-image branching of the biliary tracts on intraoperative cholangiography in a patient with left isomerism [8]. In our case, the liver was malpositioned, but intraoperative cholangiography was not performed to prove or exclude a biliary tract anomaly. Another common association with left isomerism is congenital heart disease, with an incidence between 50% and 90%, commonly including atrial septal defect (ASD), common atrioventricular canal, and partial anomalous pulmonary venous return [5]. Interestingly, our case had a normal echocardiographic finding except for patent ductus arteriosus.

Intestinal malrotation, including nonrotation, is also associated with situs ambiguous, whether right or left isomerism, in up to 70% of cases [9,10]. Congenital duodenal obstruction secondary to a duodenal web can be associated with intestinal malrotation and is a presenting feature in situs ambiguous, occurring in 1:10,000 to 1:40,000 births [10]. A similar finding of nonrotation in association with a duodenal web was observed in our case. Clinically, the patient presents with biliary vomiting and constipation, like our case who failed to pass meconium for five days.
Tomas Mujo [2] reported a similar case of a female newborn with situs ambiguous, intestinal nonrotation, and an obstructing duodenal web, who presented with repeated bilious vomiting and absence of stooling for two days. X-ray findings also revealed a double-bubble sign in the right hemiabdomen with an inverse relationship between the stomach and liver. Unlike our case, Tomas’s case had a spleen in the right upper quadrant without spleniculi, confirmed by abdominal ultrasound.

Francesca Destro et al. [11] in 2017 reported two cases of situs ambiguous associated with a choledochal cyst in 8 and 18-month-old boys, both diagnosed antenatally. The first case was asymptomatic, while the second presented at 18 months with failure to thrive, abdominal distention, and hepatomegaly. In both cases, ultrasonography and MRI confirmed the diagnosis of situs ambiguous with dilated intrahepatic bile ducts associated with a type I choledochal cyst.

In the same year, Shagun Aggarwal [12] reported a case of situs ambiguous presenting as fetal hydrops due to an associated complex cardiac defect. The operative procedure for Tomas’s case was excision of the web with primary anastomosis and appendectomy, while in Francesca’s cases, intestinal continuity was achieved by Roux-en-Y fashion after excision of the choledochal cyst. In our case, we performed a diamond duodeno-duodenostomy with a limited web excision to avoid injury to a possible anomalous ampulla.

Regarding the outcome of the reviewed cases, Tomas’s case did well postoperatively but without a recorded follow-up time. The two cases reported by Francesca Destro also did well postoperatively with follow-up periods of four years and five months, respectively. Although the association of duodenal atresia with situs ambiguous is rare, it is more frequently

Table 1: Review of the reported cases of duodenal atresia with situs inversus

<table>
<thead>
<tr>
<th>Author &amp; year of publication</th>
<th>Age</th>
<th>Gender</th>
<th>Clinical presentation</th>
<th>Situs variant</th>
<th>Type of duodenal pathology</th>
<th>Operative procedure</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Akhtar Nawaz[13] 2005</td>
<td>4 days</td>
<td>female</td>
<td>Bile-stained vomiting, failure to pass meconium</td>
<td>Situs inversus abdominus</td>
<td>Duodenal diaphragm with a central opening</td>
<td>Side to side duodeno-duodenostomy+ appendectomy</td>
<td>Well with 3 weeks of follow-up</td>
</tr>
<tr>
<td>Craig Brown[14] 2009</td>
<td>1 day</td>
<td>female</td>
<td>Bilious vomiting</td>
<td>Situs inversus abdominus</td>
<td>Obstructing duodenal web</td>
<td>Web resection with primary anastomosis</td>
<td>Well</td>
</tr>
<tr>
<td>Raghu Shankar[15] 2012</td>
<td>2 days</td>
<td>male</td>
<td>Bilious aspirate, failure to pass meconium</td>
<td>Situs inversus abdominus</td>
<td>Obstructing duodenal web</td>
<td>Side to side duodeno-duodenostomy</td>
<td>Well</td>
</tr>
<tr>
<td>Shuai Qiang[16] 2020</td>
<td>1 day</td>
<td>male</td>
<td>Bilious vomiting</td>
<td>Situs inversus abdominus</td>
<td>Duodenal stenosis with a Ladd band obstruction</td>
<td>Diamond duodeno-duodenostomy+ Ladd’s procedure</td>
<td>Well with 4 years of follow-up</td>
</tr>
<tr>
<td>Murtadha A Alshaikh[17] 2021</td>
<td>1 day</td>
<td>male</td>
<td>Bilious vomiting</td>
<td>Situs inversus totalis</td>
<td>Type III duodenal atresia</td>
<td>Side-to-side duodeno-duodenostomy with Ladd’s procedure</td>
<td>Well with 7 months of follow-up</td>
</tr>
<tr>
<td>Fatima Al Zahra[18] 2022</td>
<td>7 days</td>
<td>male</td>
<td>Bilious vomiting, failure to pass meconium</td>
<td>Situs inversus incompletes</td>
<td>Duodenal web with tiny central aperture</td>
<td>Web resection with Kimura duodeno-duodenostomy</td>
<td>Well</td>
</tr>
<tr>
<td>Gogan M[19] 2023</td>
<td>6 days</td>
<td>male</td>
<td>Bilious vomiting, failure to pass meconium</td>
<td>Situs inversus totalis</td>
<td>Annular pancreas obstructing the 2nd and 3rd part of duodenum</td>
<td>Diamond duodeno-jejunostomy</td>
<td>Well with 6 months follow-up</td>
</tr>
</tbody>
</table>
CONCLUSION

To conclude, situs ambiguity is a rare form of situs anomaly with few reported cases worldwide. It has two subtypes: left and right isomerism, based on the anomaly with few reported cases worldwide. It has

REFERENCES


