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Case Report

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

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KEYWORDS

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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.

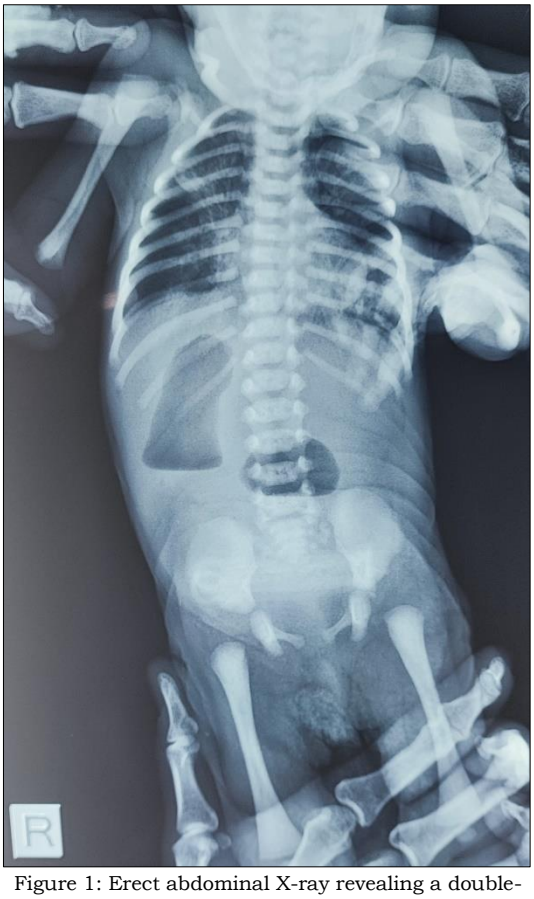

Figure 1: Erect abdominal X-ray revealing a double-bubbled sign in the right hemiabdomen, with a liver shadow on the left side.

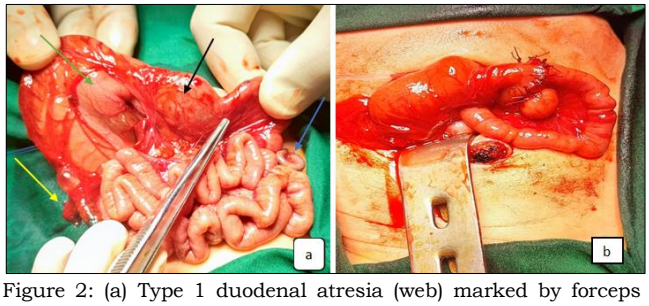

Figure 2: (a) Type 1 duodenal atresia (web) marked by forceps causing a dilated stomach (green arrow) located on the right hemiabdomen, with a dilated first part of the duodenum (black arrow). The appendix was in the left hypochondrium (blue arrow). One of the spleniculi that located close to greater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

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

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

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

encountered with other situs anomalies. The following table illustrates the reported cases of duodenal atresia with situs inversus (Table 1).

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 anatomy of the spleen. Clinically, it can present with features of intestinal obstruction when associated with duodenal atresia.

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REFERENCES

1. Lambert TE, Kuller J, Small M, Rhee E, Barker P. Abnormalities of Fetal Situs: An Overview and Literature Review. Obstet Gynecol Surv. [Internet] 2016 [cited 2024 Jan 21];71(1):33–8. Available from: https://journals.lww.com/obgynsurvey/fulltext/2016/01000/abnormalities_of_fetal_situs__an_overview_and.18.aspx
2. Mujo T, Finnegan T, Joshi J, Wilcoxen KA, Reed JC. Situs ambiguous, levocardia, right sided stomach, obstructing duodenal web, and intestinal nonrotation: A case report. J Radiol Case Rep. 2015;9(2):16–23.
3. Worsley C, Weerakkody Y. Heterotaxy syndrome. Radiopaedia.org [Internet] 2009 [cited 2024 Jan 19]; Available from: http://radiopaedia.org/articles/7420
4. Cullis PS, Siminas S, Losty PD. Is Screening of Intestinal Foregut Anatomy in Heterotaxy Patients Really Necessary?: A Systematic Review in Search of the Evidence. Ann Surg. [Internet] 2016 [cited 2024 Jan 28];264(6):1156–61. Available from: https://pubmed.ncbi.nlm.nih.gov/26704743/
5. Applegate KE, Goske MJ, Pierce G, Murphy D. Situs revisited: imaging of the heterotaxy syndrome. Radiographics. [Internet] 1999 [cited 2024 Jan 28];19(4):837–52. Available from: https://pubmed.ncbi.nlm.nih.gov/10464794/
6. Narasimhan V, Hjeij R, Vij S, Loges NT, Wallmeier J, Koerner-Rettberg C, et al. Mutations in CCDC11, which encodes a coiled-coil containing ciliary protein, causes situs inversus due to dysmotility of monocilia in the left-right organizer. Hum Mutat. [Internet] 2015 [cited 2024 Jan 28];36(3):307–18. Available from: https://europepmc.org/article/med/25504577
7. Vetrini F, D'Alessandro LCA, Akdemir ZC, Braxton A, Azamian MS, Eldomery MK, et al. Bi-allelic Mutations in PKD1L1 Are Associated with Laterality Defects in Humans. Am J Hum Genet. [Internet] 2016 [cited 2024 Jan 28];99(4):886–93. Available from: https://pubmed.ncbi.nlm.nih.gov/27616478/
8. Fulcher AS, Turner MA. Abdominal Manifestations of Situs Anomalies in Adults. https://doi.org/101148/rg226025016 [Internet] 2002 [cited 2024 Feb 2];22(6):1439–56. Available from: https://pubs.rsna.org/doi/10.1148/rg.226025016
9. Jo DS, Jung SS, Joo CU. A case of unusual visceral heterotaxy syndrome with isolated levocardia. Korean Circ J. [Internet] 2013 [cited 2024 Feb 2];43(10):705–9. Available from: https://pubmed.ncbi.nlm.nih.gov/24255657/
10. Eksarko P, Nazir S, Kessler E, Leblanc P, Zeidman M, Asarian AP, et al. Duodenal web associated with malrotation and review of literature. J Surg Case Rep. 2013;2013(12):7–9.
11. Destro F, Maestri L, Napolitano M, Meroni M, Pansini A, Riccipetitoni G. Choledochal cyst, polysplenia and situs ambiguous: A rare and new association. J Pediatr Surg Case Rep. [Internet] 2017;24:35–9. Available from: http://dx.doi.org/10.1016/j.epsc.2017.06.010
12. Aggarwal S. A Case of Situs Ambiguous and Complex Cardiac Defect Presenting as Fetal Hydrops. Indian J Cardiovasc Dis Women. WINCARS 2017;02(02):060–3.
13. Nawaz A, Matta H, Hamchou M, Jacobez A, Trad O, Al Salem AH. Situs inversus abdominus in association with congenital duodenal obstruction: A report of two cases and review of the literature. Pediatr Surg Int. 2005;21(7):589–92.
14. Brown C, Numanoglu A, Rode H, Sidler D. SAJS Situs inversus abdominalis and duodenal atresia. South African J Surg. 2009;47(4):127–30.
15. Shankar R, Rao SP, Shetty KB. Duodenal atresia in association with situs inversus abdominus. J Indian Assoc Pediatr Surg. 2012;17(2):71–2.
16. Qiang S, Fan M, Cui Q, Li Z, Zhou Y, Li Q, et al. Management of duodenal atresia associated with situs inversus abdominus: A case report. Med. (United States) 2020;99(31):E21439.
17. Alshaikh MA, Al Ghadeer HA, Alabad H, Almohsin M, Al Ali RA. Situs Inversus Totalis in Association With Duodenal Atresia. Cureus. 2021;13(9).
18. Zahra F Al, Khan NA, Akhtar N. An unusual association: duodenal atresia and situs inversus incompletes-a case report and discussion. Int J Contemp Pediatr. 2022;9(5):515.
19. Gogan MVLSB, Lafia KT, Amoussou CAM, Metchihoungbé CS, Dossou MGIS, Fiogbé MA. Duodenal atresia, annular pancreas, and situs inversus totalis- rare association in a newborn: A case report. J Neonatal Surg. 2023;12(2023):5–7.