

Case Report

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Duodenal atresia, annular pancreas, and situs inversus totalis- rare association in a newborn: A case report

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KEYWORDS

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Annular pancreas,
Situs inversus,
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Neonate

ABSTRACT

Background: Simultaneous occurrence of duodenal atresia, annular pancreas, and situs inversus totalis is an exceedingly rare clinical condition. This may pose management-related challenges to the treating surgeon.

Case Presentation: We report a case with a preoperative diagnosis of duodenal atresia and situs inversus. Intraoperatively, an annular pancreas encircling the duodenum was found. Instead of duodenoduodenostomy, a duodenojejunal anastomosis was performed. The postoperative course remained uneventful. The patient is doing fine on a 6-month follow-up.

Conclusion: The simultaneous presence of multiple anomalies in this patient prompted us to utilize an alternative approach to relieve the duodenal obstruction. This report highlights the importance of tailoring surgical decisions based on the individualized needs of the patient.

INTRODUCTION

Duodenal atresia or duodenal stenosis is frequently associated with an annular pancreas or intestinal malrotation. However, the simultaneous occurrence of duodenal atresia, annular pancreas, and complete situs inversus is rarely reported in the literature. [1] Surgical management in this setting remains a challenge for the surgeon and often alternate treatment options have to be incorporated in the treatment plan. [1-3] This case is being reported to highlight the concurrence of multiple rare anomalies in a single patient with duodenal atresia that required duodenojejunostomy in place of a duodenoduodenostomy.

CASE REPORT

A 6-day-old male neonate weighing 1835 g, born via a vaginal delivery on the 36th gestational week to a 30-year-old mother, was referred from a periphery health facility for management of bilious vomiting. The baby was not exposed to known teratogens. The last antenatal scan picked polyhydramnios; otherwise, the antenatal period remained uneventful. There was no family history of malformations, and the other two siblings were fine.

On physical examination, the baby was in good clinical condition with a soft and non-distended

abdomen. Rectal stimulation revealed no meconium in it. X-ray of the chest and abdomen noted the tip of the heart on the right side, a distended stomach with an air pocket on the right side, and an absence of gas shadows in the rest of the abdomen (Fig. 1). A transthoracic cardiac Doppler ultrasound was performed and noted dextrocardia and a 7.43 mm inter-atrial communication with a left-to-right shunt without evidence of pulmonary hypertension. The diagnosis of probable duodenal atresia with situs inversus totalis was made.

After optimization, the baby underwent operation through a left supra-umbilical transverse laparotomy that showed the liver and cecum on the left side of the abdomen; whereas, the stomach and spleen were on the right side. The proximal duodenum was dilated with an annular pancreas encircling the 2nd and 3rd parts of the duodenum (Fig. 2). A duodenojejunostomy was performed in the manner of a Kimura diamond duodenoduodenostomy. An appendectomy was also performed. The immediate postoperative course was unremarkable however the baby developed bowel evisceration through the surgical wound on the 5th postoperative day, which was addressed by re-suturing the abdominal wound. The postoperative course remained uneventful and the patient was allowed orally on the 7th postoperative day. The patient was discharged in good

clinical condition. The baby is thriving well and doing fine on a 6-month follow-up.



Figure 1: Thoraco-abdominal radiography made on day 4 of life before admission.



Figure 2: Intraoperative image showing annular pancreas and duodenal atresia.

DISCUSSION

Situs solitus refers to the normal anatomic arrangement of thoracic and abdominal organs whereas its mirror arrangement is called situs inversus. [2] Heterotaxy of both abdominal and thoracic viscera is termed situs inversus totalis. [3] Its incidence is approximately one in 5,000 to 20,000 live births. [4] Most cases are associated with other congenital malformations, such as cardiac anomalies, duodenal atresia, annular pancreas, biliary atresia, preduodenal portal vein, diaphragmatic hernia, and renal dysplasia [Table 1]. [8-14] The index case had duodenal atresia, annular pancreas, and cardiac defects associated with situs inversus totalis.

Table 1: Duodenal atresia with annular pancreas and situs inversus in the literature

| Author and Year | Age, Sex | Situs inversus | Associated anomalies |
|-------------------------------|----------|-------------------|--|
| Adeyemi SD 1988 [8] | 7d, F | Abdominis | Duodenal atresia, complete annular pancreas surrounding duodenum |
| Adeyemi SD 1988 [8] | 12d, M | Abdominis | Duodenal atresia, annular pancreas incomplete, mucosal diaphragm |
| Adeyemi SD 1988 [8] | 8d, M | Abdominis | Duodenal atresia, complete annular pancreas surrounding duodenum |
| Iuchtman et al. 1993 [9] | 11d, F | Abdominis | Incomplete annular pancreas and duodenal diaphragm with internal hernias |
| Chang et al. 1993 [10] | - | Abdominis | Pulmonary stenosis, AV canal, asplenia, annular pancreas and duodenal atresia |
| Kayhan et al. 2009 [11] | 70y, F | Thoraco-abdominal | Duodenal stenosis with incomplete annular pancreas, preduodenal portal vein and polysplenia |
| Park et al. 2009 [12] | 6d, F | Abdominis | Polysplenia with incomplete annular pancreas and duodenal diaphragm |
| Ben Ahmed et al. 2012 [13] | 18d, F | Thoraco-abdominal | Polysplenia with incomplete annular pancreas, duodenal diaphragm, and intestinal malrotation |
| Wabada et al. 2015 [2] | 10d, M | Abdominis | Duodenal atresia with preduodenal portal vein, annular pancreas, and intestinal malrotation |
| Pattanshetti et al. 2020 [14] | 3y, M | Abdominis | Duodenal stenosis with incomplete annular pancreas and polysplenia |
| Our case 2023 | 6d, M | Thoraco-abdominal | Duodenal atresia, complete annular pancreas surrounding duodenum |

Talabi et al [5], in a literature review, found 25 neonates with duodenal atresia and situs inversus totalis; of these, only 04 neonates also had annular pancreas. The simultaneous occurrence of these anomalies is exceedingly rare. A review of the English medical literature revealed only 11 such cases (Table 1).

In the context of duodenal atresia, these anomalies are usually identified most at surgery [7]; however, in the index case, we had a suspicion of situs inversus totalis preoperatively based on findings of the right-sided apex of the heart with right-sided gastric bubble. Nevertheless, the annular pancreas was identified only intraoperatively.

Preoperative identification of the situs inversus is crucial as it helps in surgical decision-making and tailoring the surgical procedure. [8] Similarly, in our case, it guided the surgeon to go for a left-sided transverse laparotomy instead of the standard right one. Additionally, the presence of an annular pancreas covering the 2nd and 3rd parts of the duodenum also prompted us to go for a diamond duodenojejunostomy instead of a standard diamond-shaped duodeno-duodenostomy.

The situs inversus totalis associated with duodenal atresia and an annular pancreas is complex, rare, and constitutes a challenge for the surgical team. This report highlights the importance of tailoring surgical decisions based on the individualized needs of the patient.

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