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Radiological and Clinical Aspects of Duodenal Atresia Associated with Situs Inversus: A Case Report

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Abstract **Case Report**

Duodenal atresias are rare digestive embryopathies that present as typical high intestinal obstructions. They are associated in 30 to 50% of cases with other anomalies such as Trisomy 21, situs inversus, congenital heart diseases, digestive tract anomalies, and biliopancreatic abnormalities, among others. Situs inversus is a particularly rare association. Radiologists must have a thorough understanding and be able to identify associated malformations to provide useful interpretations. The purpose of this work is to present their clinical and radiological aspects, as well as management approaches. The diagnosis is straightforward in the presence of bilious vomiting with a flat abdomen and findings from plain abdominal radiography. Ultrasound facilitates the identification of any associated malformations, including situs inversus.

Keywords: Duodenal atresias, interpretations, heart diseases.

INTRODUCTION

Duodenal atresia associated with situs inversus is a rare combination. Intestinal obstruction due to malrotation or intestinal atresia occurs in about 6 per 10,000 live births. Duodenal atresia represents up to 60% of intestinal atresias; malrotation is a congenital anomaly of intestinal positioning and can lead to volvulus of the midgut around a narrow-based mesentery [1]. It is estimated that an asymptomatic rotational anomaly occurs in 1 in 200 live births; however, symptomatic malrotation is less frequent (1 in 6,000 live births) [2].

The exact cause is unknown but is believed to be due to a defect in the formation or involution of an intestinal segment following a mesenteric vascular accident in utero, resulting in ischemia or necrosis of an intestinal segment occurring after organogenesis [3].

Diagnosis is usually suggested in the presence of neonatal obstruction with a flat abdomen, most often bilious vomiting, with or without meconium elimination abnormalities, and is confirmed by a thoraco-abdominal radiograph showing the characteristic "double bubble" image, with contrast imaging used if necessary. Prenatal diagnosis occurs in 80 to 90% of cases (diagnosed by ultrasound in the seventh month or earlier showing the characteristic "double bubble" image). Differential diagnoses include pyloric stenosis in incomplete diaphragms with late presentation, other intestinal atresias, intestinal volvulus on a common mesentery, and duodenal duplication [4].

The rarity of this association warrants this case report, which aims to describe the radiological and clinical aspects and management.

PATIENT AND OBSERVATION

Observation

The patient is a 2-day-old female newborn, from a non-consanguineous marriage, with a monitored pregnancy, delivered at 36 weeks. She was delivered by cesarean section due to the threat of premature labor and severe polyhydramnios, with a birth weight of 2500 g and an APGAR score of 8/9/10. The parents reported concerns about the absence of meconium passage at birth, accompanied by postprandial bilious vomiting. No particular medical history was found.

Clinical Findings:

The clinical examination revealed a general state of deterioration marked by dehydration, and subicterus in a non-febrile context. The thorax was

unremarkable. The abdomen showed an epigastric bulge with a soft, non-distended abdomen that was examined without resistance, and a silent auscultation. On rectal examination, the anal margin was clean, the anal sphincter was tonic, the mucosa was soft, the rectum was empty, and the Douglas pouch was not distended with a negative probe test. No clinically detectable malformations were noted.

Diagnostic Approach

Two Exams Were Requested:

An ultrasound and an abdominal X-ray without preparation the ultrasound, performed with a high-frequency probe, revealed gastric dilation of the duodenum upstream of a short stenosis of approximately 15 mm. It also highlighted a mirror-image arrangement of the intra-abdominal organs (situs inversus).

The abdominal X-ray showed a "double bubble" appearance, suggesting a diagnosis of duodenal atresia.

Therapeutic Intervention

Under general anesthesia, the newborn was placed in a dorsal position on a surgical table, with the insertion of a nasogastric tube and bladder catheter.

A laparotomy was performed through a suprapubic incision. The abdominal cavity opening revealed a significantly dilated stomach and the first two duodenal segments, with duodenal and jejunal atresia and malrotation of the common mesentery associated with situs inversus. The surgical procedure involved resection of the duodenal and jejunal atresia segments, followed by a termino-lateral and duodeno-jejunostomy anastomosis. A final check of the bowel loops showed no abnormalities.

Postoperatively, the patient was monitored in pediatric intensive care and received analgesics (Perfalgan intravenously, 15 mg/kg every 6 hours), gastroprotectors (omeprazole 1 mg/kg/day IV), a bi-antibiotic regimen (C3G 50 mg/kg/day, Flagyl 20-30 mg/kg/day), and was kept NPO (nil per os) for 6 days.

Follow-up and Outcome of the Therapeutic Intervention: The postoperative course was marked by hypovolemic shock on the fifth day, which led to the newborn's death.

Informed Consent

After discussions with the medical team, the parents of our patient had freely consented to the therapeutic protocols and expressed their approval of the established plan.

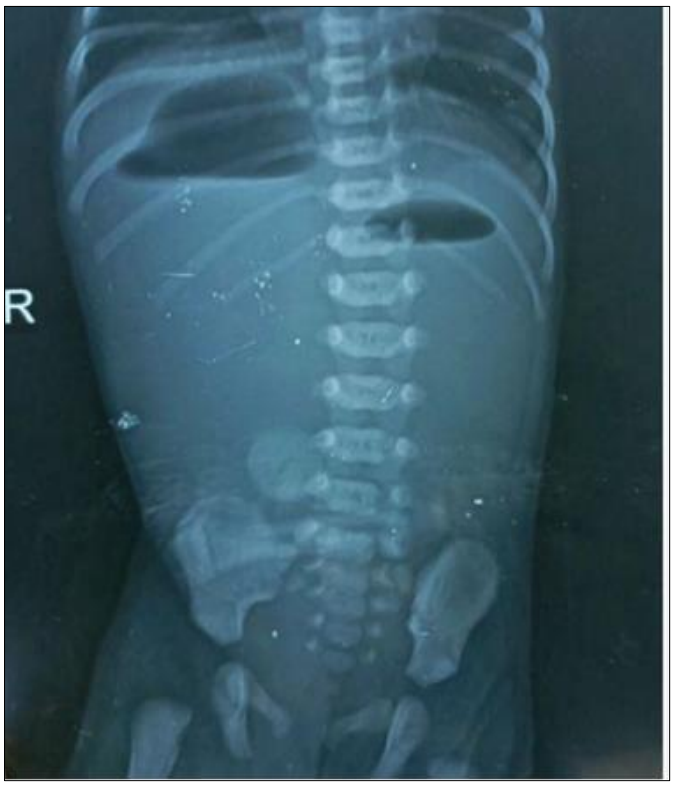

Figure 1: Abdominal X-ray without preparation

Shows a "double bubble" image. This image indicates a large gastric air dilation predominantly on the right and a small duodenal dilation on the left (situs inversus).

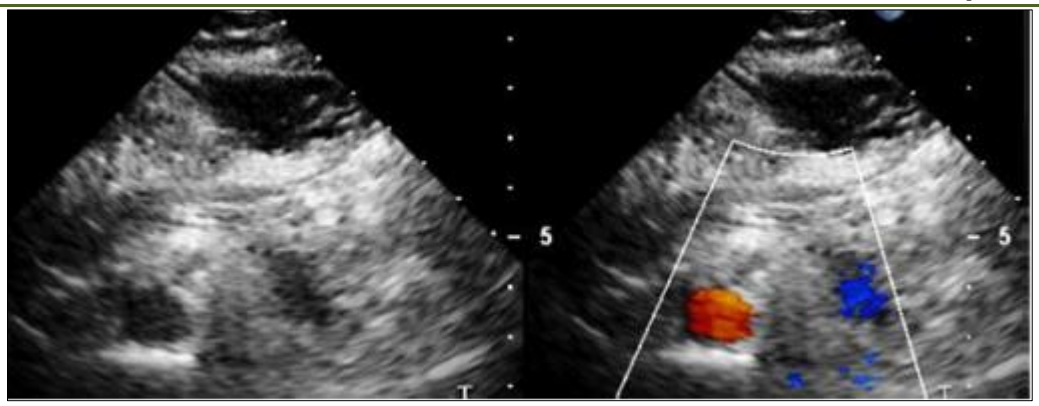

Figure 2: Abdominal Ultrasound with a Slice through the Aorta and IVC

Shows the aorta located on the right and the inferior vena cava on the left.

DISCUSSION

Our study highlights the role of imaging in diagnosing a rare digestive malformation association not frequently encountered in our daily practice. It underscores, however, the limitations in management. This can be attributed partly to the frequent poly-malformative nature of such clinical presentations; some of these malformations are not clinically detectable. Additionally, the difficulty of providing postoperative nutrition to these patients is a challenge, as appropriate parenteral nutrition is often not only unavailable but also inaccessible to many social strata. Congenital intestinal atresia occurs in 1 in 20,000 live births. The causes of these atresias are varied, including intrauterine mesenteric vascular accidents, genetic anomalies, or states of hypercoagulability [5].

Viral theories implicating the varicella virus in the development of these atresias have also been proposed. The damage caused to the enteric plexus neurons by this aggressive virus could lead to poor development of the vessels and the appearance of ischemic lesions, resulting in intestinal atresia [6, 7]. The incidence of this embryopathy is between 1 in 10,000 and 1 in 6,000 live births, with a male-to-female ratio close to 1 [8]. Clinically, the newborn presents with very early vomiting, most often bilious, without abdominal distension.

Radiologically, the plain abdominal X-ray confirms the diagnosis before any clinical signs appear. It shows the characteristic "double bubble" appearance, which is pathognomonic for duodenal obstruction. This image indicates a large gastric dilation, predominantly on the left, and a small duodenal dilation on the right. It may appear purely gaseous or hydroaeric depending on the imaging angle, whether the patient is upright or supine, and whether or not gastric aspiration has been performed. If the newborn has not been aspirated or has not vomited excessively beforehand, gastric aspiration might minimize the double bubble effect, and a few cubic centimeters of air might be introduced via the gastric tube. The nature of the obstruction is suggested by the presence or absence of air downstream of the obstruction. The absence of air downstream supports a complete obstruction, such as atresia.

Abdominal ultrasound, when performed by skilled practitioners, can show the image of the stomach and duodenum dilated upstream of an obstruction, the position of the superior mesenteric vessels, and can also identify any associated malformations. Newborns typically present with very early vomiting, usually bilious, weight loss, and without abdominal distension [9].

The prognosis depends on the remaining length of the small intestine and the presence of the ileocecal valve. The short bowel syndrome is a fatal complication requiring total and prolonged parenteral nutrition. The rectal tube test was negative, which is consistent with high neonatal obstructions (obstructions with a flat abdomen). The Louw classification, modified by Grosfeld, categorizes duodenal atresias into four types [10]:

Type I: Internal membrane with continuity of the serosa, no mesenteric defect.
Type II: Serosal discontinuity with a cord between the proximal and distal ends.
Type III: Serosal discontinuity with a mesenteric defect (subdivided by Grosfeld into A and B):
Type IIIa: Mesenteric defect only.
Type IIIb: Apple peel deformity.
Type IV: Complete atresia; multiple atresias.

In our case, it was a complete atresia type IV (Figure 1) associated with situs inversus. The surgical intervention is a duodeno-duodenostomy, where the proximal and distal duodenal pouches are opened and joined, effectively bypassing the atretic segment. This procedure can be performed either by laparotomy or laparoscopy [10]. In our context, laparotomy remains the preferred approach for newborns.

CONCLUSION

The association between duodenal atresia and situs inversus is rarely described. These two entities, with different etiopathogenesis, seem to have distinct origins when associated. It is crucial for all physicians to be aware of this association, which should be considered in cases of intestinal obstruction with a flat abdomen, very early bilious vomiting, weight loss, and without abdominal distension. This may indicate the need for diagnostic imaging. The outcome depends on the timeliness of the diagnosis, the surgical intervention, and effective postoperative care and nutrition. Improving postoperative prognosis remains a significant challenge.

Conflicts of Interest: The authors declare no conflicts of interest.

Contributions of the Authors: All authors contributed to the conduct of this work. They have read and approved the final version of the manuscript.

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