Annular pancreas in neonate patients
Vomiting and weight loss in a 13-day newborn

Pâncreas Anular no período neonatal
Vômitos e perda de peso em neonato com 13 dias de vida

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ABSTRACT

Objective: Reinforce that, despite being a rare condition, the annular pancreas must be considered a differential diagnosis for intestinal obstructions in the newborn period. **Case description:** This case report refers to a 13-days newborn admitted to the emergency room presenting with postprandial vomiting and progressive weight loss since maternity discharge. The patient did not respond to initial interventions that included IV antibiotics and nasogastric tube insertion. A contrast study of the upper digestive tract (esophagus, stomach, and duodenum) found a luminal narrowing on the second duodenal portion. The patient was submitted to an exploratory laparotomy which found a pancreatic-tissue ring involving the second part of the duodenum. Despite rare, we reinforce that the annular pancreas must be considered a differential diagnosis for intestinal obstructions in the newborn period. **Comments**: Annular pancreas is a rare congenital defect in which a ring of pancreatic tissue encircles the duodenum, causing different degrees of intestinal obstruction.

Keywords: Pancreas, Pancreatic diseases, Intestinal obstruction, Newborn.

RESUMO

Objetivo: Evidenciar que, apesar de condição rara, o pâncreas anular deve se firmar como diagnóstico diferencial das obstruções intestinais no período neonatal. **Descrição do caso:** Este relato aborda o caso de uma paciente de 13 dias de vida admitida no pronto atendimento com queixa de volumosos vômitos pós-prandiais e perda de peso progressiva desde a alta da maternidade. Apesar das medidas iniciais, com ressuscitação volêmica, antibioticoterapia e passagem de sonda nasogástrica para descompressão, a paciente evolui sem melhora. Exame contrastado de esôfago-estômago-duodeno detectou estreitamento luminal da segunda porção duodenal. Paciente submetida à laparotomia exploradora, que evidenciou anel de tecido pancreático estreitando o trânsito intestinal na região. **Comentários**: O pâncreas anular é uma anomalia congênita rara na qual um anel de tecido pancreático envolve a porção descendente do duodeno, causando graus variados de obstrução intestinal extrínseca**.**

Palavras-chave: Pâncreas, Pancreatopatias, Obstrução intestinal, Recém-nascido.

CASE REPORT:

A female full-term appropriate for gestational age newborn patient checked in at an emergency care unit at 13 days of age, presenting with daily postprandial vomiting within 15 to 20 minutes after nursing. The patient had lost 6% of her weight compared to that of checkout from the maternity unit. Her mother had received routine prenatal care with no complications. She had been on carbamazepine due to epilepsy history and denied using other drugs. After birth, the patient remained hospitalized in the maternity unit for seven days due to respiratory discomfort, requiring supplemental oxygen and need of 48 hours of phototherapy due to neonatal jaundice with ABO incompatibility. Furthermore, the patient was submitted to a repeated newborn bloodspot workout due to initial altered results in hypothyroidism and aminoacidopathies screening. The patient also had a bone deformity in the sacral region and was referred to follow-up care to confirm a diagnosis of a hemangioma.

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On physical examination, the patient was hypoactive, pale and dehydrated, but maintained symmetrical peripheral pulses, capillary refill time under 3 seconds and normal blood pressure. Abdominal palpation was normal and no other abnormal findings were identified. She exhibited two episodes of vomiting during the initial examination, having been nursed immediately before entering the doctor's office.

Laboratory exams revealed that the patient was alkalotic and had increased lactate. Urine test type 1 results indicated hematuria and leukocyturia (Table 1). A lumbar puncture was not performed to rule out meningitis due to the sacral bone deformity. The patient was started on cefotaxime and oxacillin for presumed late neonatal sepsis, with a working diagnosis of urinary tract infection, and was admitted to the intensive care unit.

Table 1.
Laboratory examinations at the ER

Blood count	Hb 17,10 | Ht 47,80 | WBC 15.800 | platelets 452.000	**CRP**	79,80 mg/dL
Blood Culture	Negative	**Venous Lactate**	29 mmol/L
Urinalysis	Turbid, nitrite +, ketone +, protein +/4, RBC 123.000, WBC 190.000	**Creatinine**	0,77 mg/dL
Urine culture	Negative	**BUN**	29 mg/dL
Na	131 mEq/L	**Glucose**	90 mg/dL
K	4,80 mEq/L	**Cai**	0,96 mEq/L
Arterial Blood Gas	pH 7,65 | pCO2 30 | pO2 53 | HCO3 34 | BE +13,20	**Mg**	1,50 mEq/L

Laboratory tests collected at the admission on the Emergency Room (ER), with respective units of measurement

RBC: red blood cells | WBC: white blood cells | Na: sodium | K: potassium | CRP: C-reactive protein | BUN: blood urea nitrogen | Cai: ionic calcium | Mg: magnesium

She persisted with vomiting, requiring a nasogastric tube to decompress the stomach. After this procedure, the thoracoabdominal X-ray revealed great gastric distension. The possibility of mechanical obstruction of the gastrointestinal tract was considered and a bedside abdominal ultrasound was performed, which indicated no signs of pyloric stenosis. After the pediatric surgical team's evaluation, an upper gastrointestinal barium swallow test was ordered, revealing a narrowing of the second duodenal portion, with the passage of only a small amount of contrast.

A diagnostic hypothesis of duodenal membrane was suggested and an upper digestive endoscopy was scheduled for surgical evaluation. During the procedure, an extrinsic obstruction was identified and a conversion to open laparotomy was made. A right transverse incision was made and a ring of pancreatic tissue involving the second portion of the duodenum was identified. A conventional diamond-shaped duodenoduodenostomy was performed without complications. This technique consists of a longitudinal incision on the cranial portion of the obstruction and a transversal incision on the distal side. After that, an interrupted suture is performed initially on the posterior duodenal wall and then on the anterior wall. In that way, we can bypass the obstruction, creating a new lumen for the digestive tract.

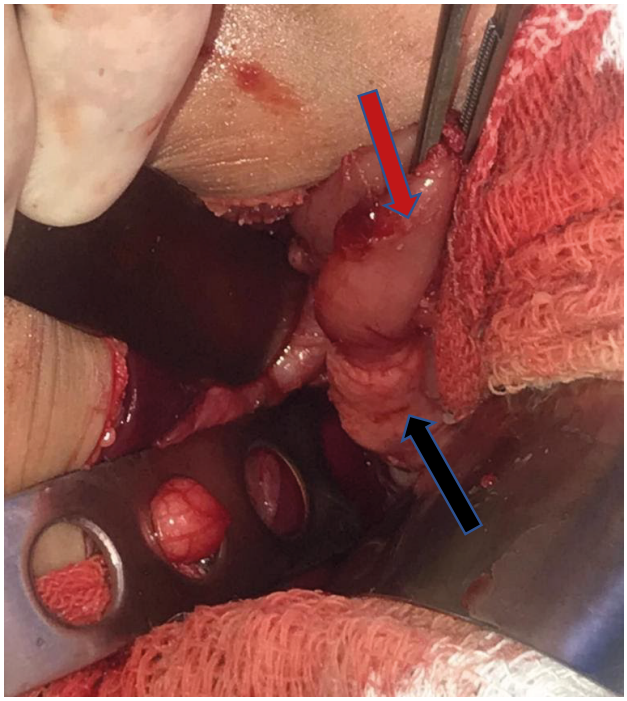

Figure 1: Intra-operatory image representing pancreatic tissue (black arrow) involving the duodenum (red arrow)

DISCUSSION

An annular pancreas is a congenital malformation characterized by a band or ring of pancreatic tissue surrounding the lower portion of the duodenum, causing different degrees of extrinsic intestinal obstruction. It results from a failed fusion of the dorsal and ventral pancreatic leaflets during the embryonic stage, between four and eight weeks of gestational age.

Clinical presentation can vary significantly, from asymptomatic cases to complete intestinal obstruction. The most common symptom is postprandial vomiting. The age of diagnosis also varies from newborns to elderly patients, according to the degree of obstruction.

This anomaly is responsible for about 1% of intestinal obstructions in the neonatal period. Although rare, its correct diagnosis is essential because 40% of cases present themselves in situations that require immediate surgery.

The pancreas is formed between the fourth and eighth week of embryonic life, resulting from the fusion of a dorsal portion – which will form the body and tail – and a ventral bifid portion – which will form the head. Usually, the two components of the ventral pancreatic portion fuse and rotate around the duodenum, placing themselves under the dorsal pancreatic portion. Rarely, these two components migrate in opposite directions, surrounding the duodenum and thus forming a pancreatic ring.

The finding of the annular pancreas is often associated with polyhydramnios in gestational ultrasound, particularly in complete rings. Furthermore, it may be associated with other congenital anomalies in up to 71% of cases. The literature mentions intestinal duplication, intestinal malrotation, and even Meckel's diverticulum.[5,8] There is also a strong association with Down syndrome.[9] In the case reported, however, the other malformation was a bone deformity in the sacral region, not usually associated with the annular pancreas.

About two-thirds of patients are asymptomatic throughout life. The pancreatic ring is partial in about 75% of cases, an anatomical disposition that can be asymptomatic and diagnosed only in adulthood or in necrotic studies. However, if the pancreatic ring causes duodenal obstruction, symptoms may appear soon after birth and are often indistinguishable from duodenal atresia or intestinal volvulus.[4] In these cases, the patient presents with food intolerance, vomiting, and abdominal distension, such as in the case described. In the neonatal period, the primary differential diagnoses are pyloric stenosis, intestinal atresia, and intestinal volvulus. In addition, a substantial number of adults with this condition develop neoplasms (24%), especially biliary and pancreatic adenocarcinoma. [1,10]

The diagnosis can be suggested through imaging tests such as X-ray, barium swallow, and Computed Tomography (CT), but the definitive diagnosis is surgical.[2] Although endoscopic approaches are described, open surgical management is currently considered the first-line treatment. The purpose of surgery is to unclog the duodenum or intestinal loop, bypassing the pancreatic ring. Removing the ring is avoided as this procedure is more associated with pancreatitis and fistula formation. In neonates, a duodenum-duodenum anastomosis is the procedure of choice.[6]

Prognosis depends on the age at onset of symptoms, early diagnosis through prenatal ultrasound, and association with other congenital abnormalities. Mortality is higher with associated congenital cardiac abnormalities than with duodenal obstruction, whether complete or incomplete.[7]

CONCLUSION

This report highlights the importance of investigating the causes of intestinal obstruction in newborns with persistent vomiting. An annular pancreas, although rare, can present with these symptoms and represents a surgical emergency.

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SCR: Data collection, translation
LGB: Writing and edition
CETB: Data collection, translation
GFP: Supervision, general discussion
GAC: Review, supervision
VMCM: Supervision, general discussion

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