Annular Pancreas: Unusual Presentation of a Rare Malformation

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Abstract

A case of a 6-month-old female infant presenting postprandial vomiting and poor weight gain is presented. She was malnourished and dehydrated with no abdominal distension or neurological signs. Laboratory testing revealed hypochloremic hypokalemic alkalosis. After the exclusion of infectious disease and food intolerance, plain abdominal X-rays revealed a "double bubble" sign with distal air, thereby suggesting partial duodenal obstruction. An upper gastrointestinal contrast study confirmed this diagnosis. She was then referred to a tertiary hospital for surgical treatment. At surgery, a duodenum-duodenostomy was performed due to a complete annular pancreas, with an excellent outcome. Annular pancreas is a rare entity. When symptoms arise, they usually occur in the neonatal period. As the present case illustrates, it may be symptomatic at later ages, requiring a high grade of suspicion in order to diagnose and treat it early.

Keywords: Duodenal Obstruction/congenital; Duodenal Obstruction/diagnosis; Duodenal Obstruction/therapy; Infant; Pancreas/abnormalities

Introduction

Congenital intestinal obstruction of the duodenum occurs in one out of 2,500 to 10,000 births.[1] The typical clinical presentation occurs in the neonatal period with early and frequent vomiting. However, the associated symptomatology depends not only on the degree of stenosis but also on the type of anatomical defect that may be complete or incomplete, extrinsic or intrinsic. Duodenal atresia, including duodenal membrane, are classified as intrinsic defects, whereas intestinal malrotation, duodenal duplication, annular pancreas, and preduodenal portal vein are considered extrinsic defects.[1-3]

The annular pancreas is a rare congenital malformation that occurs in about one out of 10,000-20,000 live births. It is characterized by a band of pancreatic tissue that partially or totally surrounds the second portion of the duodenum, causing a variable degree of obstruction.[4]

Herein, we report a clinical case of complete annular pancreas that resulted in partial duodenal obstruction presenting at a late age.

Case Report

We report on a case of a caucasian 6-month-old female, as the second daughter of healthy parents. She was born from a term gestation with adequate follow-up and no intercurrences. The somatometry at birth was appropriate for gestational age; the neonatal period had no complications with the passing of meconium in the first 24 hours of life.

At 4 months of age, when she started food diversification, intermittent postprandial vomiting developed, predominantly after pasty meals (soup, baby cereals, or fruit). Episodes of vomiting became progressively more intense (in frequency and content). At 6 months of age, the weight decreased from percentile (P) 50 to under P3 and height from P50 to P15. The bowel habit was irregular but with normal consistency stools.

At admission, she was malnourished with loss of muscle mass and subcutaneous fat. The abdomen was depressed with no palpable masses or organomegaly. The remaining examination was unremarkable. Laboratory testing revealed hypochloremic hypokalemic metabolic alkalosis (pH 7.59, chlorine 91 mmol/L, potassium 3.28 mmol/L). Renal and hepatic functions were within the normal ranges. There was no evidence of infectious disease and no alterations in food specific immunoglobulins or celiac disease specific antibodies.

Abdominal ultrasound showed the dilatation of the stomach with water and gas content. Plain abdominal X-rays revealed a "double bubble" sign (gastric and duodenal dilatation) with distal air, suggesting partial

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duodenal obstruction (Fig. 1). The upper gastrointestinal contrast study demonstrated an exuberant dilatation of the stomach and first duodenal portion, with the filiform progression of contrast to the second portion of the duodenum after about 40 minutes of the beginning of the examination, confirming the diagnosis of partial duodenal obstruction (Fig. 2). The child was then referred to a tertiary hospital where she underwent surgical treatment that revealed a "complete" annular pancreas that was managed with a side to side duodenum-duodenostomy.

The postoperative course was uneventful with oral tolerance from the fourth day. Regular surveillance by a pediatrician has shown excellent weight improvement (Fig. 3).

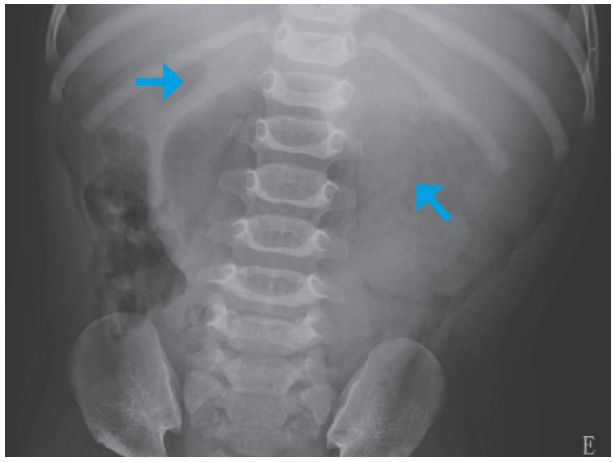

Figure 1. Plain abdominal X-ray showing the "double bubble" sign due to the dilatation of the stomach and the first portion of the duodenum (arrows); distal air suggests a partial duodenal obstruction.

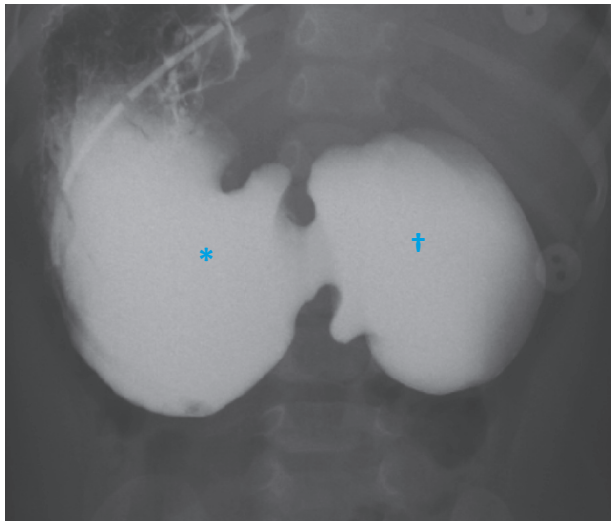

Figure 2. Upper gastrointestinal contrast study (posteroanterior view) showing the marked distension of the stomach (*) and the first duodenal portion (†), with the filiform progression of the contrast to the second portion of the duodenum (not clearly visible in this image).

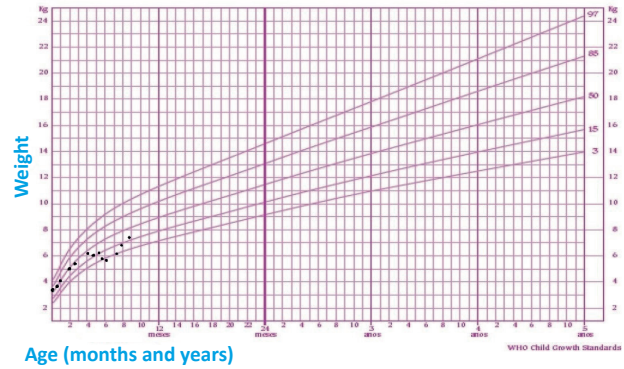

Figure 3. Weight percentiles curve. Decrease in weight at 4 months of age and subsequent recovery after surgery performed at 7 months of age.

Discussion

Intestinal obstruction is a common neonatal surgical emergency.[5] The annular pancreas is a rare malformation, accounting for approximately 1% of the causes of bowel obstruction in newborns.[6]

The pancreas develops between the sixth and eighth week of gestation. Its final location and shape result from the fusion of two evaginations, the dorsal and ventral, of the primitive duodenum. During this process of embryogenesis, the ventral evagination, located to the right of the duodenum, follows a rotational movement posterior to the second portion of the duodenum and merges with the dorsal evagination. The annular pancreas results from an error in the rotation of ventral evagination in this process of morphogenesis, resulting in the circular involvement of the second portion of the duodenum by pancreatic tissue.[7] This ring may be complete or incomplete (in 25% and 75% of cases, respectively).[6,8] There are several theories proposed to explain the origin of the annular pancreas but there is no clear explanation of the mechanism underlying this malformation.[9]

The prenatal diagnosis of this malformation is extremely rare, but it can be performed from the second trimester of pregnancy through the identification of two characteristic findings on the obstetric ultrasound: the "double bubble" sign and the presence of hyperechogenic bands around the duodenum.[6]

The age of the clinical presentation of annular pancreas depends on the degree of constriction that confers on the duodenum and on the coexistence of other malformations.[10] The degree of constriction is variable and there is often an associated duodenum atresia.[2,3]

In more than two thirds of cases, the diagnosis of intestinal obstruction occurs in the neonatal period, with patients presenting non-biliary vomiting, localized

abdominal distension, decreased bowel movements, and food intolerance.[9,11,12] Non-biliary vomiting is more frequent in children with an annular pancreas than in those with duodenal obstruction of other causes, since in the former the obstruction is usually upstream of the ampulla.[13] There may also be a history of prematurity, low birth weight, and/or polyhydramnios. In cases of incomplete obstruction, the clinical presentation may be subtler with recurrent vomiting.[2.11]

The diagnosis of annular pancreas is usually intra-surgical during the exploration of the duodenum-cephalopancreatic region.[12,14] However, the diagnosis of duodenal obstruction can be made by clinical and imaging data. The "double bubble" sign in the plain abdominal X-rays corresponds to the gastric and duodenal distension (in the absence of distal air is the pathognomonic of complete duodenal obstruction; with distal air being suggestive of partial obstruction), a finding not specific of an annular pancreas. A contrast study demonstrates circumferential narrowing of the second portion of the duodenum. Abdominal ultrasound has a relatively low diagnostic accuracy because pancreatic ring observation is not always possible. However, the typical abnormal pancreas head conformation of "crocodile jaw" associated with a dilated duodenum may be visible. Computed tomography and magnetic resonance imaging may show pancreatic tissue around the duodenum; but it should not be performed routinely.[15,16] In neonates, a plain abdominal X-ray demonstrating the "double bubble" sign with no distal air or, if present, with an upper digestive contrast study revealing partial duodenal obstruction is enough to indicate surgery.[14]

Initially, the surgical approach was based on the resection of the pancreatic ring, in order to release the duodenum. However, this method was associated with severe complications such as pancreatic fistulas and pancreatitis.[12-14] In addition, duodenal membranes and duodenal stenosis are often associated with the annular pancreas so this technique *per se* does not guarantee duodenal patency.[17] Performing a bypass through duodenostomy is the procedure of choice for the treatment of duodenal obstruction by an annular pancreas. During the procedure, the surgeon should be alert to other existing malformations.[7,13] With the enhancement of surgical and anesthetic techniques, neonatal intensive care and nutrition, together with the meticulous treatment of other malformations associated with the annular pancreas, there has been a substantial improvement in surgical morbidity and mortality rates in the last decades.[11,13]

The purpose of this article is to share a clinical case of a rare malformation with an unusual and insidious clinical onset. The diagnosis of partial congenital duodenal obstruction can be challenging; however, with a detailed clinical history associated with a careful choice of diagnostic tests, the diagnostic accuracy increases. Surgery is generally the specific diagnostic method and therapy of choice for the symptomatic annular pancreas. The procedure presents an excellent outcome and, when performed early, avoids complications such as hydroelectrolytic, metabolic, or ponderal imbalances.

WHAT THIS CASE REPORT ADDS

- The approach of the child with persistent vomiting requires detailed clinical data acquisition and a meticulous objective examination associated with the support of laboratory and imaging studies.
- Although it usually presents in the neonatal period, congenital partial duodenal obstruction should be taken in account even in older children with recurrent vomiting.
- Despite being not the main etiology, the annular pancreas may be the cause of partial duodenal obstruction and dictates surgical treatment that is associated with excellent outcome.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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Pâncreas Anular: Apresentação Pouco Habitual de uma Malformação Rara

Resumo:
Descreve-se o caso clínico de uma lactente de seis meses de idade com vómitos pós-prandiais e má evolução ponderal com dois meses de evolução, coincidente com início da diversificação alimentar. Apresentava-se desnutrida, desidratada, sem distensão abdominal ou alterações neurológicas. Os exames laboratoriais revelaram alcalose hipoclorémica e hipocaliémica. No internamento manteve vómitos pós-prandiais e obstipação. Após exclusão de causas infeciosas e de intolerância alimentar, realizou estudo esofagogastroduodenal contrastado que demonstrou dilatação acentuada do estômago e primeira porção do duodeno com passagem filiforme do contraste para a segunda porção duodenal. Com o diagnóstico de obstrução duodenal parcial, foi transferida para um hospital terciário onde foi submetida a intervenção cirúrgica na qual se identificou pâncreas anular completo, sendo efetuada duodeno-duodenostomia latero-lateral com excelente evolução pós-operatória.
O pâncreas anular é uma entidade rara, assintomática na maioria dos casos. Quando associado a sintomatologia esta ocorre tipicamente no período neonatal, raramente surgindo em idades mais tardias.

Palavras-Chave: Lactente; Obstrução Duodenal/congénito; Obstrução Duodenal/diagnóstico; Obstrução Duodenal/tratamento; Pâncreas/anomalias congénitas