

An Unusual Presentation of Annular Pancreas: A Case Report

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Abstract

Annular pancreas (AP) is a rare congenital malformation resulting from failure of pancreas ventral anlage rotation with the duodenum. This leads to a ring of pancreatic tissue that envelops the duodenum. Clinical manifestations of AP most commonly develop in infancy or early childhood but can present at any age. The diagnosis of AP, usually suggested by an upper Gastrointestinal (GI) series or abdominal Computerized Tomography (CT) scan, but surgery is considered the gold standard diagnostic method. Surgical bypass of the annulus in all patients with symptomatic AP is recommended. We report a 1 year old girl who presented with intermittent, non projectile, non bilious vomiting that occurred 1h to 2h after feeding since neonatal period. Upper GI contrast study demonstrates, a dilated duodenal bulb associated with narrowing of post bulbar area. The patient underwent surgical correction of the obstruction. A bypass of the ectopic pancreas tissue was performed by duodenoduodenostomy. Considering the rarity of this congenital abnormality, presenting with chronic partial duodenal obstruction, and its successful correction by surgical means have prompted us to report the case.

Key words: Annular pancreas, Duodenal obstruction.

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Received date: Nov 29, 2014 ; Accepted date: Dec 12, 2014

Introduction

Annular pancreas is a rare congenital abnormality which is characterized by a thin, flat band of pancreatic tissue surrounding the second part of duodenum, and may lead to partial or complete duodenal obstruction (1, 2). The age at presentation is related to degree of duodenal obstruction and associated malformations (for example, duodenal atresia) (3, 4).

Case report

The patient was a 1-year-old girl who presented to our department with intermittent, non-bilious vomiting occurring after the ingestion of breast milk and liquid diet.

He was born at 39 weeks' gestational age with a birth weight of 3.5 kg. The pregnancy was uncomplicated. Vomiting began from ten days of age. By the time of admission, at 12 months of age, the patient had intermittent non-projectile, non-bilious vomiting that occurred 1h to 2h after feeding. She appeared hungry immediately after vomiting. The patient had been offered several different formulas and anti-reflux treatments, but these did not result in symptomatic improvement. Physical examination on admission revealed slight upper abdominal distention. The infant admission weight was 8.5 kg, and otherwise was healthy. Laboratory studies were within normal limits.

Upper Gastrointestinal study (UGI) demonstrated mild distention of the stomach as well as distended duodenal bulb associated with narrowing of post-bulbar area and slow contrast flow through narrow portion (indicative of duodenal stenosis). The duodeno-jejunal junction was at normal position (Figure.1). Upper endoscopy showed marked dilation of the duodenal

bulb, with complete disappearance of the mucosal duodenal sketch that can be attributable to its hypertrophied wall (Figure.2).

The patient underwent surgery on the third day of hospital admission, an incomplete annular pancreas with duodenal stenosis in the second portion of the duodenum, proximal to the ampulla of Vater was observed. A bypass of the ectopic pancreatic tissue was performed by a duodenoduodenostomy. The postoperative course was uncomplicated, and oral feeding was tolerated from the third day. The patient was discharged from hospital on the seventh day after operation. On follow-up, patient was asymptomatic and had acceptable weight gain.

Fig. 1: Upper GI series



Fig. 2: Duodenal bulb in upper endoscopy

Discussion

Symptomatic AP may present at any age. Approximately one – third of the cases present during the neonatal period and half during the first year of life (3,4). It is estimated that approximately two – thirds of patients remain asymptomatic. The age of presentation is related to the severity of duodenal obstruction and coexistent malformations (5). The ventral portion of the pancreas, during migration in the second month of gestation can become misaligned and encircle the second portion of the duodenum, and then fuse with the dorsal aspect of the pancreas (6). Such a ring of pancreas almost invariably is accompanied by, some degree of constriction in the duodenum. If the constriction is minimal, there is little or no intestinal obstruction. A more insidious form of chronic, partial duodenal obstruction similar to our case may also occur. At laparotomy, in our case, an incomplete annular pancreas with stenosis at the second portion of the duodenum, proximal to the ampulla of Vater was detected. Annular pancreas may occur isolated or together with other congenital malformations. The most common associated malformations are usually related

to gastrointestinal system, including duodenal abnormalities (such as web, stenosis and atresia), intestinal malrotation, imperforated anus, tracheoesophageal fistula and Hirschsprung's disease (7, 8). Any associated anomaly was not found in this presented case. Clinical manifestations of AP in more than of two – thirds of children are in the neonatal period typically with features of intestinal obstruction including bilious vomiting, feeding intolerance, and abdominal distention (9).

After the neonatal period, symptoms differ, include recurrent vomiting and chronic gastric distention (10). Recurrent attack of non bilious vomiting and intolerance of solid foods since neonatal period were the most significant symptoms in our case. A plain abdominal X-ray or ultrasound in symptomatic neonates will show the classic double bubble sign, but this finding is not specific for AP (11-14). However, because all patients in this age group with complete or partial duodenal obstruction require surgery, no further testing is usually required. Although in older children, surgery remains the gold standard method of diagnosis, the diagnosis is usually made with an upper GI series or an abdominal CT scan. When the UGI series or abdominal CT scan shows descending duodenal narrowing, AP should be considered in differential diagnosis (15, 16). In children, duodenojejunostomy is the preferred surgical modality, whereas adults more often required endoscopic pancreatobiliary procedures, cholecystectomy, and other pancreatobiliary surgery (17, 18).

Pancreatic annulus often contains a duct that is sometimes adherent tightly to the duodenal wall and also, frequent associated duodenal malformation explains the abandonment of pancreatic ring resection as a method of surgery (19-21).

Conclusion

We reported this case to show that incomplete duodenal obstruction due to annular pancreas have a more insidious presentation, making diagnosis and early intervention more challenging, as it can be easily mistaken with the much more common conditions such as infantile pyloric stenosis and gastro esophageal reflux.

Conflict of interests: None

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