Case report

A Rare Cause of Sub-Acute Proximal Intestinal Obstruction Due to Annular Pancreas

Weledji EP, Ngowe M, Mokake M

Department of Surgery, Regional Hospital Buea, Cameroon

Correspondence to: E P Weledji, P.O Box 126, Limbe, Cameroon. Email:elroypat@yahoo.co.uk

Summary

Background: Annular pancreas is a rare congenital anomaly due to an abnormal fusion between the tip of the ventral pancreatic bud and the duodenum at about the 7th gestational week and, presenting usually with symptoms due to duodenal obstruction. We report a case of a 2- year old girl who presented electively for investigations of symptoms of a sub-acute proximal intestinal obstruction. Investigations revealed a partial duodenal obstruction and an exploratory laparatomy surprisingly showed a partially obstructing annular pancreas for which she underwent a bypass procedure. A precise preoperative diagnosis of annular pancreas can be difficult without good imaging and the diagnosis and surgical management decision may only be made at laparatomy. Children with atypical or mild symptoms of intestinal obstruction associated with failure to thrive should be investigated fully for a probable mechanical cause.

Keywords: Annular pancreas, Duodenal obstruction, Surgical bypass

Introduction

Annular pancreas is a rare congenital anomaly diagnosed with nearly equal frequency in children and adults (1, 2). This abnormality, although at times clinically silent, may be the cause of a broad spectrum of disease. Complications range from neonatal intestinal obstruction to more complex pathologies in the adult such as recurrent pancreatitis, duodenal or gastric ulceration from duodenal stenosis, obstructive jaundice and malignancy (2-4). In cases of neonatal obstruction, annular pancreas is an important structural and anatomical cause that must be identified and treated appropriately (3). Obviously if obstruction is complete or if ampulla of Vater is involved symptoms are present in the first days of life (2-4). Preampullary duodenal obstruction presenting with non-bilious vomiting occurs commonly and is mostly partial (7). Currently, the majority of cases are diagnosed early in life, and prenatal diagnosis with 2-D ultrasound or specific markers is becoming increasingly important (5). After evaluation, these patients can be managed safely with surgical bypass of the annulus to restore intestinal continuity. Duodeno-duodenostomy is the appropriate treatment (1-7). Prognosis is excellent, despite the frequent association with chromosomal anomalies and major congenital malformations (2, 6).

Case Presentation

A 2- year old girl presented with a history of recurrent episodes of post-prandial vomiting and flatulence since she was about 2 months old. This was associated with loss of appetite, malnutrition, and a gradual failure to thrive. She was not constipated and occasionally had diarrhoea. She was born at term from a normal vaginal delivery and her four siblings are normal. On physical examination she was pale, weak, undernourished with an asthenic habitus and, small for age. She had a distended non-tender upper abdomen with visible and palpable peristalsis. There was no hepatosplenomegaly. No congenital abnormality was noted. An abdominal ultrasound scan revealed an enlarged gastric pouch, stasis of food, normal pylorus with increased peristaltic movements in the antrum suggesting a stenosis of the second part of the duodenum. A plain abdominal X-ray and contrast barium radiography confirmed the obstructed second part of the duodenum with the classical ‘double-bubble sign’ indicating a dilated duodenal bulb and stomach proximal to the obstruction. The differential diagnosis included a duodenal atresia, a stenosis or a duodenal web. She was mildly anaemic with a haemoglobin level of 9.6 gm/dl. She was optimized for an exploratory laparatomy with intravenous hydration and nasogastric suction.

Insert Figure 1 here

Laparatomy revealed a dilated stomach, normal pylorus, dilated first and second part of duodenum. The jejunum and ileum were of normal calibre and exploration of other intra-abdominal organs revealed no other congenital anomalies. Following Kocherisation of the duodenum an annular pancreas was seen partially encircling the second part of the duodenum dorsally (Figs 2a, b).

Insert Figure 2a and 2b here

A simple bypass of the obstruction by an antecolic gastro-jejunostomy was done. The nasogastric tube was left in situ until ileus resolved and enteral feeding was commenced on day 4. She had no post-operative complication and was discharged on the 8th post-operative day.

Discussion

The incidence of this congenital anomaly is reported as 1–3 in 20,000, and some studies have shown that it is more common in males (1, 2). The nature of presentation differs according to the patient's age, although most presentation occurs in infancy or early childhood. The symptoms in general depend on the degree of duodenal obstruction (7, 9). Signs of neonatal intestinal obstruction may initially be non-specific, including poor feeding, vomiting, and irritability. If proximal to the ampulla of Vater, vomiting may be non-bilious, and therefore could be confused with less severe, non–life-threatening conditions (7). Children frequently present with gastrointestinal symptoms, including poor feeding, vomiting, and abdominal distension. In Africa, most patients present late, with malnutrition, failure to thrive, bile-stained vomiting, and less frequently abdominal cramps (9). For some patients as in this case, presentation is delayed by parents because of the non-acute nature of the intestinal obstruction which could be mistaken for an eating or feeding disorder. Thus, these patients will likely be more unwell at presentation and may even die before they reach the hospital. There is a strong association between annular pancreas and other congenital abnormalities with up to 71% of cases have coexisting congenital anomalies (6). The most common association is with Down's syndrome. However, there may be a wide range of associated cardiac and gastrointestinal anomalies (including Hirschsprung's disease and imperforate anus), as well as tracheo-oesophageal fistula and oesophageal atresia (6, 8). Clinical examination findings will vary according to the age at presentation and the extent of systemic upset. In clinically silent cases of annular pancreas, no specific intervention is required (9). Specific management will depend upon the nature of the complication or symptoms caused by the annular pancreas. Successful treatment of neonatal intestinal obstruction requires rapid identification, resuscitation, and definitive management. In addition to the technical skill required to bypass the obstruction caused by aberrant pancreatic tissue, appropriate peri-operative care is required to ensure a successful outcome after surgery (1-9).

The frequently done simple bypass procedures (duodeno-duodenostomy, duodeno-jejunostomy and gastro-enterostomy) have the best results (9, 10). A gastro-jejunostomy was fashioned in this case because of our familiarity with the procedure. Separation of the annular pancreas from the duodenum is associated in 50% of cases with serious complications due to the abnormal proximity of the pancreatic duct.(1) Resection is contraindicated because of risk of post-operative pancreatitis /fistula or a co-existing duodenal web (mucosal diaphragm) or erosion of duodenal wall by penetrating annular pancreas.(1,7-10) The prognosis after surgery for duodenal obstruction is good (100% survival in one series).(1,2,6) However, overall outcome will also be affected by the severity of associated congenital anomalies.(1,2) A close long- term follow-up is

essential for infants and children treated for annular pancreas as many of them (80%) can be expected to develop complications such as obstructive jaundice, upper gastrointestinal motility disorder, failure to thrive, and chronic diarrhea (6). The later three complications may be due to the altered gastrointestinal anatomy.

Conclusion

Annular pancreas is a recognized cause of partial duodenal obstruction in children and by presenting as a salient less life-threatening condition is usually diagnosed late. It would seem advisable that a child with recurrent episodes of vomiting, poor feeding accompanied by developmental retardation should be fully investigated for a possible mechanical cause. Surgeons should always keep this rare diagnosis in mind when a patient present with a sub-acute proximal intestinal obstruction. These patients can be managed safely with surgical bypass of the annulus to restore intestinal continuity but long-term follow-up is essential.

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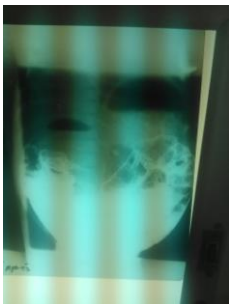

Fig1. Contrast Barium meal “Double bubble” sign

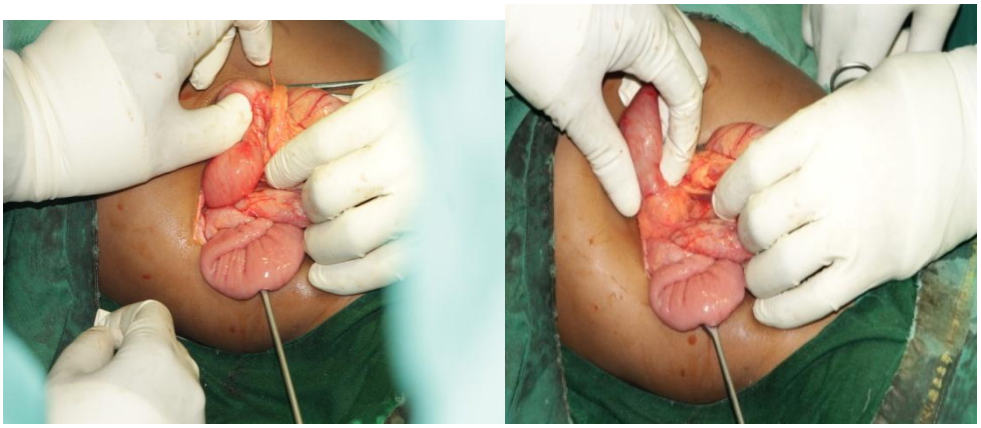

Fig 2a and b: Annular pancreas partially obstructing 2nd part of duodenum