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Annular Pancreas in Adults

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Abstract **Case Report**

Annular pancreas is a rare congenital anomaly characterized by the presence of ectopic pancreatic tissue around the duodenum. Usually diagnosed in the neonatal period, the annular pancreas can be responsible for intestinal obstruction in 1% of cases. In adults it is asymptomatic in 50% of cases, as it can be responsible for duodenal stenosis, pancreatitis, or duodenal or gastric ulcerations. We report two cases of duodenal stenosis secondary to an annular pancreas diagnosed in two adults in whom a surgical bypass was performed allowing resolution of symptoms.

Key words: Annular pancreas, duodenal stenosis, gastro-Entero anastomosis.

INTRODUCTION

Annular pancreas is a rare congenital anomaly. Its annual incidence is estimated at about 1/50,000 births and it represents 10% of duodenal strictures [1]. The annular pancreas can remain totally asymptomatic, only 33% of cases are symptomatic and will be responsible for a wide variety of clinical manifestations of varying severity, which makes its diagnosis difficult [2, 3]. Cross-sectional imaging such as computed tomography (CT) and magnetic resonance imaging (MRI) plays a fundamental role in the positive diagnosis by showing the pancreatic parenchyma islands encircling the 2nd duodenum [4].

MEDICAL OBSERVATIONS

Clinical Situation N°1

An 18-year-old patient was referred to our clinic for the management of epigastric pain associated with nausea and vomiting, which had been progressively worsening for one year. Lateral duodenoscopy revealed a significant concentric narrowing of the second portion of the duodenum.

Abdominal computed tomography (CT) scan showed an annular pancreas encircling the second portion of the duodenum (Figure 1).

Laparotomy confirmed the diagnosis of annular pancreas with dilatation of the first and second portion of the duodenum upstream of an area narrowed by the pancreatic ring that completely surrounded the duodenum and was continuous with the head of the pancreas (Figure 2). No other congenital anomalies of the intra-abdominal organs were noted. A gastroentero-anastomosis was performed without technical difficulty. The patient recovered well and was discharged from the hospital 4 days later. He remains in good general health 8 years after surgery.

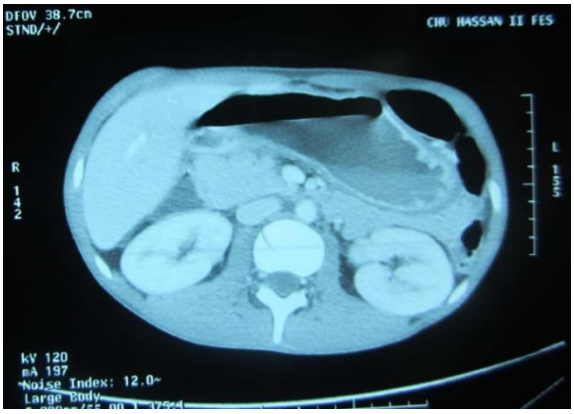

Fig-1: Computerized tomography of abdomen showing the annular pancreas encircling the second part of the duodenum.

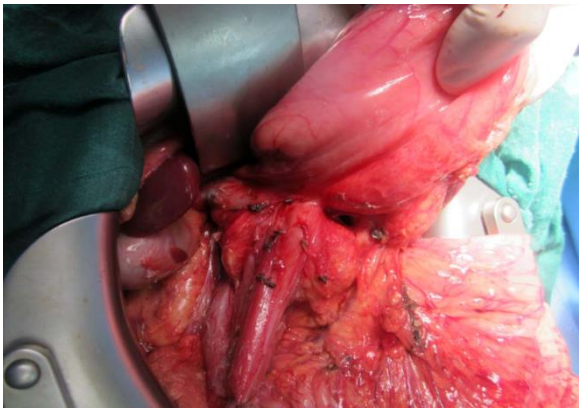

Fig-2: Intra-operative photograph showing an annular ring of pancreatic tissue around the duodenum

Clinical Situation N°2

A 58-year-old patient, cholecystectomized, consulted for an antro-pyloric stenosis syndrome made of chronic late post prandial vomiting with positive fasting. A Endoscopy showed a regular post-bulbar stenosis that could not be crossed. On abdominal CT, it was an extrinsic duodenal stenosis upstream of an annular aspect of the pancreas (Figure 3). On surgical exploration, an annular pancreas responsible for duodenal stenosis was found

(Figure 4). A gastro-entero anastomosis bypassing the duodenal stenosis was performed with simple post-operative follow-ups.

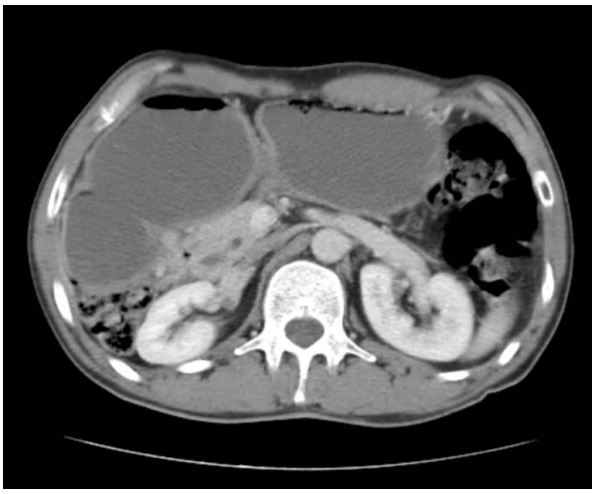

Fig-3: Abdominal CT showing an extrinsic duodenal stenosis upstream of an annular aspect of the pancreas

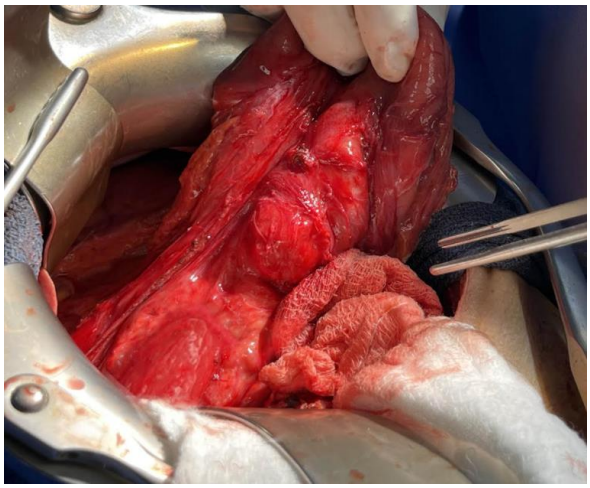

Fig-4: Intra-operative photograph showing an annular pancreas responsible for duodenal stenosis

DISCUSSION

Annular pancreas (AP) is a rare disease entity; originally described by Tiedemann in 1818 and named by Ecker in 1864 [5, 6]; older studies have reported its prevalence as three of 20,000 autopsies and three of 24,519 surgical cases [7]. However, with the availability of new radiological modalities, the prevalence of AP has increased slightly and estimated to be about 1 case/1000 [8]. It affects both sexes with a slight male preponderance [9].

Embryologically; during the first four to eight weeks; the pancreas develops normally as a result of rotation and fusion of the dorsal and ventral pancreatic buds due to expansion of the duodenum. The ventral bud develops in the lower part of the head and the uncinate process of the pancreas, with the dorsal bud developing in the body and tail of the pancreas. The development of the AP is secondary to a migration abnormality resulting from the inability of the ventral bud to rotate and expand to envelop the second part of the duodenum partially or completely [10].

The diagnosis of annular pancreas is usually made in the neonatal period representing 1% of the causes of intestinal obstruction in children and will be responsible for non-bilious vomiting since the obstruction is supra-papillary [1, 3]. It is most often associated with other congenital malformations such as heart disease, Hirschsprung's disease and imperforate anus, tracheoesophageal fistulas, esophageal atresia, and chromosomal abnormalities, the most common of which is trisomy 21, then 18 and 13 [1, 11]. In adults, it is rare and appears between the ages of 20 and 50, most

often through duodenal stenosis. Other manifestations are pancreatitis, peptic ulcer disease, and obstructive jaundice; it may also remain asymptomatic in more than half of the cases [12].

The diagnosis of AP is usually made incidentally or during exploration of clinical manifestations, and can be made prenatally, preoperatively, or intraoperatively by many distinct invasive or noninvasive diagnostic techniques [13].

Useful diagnostic modalities for this condition in the prenatal or neonatal period include ultrasound or plain abdominal radiographs, which usually show the classic "double bubble" sign in duodenal obstruction and may be visible as early as the 2nd trimester of pregnancy [4, 13].

In adults, the diagnosis is usually made by computed tomography or magnetic resonance imaging. Other imaging techniques available to aid in the diagnosis of AP are magnetic resonance cholangiopancreatography, endoscopic retrograde cholangiopancreatography, or echo-endoscopy. More recent imaging techniques have been developed such as positron emission tomography (PET)/CT incorporating the radiotracers choline C-11 and fluciclovine F-18 and have contributed to the diagnosis of AP. Despite the many advances in diagnostic techniques, the gold standard test for the diagnosis of AP remains laparotomy, which has long been considered the "gold standard" as it allows a complete macroscopic examination of the duodenum and pancreatic head [13].

The therapeutic aspect is individualized according to the symptoms presented. Asymptomatic patients can be followed closely without surgery. When AP is associated with effective duodenal obstruction, the treatment of choice is surgery. This involves bypassing the duodenal stenosis by performing a gastroentero-anastomosis. Resection of the annular pancreatic tissue can also be performed, but has been associated with several complications; including pancreatitis, pancreatic fistula formation, and incomplete relief of the obstruction, as well as a lower rate of permanent cure [12]. Zyromski *et al*. [12] noted in their study that 20% of adult patients had complex pancreaticobiliary pathology associated with AP requiring surgical intervention corresponding to pancreaticoduodenectomy, lateral pancreatojejunostomy, biliary/pancreatic sphincteroplasty, and biliary system shunting [14].

The prognosis of annular pancreas in children is excellent, with increased overall survival rates, despite the presence of associated congenital malformations and chromosomal abnormalities, and in adults the prognosis is favorable if not complicated by underlying malignancy [13].

CONCLUSION

Although the incidence of AP in adults is very low, this diagnosis should not be ignored and should be suspected in the presence of chronic vomiting and other non-specific digestive presentations. In order to confirm this diagnosis, multiple imaging modalities are currently available to avoid surgical exploration. There are no specific guidelines or protocols for the management of AP, but if it is symptomatic a surgical bypass will be necessary.

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