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Case Report

Annular pancreas: Beneath the intestinal obstruction—A case report ☆

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ABSTRACT

Annular pancreas is an uncommon congenital anomaly which is a rare cause of congenital duodenal obstruction. It is normally identified during the neonatal period, but may also be identified in adolescence or adulthood. This diagnosis is often overlooked in adult patients who present with symptoms suggestive of duodenal obstruction. We present a case of AP detected in a 23-year-old man, with complaints of continuous vomiting and abdominal discomfort over the last 6 months. An upper gastrointestinal study revealed a constricted second part of the duodenum. A computed tomography scan revealed a complete ring of pancreatic tissue around the second part of the duodenum. Diagnostic and therapeutic surgery decompresses the external obstruction. The patient had an early post-operative activation. No specific guidelines and protocols exist about the management of such cases. Given the rarity of this congenital anomaly, presenting with chronic partial duodenal obstruction, and

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its successful surgical treatment, have prompted us to report the case along with a brief review of literature about the subject.

Introduction

The annular pancreas (AP) is termed as the formation of a thin rim of pancreatic parenchyma around the duodenum, usually, the descending or second portion, proselytizing into partial (75%) or complete (25%) obstruction of the bowel [1,2,4]. Yogi et al. [11] classified the annular pancreas into 6 sub-categories. Type 1, considered the most diverse, annular duct flows directly into the main pancreatic duct. The second type encircles the wirsung's duct and drains in the major papilla. The other 4 types are very rare. It remains one of the rarest congenital anomalies being 1 in 1000; 3 in 20,000 cases and 3 in 24,519 based on retrospective studies in different populations according to imaging, autopsy, and surgical cases, respectively [1–3]. It was 1st reported by Tiedemann and termed "Annular Pancreas" by Ecker [3–5]. It was 1905 when Vidal performed the first obstructive annular pancreas surgical treatment and the rarity of the condition was further supported by Vasconcelos and Sadek by reporting a single case in 22,243 autopsies [5]. Due to the paucity of cases, it is yet to establish whether there's any mendelian inheritance (autosomal or x-linked) in the pathogenesis of AP [2]. There are differences in the clinical presentation of AP with the variation in age, hence, it leads to variation in diagnosis and management [1–12].

Case presentation

In our case, we present a young man, 23 years of age. He arrives at the Gastroenterology Department with complaints of continuous vomiting and abdominal discomfort over the last 6 months. He recalls being always early satiated after his meals, even when they had been in small portions. When he attempts fasting, his symptoms are relieved to some extent. The vomiting has been non-projectile, non-biliary, and does not contain blood. On physical examination, the patient appeared slightly dehydrated, and the abdominal succussion splash maneuver is positive. Vital parameters: blood pressure 110/65 mmHg and regular pulse rate 98/ min. His weight was 72 kg, height was 1.75 cm, and BMI = 23.5. There was no alteration of complete blood count, basic metabolic panel, and there was no evidence of melena.

An abdominal ultrasound is performed, with insignificant findings of gastric dilatation. Consequently, the patient undergoes upper gastrointestinal endoscopy. The latter reveals a narrowing of the second portion of the duodenum, but it doesn't detect a clear cause of the narrowing. Hence, a CT scan was performed that confirms the diagnosis of the annular pancreas, encircling the second portion of the duodenum [Fig 1].

The surgical team was consulted and based on the imaging finding he was scheduled to undergo laparoscopic surgery. He was placed under general anesthesia and all aseptic precaution and preoperative surgical precaution were noted. Hasson method was followed and an 11 mm-sized camera port was inserted in the supra-umbilical region. Three 5 mm-sized trocars were placed on the right upper and left lower abdominal wall and below the xiphoid process followed by a 12 mm-sized port inserted in the right lower abdomen. Laparoscopic findings were confirmatory of the annular pancreas without additional abnormal findings. Initial step in the surgery was the truncal vagotomy followed by clearing of the greater curvature of the stomach from 10 cm above the pylorus to proximal 10 cm to proceed with gastrojejunostomy. To construct the anticolic gastrojejunal anastomosis, loop of the proximal jejunum, 50 cm distal to the ligament of Treitz, was employed. Then, a 60 mm linear stapler was used for the gastrojejunal anastomosis, then the enterotomy was closed laparoscopically in 2 layers with absorbable sutures. No surgical drain was placed. The patient had an early postoperative recovery and he could tolerate the progressively graded diet.. In the following days, he refers to the alleviation of the symptoms.

Discussion

The pancreas develops from an outgrowth of primitive foregut, ventral and dorsal buds in the fifth week of gestation [4,8]. By the seventh week of gestation, the ventral bud rotates clockwise and passes behind the duodenum to fuse with the dorsal bud. The ventral bud gives rise to the inferior part of both the uncinate process and head whereas the dorsal part forms the tail and the body of the pancreas [4,8]. There are myriads of hypotheses aiding the embryological abnormalities leading to the annular pancreas, among them Leeco's and Baldwin's theories have been mostly supported [2–5]. Leeco postulated due to the adherence of the tip of the ventral pancreatic bud to the duodenal wall and resulting in a ring duodenal encompassment due to duodenal rotation leading to annular pancreas formation, however, Baldwin reports the formation of the annular pancreas due to persistence of left pancreatic bud leading to complete encirclement of the duodenum by pancreatic tissues [2–5].

Genetic factors play a major role in the pathogenesis of the annular pancreas supported by the association of microduplication on chromosome 6q24.2 encircling the utrophin gene (UTRN) [1]. There are reported cases of the annular pancreas in both siblings and identical twins, also the mother and her progeny [1,2]. Moreover, other congenital anomalies, that is, trisomy 21 (down's Syndrome), cardiac defects, malrotation of the midgut, duodenal atresia, genitourinary anomalies, imperforate anus, and tracheoesophageal fistula are more prevalent

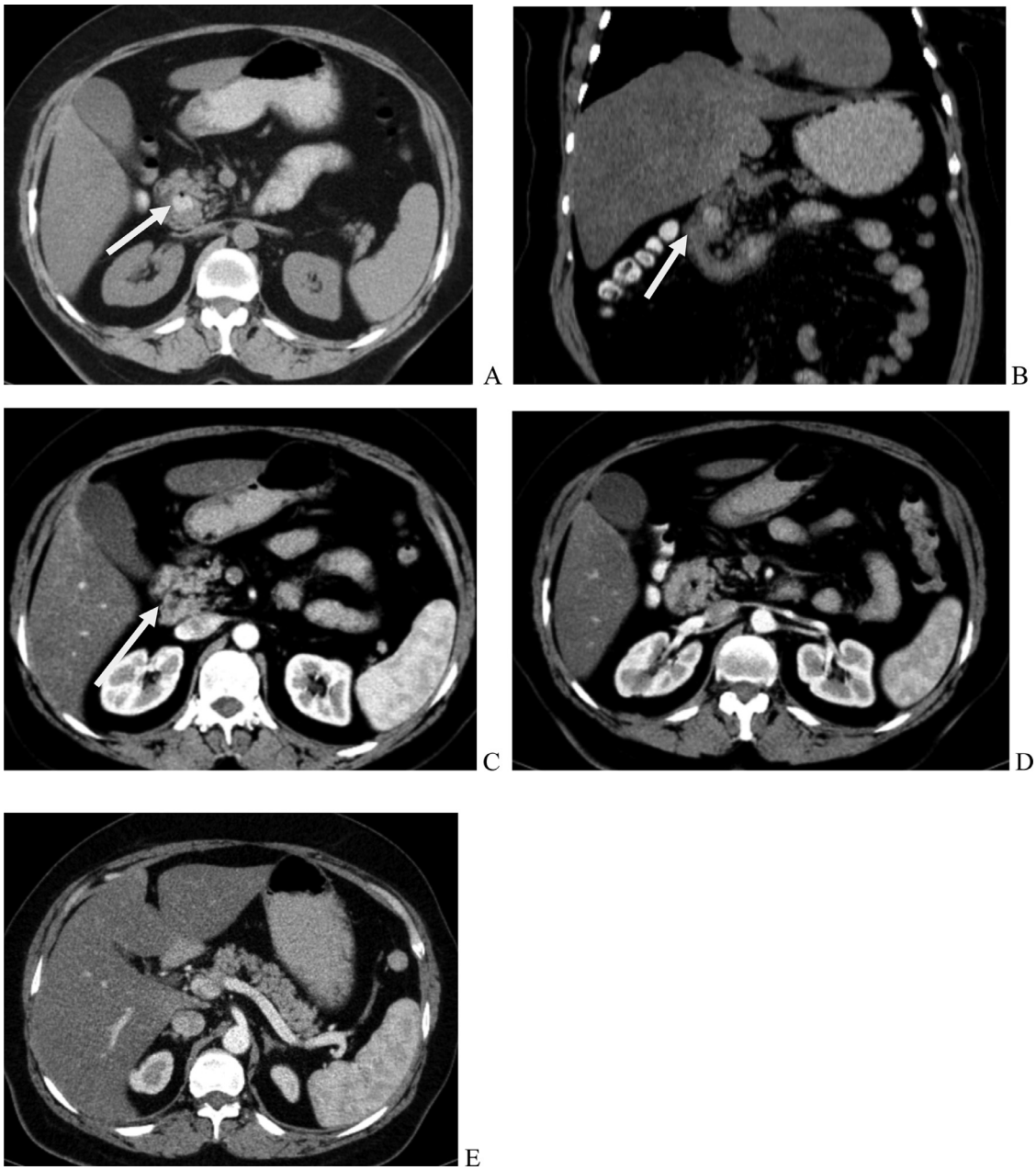

Fig. 1 – Abdominal CT axial images and coronary reconstruction with per os contrast (A and B) and with intravenous contrast (C and D) present the duodenum surrounded by isodense tissue with the pancreas. In images without i.v. contrast (A and B) notice the contrast applied per os in the duodenum which is surrounded by pancreatic tissue (A, B, white arrows). In the images after the application of contrast i.v. the choledochus with posterolateral position from the duodenum (C, arrow) and air in the duodenum (contrast for os is no longer presented) as well as the homogeneous amplification of the contrast by the pancreatic tissue is observed. There is also diffuse steatosis of the liver as well as another segmented pancreas without pathological changes (E).

in annular pancreas cases [1,4]. A mouse model with the defect of hedgehog signaling pathway-Sonic Hedgehog SHH and Indian hedgehog IHH the demonstrated of the annular pancreas in 85% of Shh−/− and 43% of Ihh−/− mutant mice playing a crucial role in developmental anomalies [1,2].

AP in utero presents mostly with maternal polyhydramnios due to intrinsic duodenal obstruction, whereas severe bilious vomiting presentation at a median age of 1 day after birth [2,7,10]. In neonates, more than two-thirds of AP will present with features of obstruction, that is, abdominal distention, bilious vomiting, decreased passage of meconium, and feeding intolerance [2]. Moreover, infants with annular pancreas are prone to have other congenital anomalies like Down's syndrome, esophageal and duodenal atresia, heart abnormalities, Meckel's diverticulum, pancreas divisum, and imperforate anus [2]. A plain radiograph showing double air demon-

strating duodenal obstruction that can be further evidenced by upper gastrointestinal series confirms the diagnosis of duodenal atresia; however, the etiology of it to ensure the proper treatment modality for annular pancreas is still a challenge [7]. The treatment for AP is surgical with duodenoduodenostomy being the most successful surgical option followed by division of a portion of the annular pancreas and gastrojejunostomy or duodenojejunostomy with Roux en Y loop [7].

It is established that between 50% and 75% of the annular pancreas cases remain asymptomatic until they reach the third to sixth decades and present with various complaints—peptic ulcer disease, duodenal obstruction, pancreatitis, obstructive jaundice, and extremely rare cases with pancreatic malignancy [1,8–10]. For the diagnosis of AP, the initial diagnostic modality can be ultrasonography or plain abdominal radiograph showing a double bubble sign in case of duodenal atresia [12]. CT or magnetic resonance (MR) finding shows pancreatic head enlargement with the enhancement of the duodenum second portion [9]. The classic crocodile jaw configuration is described as the presence of the pancreatic head anterior and posterior to the second part of the duodenum or the presence of pancreatic tissue posterolateral to the duodenum with clinical features correlation warrants the diagnosis [6,8,9]. MR is a better imaging modality compared to CT due to its high signal intensity on T1 fat-suppressed imaging showing pancreatic tissue surrounding the duodenum with similar findings on T2-weighted imaging including magnetic resonance cholangiopancreatography (MRCP) [6].

For the preoperative diagnosis of AP, endoscopic retrograde cholangiopancreatography(ERCP) is considered the gold standard despite the risk of acute pancreatitis or inaccessibility due to some sort of duodenal stenosis [10]. Around 37.5% of radiologically diagnosed partial AP happen to find a scanty rim of pancreatic tissue incorporated in the duodenal wall while performing ERCP [10]. However, despite the radiological advancement, in around 40% of the AP cases, surgical confirmation is needed making it gold stand as well [5,12]

The treatment option of AP differs according to its clinical presentation. While acute pancreatitis is managed symptomatically, surgical intervention with various bypass techniques (eg, duodenojejunostomy, gastrojejunostomy, and duodenoduodenostomy) is required to correct duodenal obstruction or gastric outlet obstruction [2,5,9,12]. Pancreatic resection is reserved for cases where suspicion of malignancy is unavoidable due to its adverse effects like pancreatitis, pancreatic fistula formation, or incomplete obstruction relief [5,12].

Conclusion

Through the review and examination of the 23-year-old male, a diagnosis of the annular pancreas was made. An annular pancreas is a rare abnormality that one is born with and occurs when extra pancreatic tissue forms around the small intestine. With abdominal ultrasound, we cannot make the diagnosis of an annular pancreas, but with a CT scan, we can confirm the diagnosis. Successful treatment of the annular pancreas can vary depending on the symptoms presented however a successful treatment can be through a duodenoduodenostomy. This is a surgical procedure where the portions of the duodenum before and after the obstruction are connected.

Patient consent

Informed consent has been obtained for the publication of this case report.

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