Unusual Clinical Presentation of Annular Pancreas in the Adult

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Abstract
Annular pancreas (AP) is a rare congenital anomaly, usually present in childhood, with symptoms due to duodenal obstruction; however, this condition can manifest in adulthood with abdominal pain, pancreatitis and pancreatic head mass. The authors present a case of AP observed in a 22-year-old patient that presented an unusual dual-phase clinical manifestation of duodenal obstruction in infancy that was treated by a duodenojejunostomy, and abdominal pain due to chronic pancreatitis in the adult age. MRI with cholangiopancreatography played a decisive role in achieving the correct diagnosis. The patient was treated by a pylorus-preserving Whipple procedure, with resection of the previous duodenojejunostomy. Pancreatic changes characteristic of chronic pancreatitis were demonstrated both in the AP and in the resected pancreatic segment. A marked biliopancreatic ductal anomaly not previously described in the literature was demonstrated by radiologic examination of the surgical specimen. The pathogenesis of AP, the importance of its association with benign and malignant pancreatic disease and the treatment alternatives are discussed by the authors.

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Annular pancreas (AP) is a congenital anomaly which consists of a ring of pancreatic tissue encircling the descending portion of the duodenum. This malformation once considered rare [1] has been reported with increasing frequency in the adult population [2]. In some reports, clinical manifestations appear after childhood in 50% of the cases [3].

Despite the congenital nature of the disease, the clinical manifestations may ensue at any age. Furthermore, it is estimated that two thirds of the patients remain asymptomatic for life [4]. In infancy or later in childhood, symptoms, when present, depend upon duodenal obstruction. In adulthood, clinical manifestations occur during the 3rd or 4th decades [5]. Presenting symptoms in adults are most frequently pain, vomiting, gastro-duodenal ulceration and pancreatitis [5]. Duodenal obstruction symptoms in infancy and clinical manifestations in adult age in the same patient have not been described in the literature.

We report herein a case of AP with a unique clinical presentation and an unusual anomaly of the malformed pancreatic ducts.
Case Report

A 22-year-old white man presented with a previous history of duodenal obstruction and vomiting due to AP at the 5th day of life when he was submitted to a successful duodenojejunostomy. He stayed asymptomatic until the age of 21 when he presented several episodes of postprandial epigastric pain with back irradiation and improvement in the genu pectoral position and intermittent vomiting. During the last year, he referred a 7-kg weight loss and 4 hospital admissions for pain and vomiting control. Upon admission in our unit, physical examination was unremarkable. The endoscopic evaluation of the upper gastrointestinal tract showed a dilated duodenal bulb with a wide anastomosis between the first duodenal portion and the jejunum. The minor papilla could be visualized next to the anastomosis. An upper gastrointestinal series successfully demonstrated the well-functioning duodenal-jejunum anastomosis. Abdominal CT and MRI scans demonstrated an enlargement of the head of the pancreas with dilated pancreatic ducts encircling and obstructing the duodenum (fig. 1). The $T_1$-weighted sequences revealed a decreased signal at the head of the pancreas, characteristic of chronic pancreatitis. Further evaluation of the pancreatic head and of the biliopancreatic ducts was done through MRCP. A dilated and arched anomalous pancreatic duct was identified at the head of the pancreas, confirming the previous diagnosis of AP. At the junction of the ventral and dorsal ducts there was a 10-mm length filling defect, interpreted as pancreatolithiasis. The body and tail of the pancreas presented a minimal ductal dilatation (fig. 2).

Due to the intensity of symptoms, operative treatment was indicated. On surgical exploration, a side-to-side duodenojejunostomy was found right upon the duodenum-encircling hard pancreatic tissue characteristic of chronic pancreatitis. After cholecystectomy, an intraoperative cholangiogram obtained through the cystic duct demonstrated common bile duct dilatation and a stop of the contrast medium within the pancreatic head. A pylorus-preserving pancreatoduodenectomy associated with the resection of the previously done duodenojejunostomy was performed; the reconstruction of the alimentary tract was accomplished by using a technique previously published [6].

The macroscopic inspection of the surgical specimen demonstrated pancreatic changes characteristic of chronic pancreatitis, both in the AP and in the resected pancreatic segment. No intraductal pancreatic calculi were found. A marked biliopancreatic ductal anomaly was demonstrated by radiologic examination of the surgical specimen after the injection of radio-opaque contrast media through the orifice of a pancreatic duct at the cut pancreatic surface (fig. 3). The dorsal pancreatic duct drained into a large and arcuated ventral duct which flowed, through an accessory pancreatic duct, into the common bile duct before the choledochoduodenal junction situated above the site of duodenal obstruction. The major papilla was located at the duodenum segment distal to the constriciting pancreatic ring (fig. 4).

Histologic study demonstrated chronic pancreatitis with acinar atrophy and moderate fibrosis present in the dorsal and ventral parts of the AP and in the segment of resected pancreas. Interlobular ducts, embedded in fibrous tissue, were focally ectasic and, in the dorsal and ventral parts of the AP, were occasionally lined by
Fig. 3. X-ray examination of the surgical specimen after the injection of radio-opaque contrast media through the orifice of a pancreatic duct at the cut pancreatic surface demonstrates a marked ductal anomaly. Filling defects seen in both pancreatic and common bile duct correspond to air bubbles.

Annular Pancreas

Discussion

With the improvements achieved in imaging techniques, the diagnosis of AP in adults increased in frequency [7], men being affected more commonly than women [4, 8].

Yogi et al. [8] classified AP in 6 types, depending on the site of drainage of the annular duct (table 1). The most common variety is type I, in which the annular duct flows directly into the main pancreatic duct. It is followed in frequency by type II, where Wirsung’s duct encircles the duodenum but still drains at the major papilla. The other 4 types are much less common. The marked biliopancreatic ductal anomaly demonstrated in the patient of the present report does not fit into any of these 6 types and,

squamous metaplastic epithelium. No protein plugs or calcifications were found. Both major and minor papillae were histologically normal. The islets of Langerhans in both the dorsal and ventral parts of the annulus were irregular and smaller than the round or ovoid islets present in the segment of resected pancreas. Immunostaining for insulin and glucagon demonstrated normal distribution.

An uncomplicated pancreatic fistula developed in the postoperative period and healed spontaneously before the hospital discharge at the 15th postoperative day. At the moment of this writing, the patient is free of symptoms after 15 months of follow-up.

Fig. 4. Diagram of ductal anomaly found in annular pancreas. AP = Annular pancreas; CBD = common bile duct; MP = major papilla; mp = minor papilla; MPD = main pancreatic duct.

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Fig. 4. Diagram of ductal anomaly found in annular pancreas. AP = Annular pancreas; CBD = common bile duct; MP = major papilla; mp = minor papilla; MPD = main pancreatic duct.
Table 1. Annular duct classification in annular pancreas [8]

<table>
<thead>
<tr>
<th>Type</th>
<th>Graphic representation</th>
<th>Opening site</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td><img src="image1.png" alt="Image" /></td>
<td>Wirsung's duct from dorsal site</td>
</tr>
<tr>
<td>II</td>
<td><img src="image2.png" alt="Image" /></td>
<td>Wirsung's duct encircling the duodenum</td>
</tr>
<tr>
<td>III</td>
<td><img src="image3.png" alt="Image" /></td>
<td>CBD from the dorsal site</td>
</tr>
<tr>
<td>IV</td>
<td><img src="image4.png" alt="Image" /></td>
<td>CBD without communication with Wirsung's duct</td>
</tr>
<tr>
<td>V</td>
<td><img src="image5.png" alt="Image" /></td>
<td>Santorini's duct from ventral site</td>
</tr>
<tr>
<td>VI</td>
<td><img src="image6.png" alt="Image" /></td>
<td>Santorini's duct from ventral site + pancreas divisum</td>
</tr>
</tbody>
</table>

CBD = Common bile duct.

to our knowledge, has not been previously described either.

There is still much controversy on the pathogenesis of AP. The two most important hypotheses concerning its formation are Lecco’s [9] and Baldwin’s [10] theories. Lecco postulated that adhesion of the distal tip of the ventral primordium to the duodenal wall, before its migration, originates the pancreatic obstructing ring [9], whereas Baldwin stated that persistence and further development of the left ventral bud is responsible for the formation of the annular pancreatic tissue around the duodenum [10]. Results of immunohistochemical analysis of pancreata with marked pancreatobiliary maljunction, like the one demonstrated in the present report, using an antipancreatic polypeptide antibody, suggest that the ring formation originates from the left lobe of paired ventral pancreata, supporting Baldwin’s hypothesis [11]. Although pancreatic polypeptide was not investigated in the present case, the similarity of physical characteristics of the islet cells in both the ventral and dorsal parts of the annulus also corroborates this hypothesis. However, the embryology of the AP is by no means elucidated yet, and other new hypotheses continue to be proposed [12, 13].

Clinical features of AP vary according to the time of symptom onset [2, 5]. In infants it is characterized by severe duodenal obstruction that requires immediate surgical intervention. On the other hand, in some cases, the obstruction may be of such minimal degree that the patient remains symptomless for life. When clinical manifestations ensue at adult age, symptoms include cramping epigastric pain, postprandial fullness and relief with vomiting [14]. Besides, peptic ulcer disease, acute and chronic pancreatitis, obstructive jaundice and gastric outlet obstruction may also be associated conditions in AP [15, 16]. A dual-phase clinical manifestation of AP in the same patient, combining duodenal obstruction in infancy and abdominal pain and vomiting due to chronic pancreatitis at adult age, as occurred in the patient of the present report, is most unusual and, to our knowledge, has not been previously published.

Pancreatic fibrosis due to chronic pancreatitis in AP is generally confined to the annulus and to the adjoining pancreatic head, preserving the body and tail of the gland [2, 16, 17]. Its pathogenesis is probably related to the impairment of pancreatic secretion flow through the annular duct [2, 14], while the maintenance of the secretion flow in the main pancreatic ductal system keeps the integrity of the body and tail of the pancreas [2, 14]. The only report on diffuse chronic pancreatitis associated with AP was an autopsy case of a patient with severe insulin-dependent diabetes that could possibly account for the features of chronic pancreatitis present in that patient [16].

The preoperative diagnosis of AP has evolved considerably due to the development of new diagnostic imaging tools. ERCP that has been considered the gold standard method in the diagnostic workup of AP [5, 8] is an invasive method associated with morbidity, including acute pancreatitis. Besides, ERCP may be unattainable due to subtotal duodenal stenosis determined by AP [18]. In the present case, ERCP, if feasible, would probably misdiagnose the pancreatic malformation that was confirmed by MRCP.

The main goal of surgical treatment of AP is the relief of duodenal or gastric outlet obstruction, and several procedures have been proposed with this intent [14]. Division or resection of the pancreatic annulus as used in the past [1, 2] is not advised because of a high incidence of duodenal leak, postoperative pancreatitis and pancreatic fistula [14]. Bypass surgery of the annulus by duodenostomy, gastrojejunostomy or duodenojejunostomy seems to be the preferred method of treatment [7, 14]. The latter procedure gave good results in the patient in this report for a long period of time.

Pancreatic resection for relief of duodenal obstruction in AP is rarely indicated [19]. Pancreateicoduodenectomy has been recommended when AP is associated with pancreatolithiasis [20], as initially suspected in the present
case. Only a few papers have been published on duodenopancreatectomy in patients with AP, most of them for treatment of associated neoplastic disease [18, 21, 22]. In a recent review of the English-language literature, AP was found concomitantly with 5 cases of ampullary carcinoma and 3 cases of pancreatic adenocarcinoma [18]. The differential diagnosis between focal inflammatory lesions in the head of the pancreas due to chronic pancreatitis and pancreatic cancer remains a challenging task for radiologists, pathologists and surgeons [18]. The association of AP and periampullary malignancy in adults must not be overlooked, and their coexistence must be considered until its absence is proved. Suspected cases belonging to population at risk for periampullary cancer should be treated by a pylorus-preserving pancreatoduodenectomy which can, nowadays, be performed with low morbidity and mortality rates.

References