Symptomatic annular pancreas in adult: Report of two different presentations and treatments and review of the literature

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1. Introduction

Annular pancreas (AP) is a rare congenital anomaly formed by a thin band of normal pancreatic tissue that completely or partially encircles the second portion of the duodenum and results in various degrees of obstruction [1–5]. Most cases are diagnosed early in life in patients presenting symptoms of gastric outlet obstruction [3]. In a few cases the condition may be found in adults where the symptoms are directly related to the degree of duodenal obstruction [1]. In some cases, the obstruction is not significant until inflammation of the annulus narrows the duodenum, in other cases the diagnosis is incidentally made when the patient is evaluated for symptoms of pancreatitis or peptic ulcer disease [6]. Usual symptoms in adults are abdominal pain in 70%, nausea and vomiting in 60%, and hematemesis in 10% of patients [6]. Up to 40% of cases requires surgery: duodenal-duodenostomy or duoden-duodeno-jejunal bypass. English language literature about annular pancreas etiology, diagnosis and treatment was reviewed.

No specific guidelines and protocols exist about management of AP, therefore, treatment and operative approaches must be individualized. In consideration of the possible post-operative complications, surgical treatment should be reserved in case of failure of conservative medical therapies.

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3. Results

A 53 year-old man went into ER for increasing epigastric abdominal pain and repeated vomiting during 5 h, without fever, bowel transit alteration and hematemesis. Upon the first clinical evaluation she was awake, clear headed and suffering; arterial blood pressure was 150/90 mmHg, heart rate 78 bpm rhythmic, blood O$_2$ saturation 100% in normal air. The abdomen was plain, painful at both superficial and deep palpation in the upper tract with positive Blumberg sign and peritoneal defense reaction, moderate enterocolic hypertympanism, and poor peristalsis. Thorax and heart examinations were negative, decile edemas were absent. A nasogastric tube was placed without significant stagnation. After 2 h the symptoms worsened. Abdominal ultrasonography showed distended gallbladder without stones, not dilated intra- and extra-biliary ducts, no free fluid, no relevant findings about liver, spleen, kidneys, bladder; the pancreas was not explorable. Abdominal plain radiography showed faecal stasis in the colon without pneumoperitoneum and significant hydro-air levels. Chest radiography and electrocardiogram were normal. Blood examination showed 19,100 white cells, 86.6% neutrophils, hepatic, pancreatic and renal functionality and electrolytes were normal. He was admitted in general surgery where she underwent fluid therapy, empiric antibiotic therapy and fasting. After 24 h he underwent abdominal contrast enhanced CT which showed solid pancreatic tissue surrounding the second portion of the duodenum, without obstructive findings (Fig. 1). Symptomatology progressively decreased during the following days. Five days later the bowel transit contrast radiography showed a regular passage of the contrast medium into the first digestive tract without stenosis and hydro-air levels. Upper endoscopy revealed chronic gastritis, duodenal bulbar deformation with a pseudo-diverticular sac (resulting from an old ulcer). Domperidone 20 mg was administered before the meal with benefit. The patient was discharged after 20 days in good conditions and without digestive symptoms.

A 71 year old man went into ER because of non specific abdominal pain and vomiting. Upon the first clinical evaluation he was awake, clear headed and suffering; arterial blood pressure was 135/85 mmHg, heart rate 65 bpm rhythmic, blood O$_2$ saturation 98% in normal air. The abdomen was globous, painful at deep palpation in the upper and middle tract with negative Blumberg sign, slightly positive Murphy sign, enterocolic hypertympanism, and valid peristalsis. Thorax and heart examinations were negative, decile edemas were absent. Blood examination, abdominal ultrasonography, chest radiography and electrocardiogram were normal. Abdominal plain radiography showed gastrectasia and faecal stasis in the colon without pneumoperitoneum and significant hydro-air levels. He was admitted in general surgery where he underwent contrast enhanced CT which showed sub-stenosant complete annular pancreatic solid tissue surrounding the inferior curve of the duodenal “C”, with edemic thickening of the mucosa, stasis of ingesta (like phytozeaors) in the second duodenal portion and dilatation of the first duodenal portion and of the stomach (Fig. 2). Upper endoscopy confirmed the stenosis of the second and third portion of the duodenum not passed by the conventional endoscope. Therefore, the patient underwent gastro-jejunal anastomosis, according to Roux technique, because of the impossibility to perform the duodeno-jejunal by-pass due to the extension of the stenosis. Post-operative course was uneventful and he was discharged on the 10th post-operative day in good condition and without digestive symptoms.

4. Discussion

AP is a rare congenital anomaly affecting approximately 1 in 20,000 newborns [10,11]. Tiedemann first reported this congenital anomaly in 1818 and it was named “annular pancreas” by Ecker in 1862 [1]. It is due to an embryologic migration fault and has been associated with other congenital anomalies, including Down’s syndrome, tracheoesophageal fistula, intestinal atresia, pancreas divisum and pancreaticobiliary malrotation [12]. The reported incidence in adults varies from 0.005 to 0.015% [13]. AP affects both sexes equally; however a recent review has found that symptomatic adult AP is mainly concentrated in males (71 males versus 42 females) [7].

The clinical presentation in most patients occurs between ages 20 and 60, with a particular concentration in the 30–50 year old-age range. In adult patients with AP common symptoms include cramping epigastric pain, post-prandial fullness and relief with vomiting. Associated conditions include peptic ulcer diseases, acute pancreatitis, pancreatic head carcinoma, biliary obstruction with jaundice and gastric outlet obstruction [2,7,13,14]. Pancreatitis as the initial disease expression is more commonly reported among adult patients than in children [15].

Useful diagnostic modalities for the diagnosis of AP include ultrasonography or plain abdominal radiographs, which usually show the classic “double bubble” sign when a duodenal obstruction is present. Surgical management is usually required to relieve the obstruction when the diagnosis of AP is confirmed operatively. Although surgery has been considered the “gold standard” for the diagnosis of annular pancreas [13–16], non operative modalities (CT scanning, MRI, MRCP, ERCP and EUS) may all suggest the presence of pancreatic tissue encircling the duodenum [17]. MRI and CT scanning have the advantage of being non invasive, although the correct diagnosis may be overlooked by both scan techniques if
the pancreatic tissue is present as a thin band incorporated in the duodenal wall [18].

When the AP is symptomatic and associated with an effective duodenal obstruction, the treatment of choice is the surgical procedure. The preferred treatment is a by-pass operation such as gastro-jejunalastomy or duodeno-jejunalastomy [19]. Resection of the annular pancreatic tissue may be also performed, on the other hand, has been associated with several complications including pancreatitis, pancreatic fistula formation, and incomplete relief of obstruction, as well as a lower rate of permanent cure [20].

5. Conclusion

Although the incidence of AP in adults is very low, its presence may be suspected in case of epigastric pain with unusual presentation. Advancements in imaging modalities allow to diagnose AP without the necessity of surgical exploration. No specific guidelines and protocols exist about management of AP, therefore, treatment and operative approaches must be individualized. In consideration of the possible post-operative complications, surgical treatment should be reserved in case of failure of conservative medical therapies.

Conflict of interest

All the authors declare that they have no conflict of interests.

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Ethical approval

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Author contribution

Fabio Rondelli: Participated substantially in conception, design, and execution of the study and in the analysis and interpretation of data; also participated substantially in the drafting of the manuscript.

Walter Bugiantella: Participated substantially in conception, design, and execution of the study and in the analysis and interpretation of data; also participated substantially in the editing of the manuscript.

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Nicola Avenia: Participated substantially in conception of the study and in the editing of the manuscript.
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