

## Case Report

# Annular Pancreas in a 24-Year-Old Female with Persistent Abdominal Pain

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### Abstract

Annular pancreas (AP) is a rare diagnosis in the adult population but can cause significant morbidity if not correctly identified. In adults, the most common symptoms are abdominal pain, nausea and vomiting. While these are not specific to AP, they are important clues to this diagnosis in the right clinical context. We present the case of a 24-year old female presenting with a 6-year history of progressive abdominal pain and dyspepsia in the context of an extensive negative workup. Upper gastrointestinal (GI) series and magnetic resonance imaging (MRI) revealed partial duodenal obstruction, concerning for AP. The patient was referred to surgery and underwent a laparoscopic duodenoduodenostomy confirming the diagnosis, with improvement of symptoms. While patients with chronic abdominal pain and vague gastrointestinal complaints may be diagnosed with functional bowel disorders, it is important to appropriately address the possibility of an underlying structural lesion such as AP. This strategy is not only cost-effective but also saves the patient discomfort associated with unnecessary procedures and allows for a timely intervention.

### Introduction

Annular pancreas (AP) is a rare congenital abnormality in which a ring of pancreatic tissue surrounds the descending portion of the duodenum causing duodenal obstruction. The exact prevalence of annular pancreas is difficult to determine as the presentation ranges with the degree of duodenal compression. As such, patients may present on a spectrum, ranging from complete intestinal obstruction in the neonatal period to completely asymptomatic [1, 2]. In adults, autopsy based prevalence studies reported annular pancreas in 5–15 cases per 100 000 adults [3]. Despite this, the incidence in adults remains low with estimates at 0.05 to 0.015% in the general population (4, 5). The pathogenesis involves embryological failure during the fourth to sixth week of gestation. Signaling cascade malfunction leads to failure of the pancreatic buds to

rotate and fuse appropriately, trapping the duodenum within pancreatic tissue [6]. AP is classically associated with other congenital disorders, such as Down's syndrome, intestinal malrotation, duodenal and esophageal atresia but these are more commonly seen in children [4, 7]. In adults, AP tends to be associated with pancreas divisum and pancreatobiliary neoplasia [7, 8].

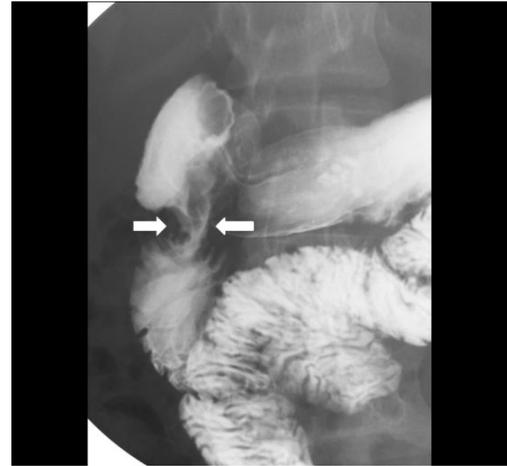
### Case Report

We report the case of a 24-year-old female who was referred for evaluation of a 6-year history of progressive abdominal pain. Her past medical history was significant for osteogenesis imperfecta with several traumatic fractures, esophagitis and gastritis. Her medications included acetaminophen, esomeprazole, and over the counter vitamins and antacids. On presentation, the patient

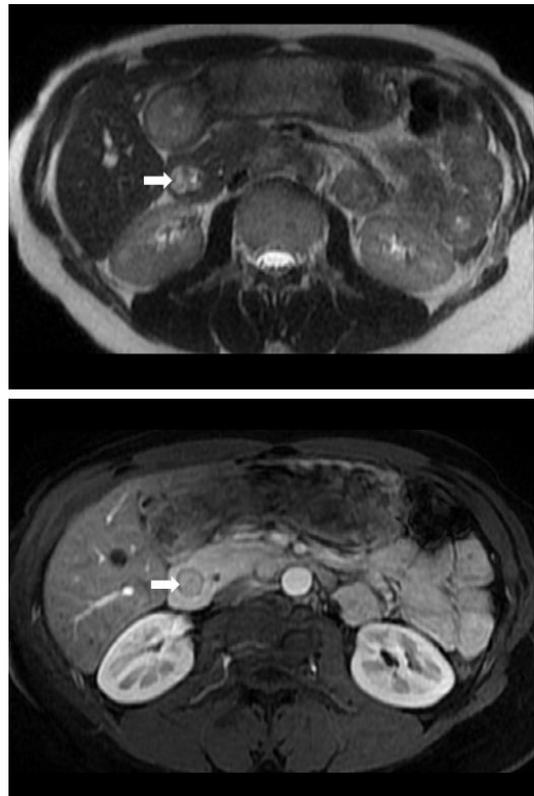
described pain located in the left upper quadrant of the abdomen (LUQ), which she characterized as intermittent and “sore-like” in nature. Eating full meals exacerbated this discomfort. She also noted a sensation as if her stomach was “not moving”, with frequent bloating. She denied all other symptoms such as nausea, vomiting, constipation, diarrhea and regurgitation. She added that the severity of the pain inhibited her sleep and also negatively impacted her ability to perform her professional duties during the day. She denied alcohol, drug and non-steroidal anti-inflammatory (NSAID) use. On examination, she was noted to be a well-developed female in no distress with normal vital signs. A complete physical examination was performed, only significant for mild tenderness to palpation of the LUQ.

The patient had recently undergone an extensive workup. This included standard blood work in addition to liver function tests, lipase, serum gastrin level, sedimentation rate, and autoimmune testing including celiac disease panel. All results were within normal limits. An abdominal computed tomography scan (CT) of abdomen and pelvis was also unremarkable. She underwent endoscopy, which was notable for mild mucosal erythema of the esophagus and stomach, with biopsy findings consistent with esophagitis and gastritis. Testing for celiac disease and *H. pylori* were negative. On colonoscopy, small bowel and colon were well-visualized and random biopsies revealed normal mucosa. The patient was started on proton-pump inhibitor therapy with little improvement in symptoms over one year. In light of this, esophageal manometry, a 24-hour pH study and a gastric emptying study were also performed prior to presentation. These studies were interpreted as negative, although the latter was not performed to standard as the patient refused solid food.

Given the extensive prior workup and persistent symptoms, functional bowel disorders were being considered highly on the differential. However, it was felt that extraluminal intestinal obstruction had not been adequately ruled out. Therefore, upper gastrointestinal (GI) series with small bowel follow-through was performed. The proximal duodenum remained narrowed throughout the series, suspicious for AP. This narrowing appeared associated with a delay in gastric emptying (Figure 1). Magnetic Resonance Imaging (MRI) of the abdomen with and without contrast showed the pancreas surrounding the cranial portion of the second portion of the duodenum causing focal narrowing consistent with AP (Figure 2 and Figure 3). The patient was referred to surgery for evaluation of likely AP. Upon confirmation on the diagnosis intraoperatively, a laparoscopic duodenoduodenostomy. Postoperatively, she reported tolerating regular food without further symptoms 6 weeks after surgery.



**Figure 1** - Upper gastrointestinal (GI) series demonstrates a fixed circumferential narrowing of the 2nd portion of the duodenum (arrow). The mucosal folds are preserved and this does not cause proximal obstruction.



**Figure 2 and Figure 3** - Axial T2 (Fig. 2) and contrast-enhanced fat-suppressed T1 weighted (Fig. 3) images demonstrate the duodenum to be completely surrounded by pancreatic parenchyma. Findings are suggestive of an annular pancreas (AP).

## Discussion

Annular pancreas (AP) is a rare diagnosis in the adult population but can cause significant morbidity if not promptly identified. Importantly, AP in adulthood can be found in both genders with similar frequency, with a mean age of diagnosis ranging from 20-60 years. In adults, the most common symptoms are abdominal pain (75%), vomiting (24%), pancreatitis (22%), or abnormal liver tests (11%), which may be seen in variable intensity depending on anatomical characteristics of the annular ring [7]. Patients with partial duodenal obstruction will commonly tolerate liquids and report bloating and distension with solids [5]. While these symptoms are not specific to AP, they are important clues to this diagnosis in the right clinical context. In the above case, the patient presented with many of the most common clinical manifestations of AP in adults.

In addition to the common symptoms, there is an association with AP and peptic ulcer disease (PUD) and gastritis, which often fails to resolve with standard therapy [5-9]. The mechanism is thought to result from functional gastric outlet obstruction leading to gastric stasis and antral distension, resulting in acid hypersecretion [3]. The patient in our reported case suffered from longstanding dyspepsia and endoscopically significant gastritis and esophagitis, likely stemming from their chronic partial duodenal obstruction. While there are many other causes of PUD and gastritis in the general population, refractory disease seen in the presence of other important symptoms raises AP on the differential diagnosis of dyspepsia.

The gold standard in diagnosis of AP is direct visualization of the annular ring, which can be achieved through laparoscopy [4]. However, imaging studies are more efficient and pose less risk. Modalities of choice include upper gastrointestinal (GI) series and CT scan. Magnetic resonance cholangiopancreatography (MRCP) may be useful when other studies are equivocal or if concomitant malignancy is suspected. It is important to note that 40% of diagnoses made via CT or upper GI series require surgery for confirmation of the diagnosis, as imaging modalities as still less sensitive [3]. When a lesion is associated with symptomatic duodenal obstruction, the treatment of choice is surgical bypass, such as gastrojejunostomy or duodenojejunostomy [10]. Resection of the annular pancreatic tissue has been associated with several complications such as fistula formation, pancreatitis or duodenal stenosis and is less frequently often performed [5]. Medical management has a limited role in symptomatic patients. In this case, CT scan was not able to identify the annular lesion, which resulted in MRI and upper GI series being pursued, with complete symptom resolution after surgical intervention.

Patients with chronic abdominal pain and vague gastrointestinal symptoms may pose a challenge for even the most experienced gastroenterologist. A routine diagnostic evaluation fails to reveal any diagnosis in up to 50% of affected patients, who are therefore said to have “functional bowel disorder” [11]. The differential diagnosis for these symptoms is broad and it is therefore paramount to perform a careful history and physical examination to guide effective diagnostic testing. While “functional bowel disorders” are being increasingly recognized and treated as a separate entity of disease in adults, it is first important to rule out structural lesions leading to chronic partial small bowel obstruction if the clinical context is appropriate. This strategy is not only cost-effective but also saves the patient considerable discomfort associated with unnecessary procedures and allows for timely and appropriate intervention.

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