

Annular Pancreas and Superior Mesenteric Artery Syndrome: Rare Etiologies and an Exceptional Association – A Case Report

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Abstract

Case Report

Annular pancreas and superior mesenteric artery syndrome are rare anatomical abnormalities, even more unusual when they coexist. This case report presents a 21-year-old patient suffering from recurrent acute pancreatitis and postprandial vomiting. Imaging revealed an annular pancreas encircling the duodenum, causing partial stenosis, and duodenal compression by the superior mesenteric artery (with an aorto-mesenteric angle reduced to 18°), characteristic of superior mesenteric artery syndrome [1, 2]. This association created a vicious cycle: duodenal obstruction and stasis promoted pancreatitis, while weight loss exacerbated mesenteric compression [3]. Initial treatment was medical (rehydration and parenteral nutrition), with surgical intervention (duodenojejunostomy) undertaken after failure of conservative measures. This case underscores the importance of a multidisciplinary approach to diagnose and manage these complex anomalies [4].

Keywords: Annular Pancreas, Superior Mesenteric Artery Syndrome, Acute Pancreatitis, Duodenal Obstruction.

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INTRODUCTION

Annular pancreas and superior mesenteric artery syndrome are two rare anatomical anomalies that can cause severe digestive symptoms.

Annular pancreas is a congenital anomaly caused by incomplete fusion of the ventral and dorsal pancreatic buds during embryonic life, resulting in a ring of pancreatic parenchyma encircling the second part of the duodenum, leading to duodenal stenosis. Imaging plays a key role in diagnosing this condition, with studies showing extrinsic, eccentric stenosis. Cross-sectional imaging techniques like computed tomography (CT) and magnetic resonance imaging (MRI) [5, 6] confirm the diagnosis by demonstrating pancreatic tissue encircling the second part of the duodenum.

Superior mesenteric artery (SMA) syndrome, or Wilkie's syndrome, is a rare condition caused by extrinsic compression of the third part of the duodenum between the superior mesenteric artery and the aorta, due to a reduced angle between these structures. Its diagnosis relies on clinical and radiological findings [7, 8].

Their coexistence is even more unusual and represents a diagnostic and therapeutic challenge. We present the case of a 21-year-old patient, with no significant medical history, admitted for recurrent acute pancreatitis and food-induced vomiting.

CASE PRESENTATION

The patient was a 21-year-old woman with no notable medical history, presenting with epigastric pain and intermittent postprandial vomiting that progressively worsened over three years. Her symptoms escalated in the nine months preceding her consultation, marked by acute epigastric pain, three episodes of mild hematemesis, and weight loss of 10 kg.

On physical examination, the patient was conscious, hemodynamically and respiratory stable, with pale conjunctiva and a body mass index (BMI) of 16 kg/m². Abdominal examination revealed epigastric tenderness, gastric splash, and peri-umbilical tympanism. Laboratory findings showed hypochromic microcytic anemia (Hb = 11 g/dL, MCV = 71 fL, MCHC = 29 g/dL) and mild lipase elevation consistent with acute pancreatitis (three episodes in total).

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Esophagogastroduodenoscopy (EGD) revealed erythematous pangastritis with normal duodenal mucosa. Gastric biopsies showed non-specific subacute gastritis.

Subsequent abdominal CT scan revealed anterior-lateral pancreatic parenchymal extension surrounding the second part of the duodenum, suggesting an incomplete annular pancreas. MRI confirmed mild hypertrophy of the pancreatic head, with no signal abnormalities, encircling the second part of the duodenum without compression, consistent with an incomplete annular pancreas. It also identified extrinsic compression of the third part of the duodenum between the superior mesenteric artery and the aorta, indicative of SMA syndrome.

The final diagnosis of incomplete annular pancreas and SMA syndrome was established. Symptomatic treatment failed, and a multidisciplinary team recommended surgical intervention. The patient underwent duodenojejunostomy.

Imaging Findings

- **Abdominal CT with 3D reconstruction:** Demonstrated an annular pancreas encircling the second part of the duodenum, causing partial stenosis [9, 10].
- **Abdominal Angio-CT:** Confirmed severe compression of the third part of the duodenum by the superior mesenteric artery, with an aorto-mesenteric angle reduced to 18° (normal $> 25^\circ$), consistent with SMA syndrome.
- **MRI:**
 - Mild hypertrophy of the pancreatic head, partially encircling the second part of the duodenum without compression. MRI findings were compatible with an incomplete annular pancreas (Figure 1).
 - Extrinsic compression of the third part of the duodenum (D3) due to SMA syndrome, explaining the clinical symptoms and requiring further CT evaluation (Figure 2).

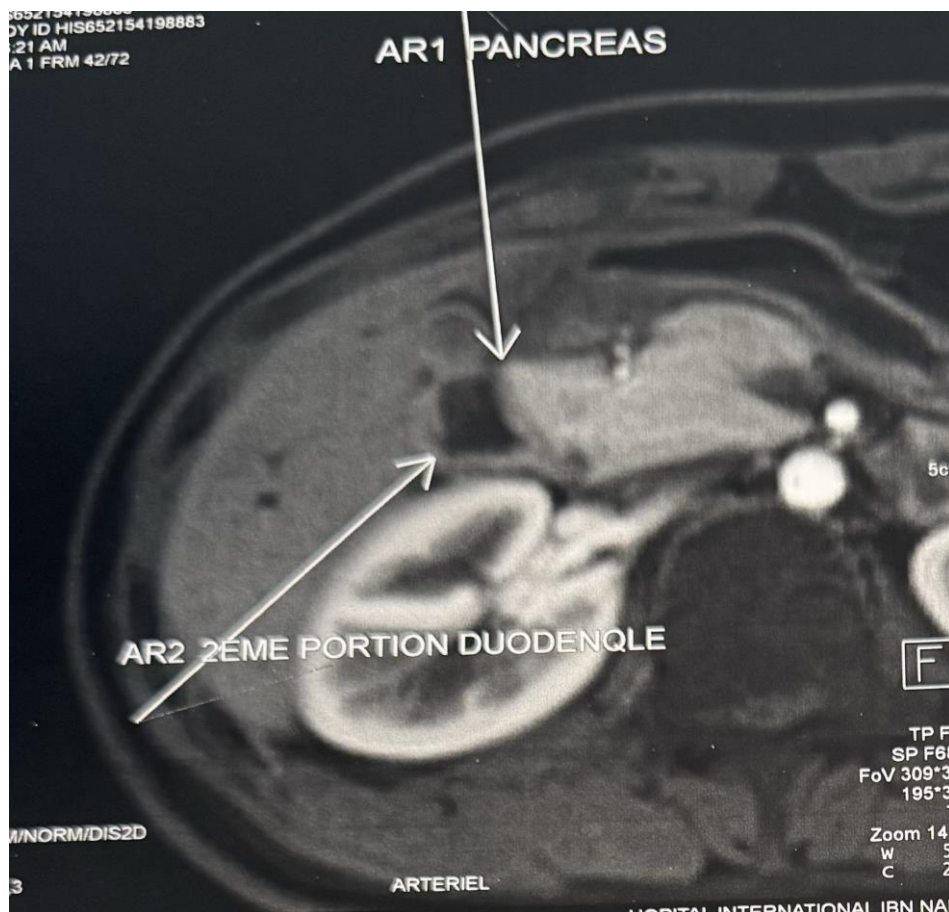


Figure 1: MRI biliary sequence showing a slight hypertrophy of its cephalic portion without associated signal abnormalities on the various sequences performed, extending anterolaterally to the second part of the duodenum, which it partially surrounds without signs of compression

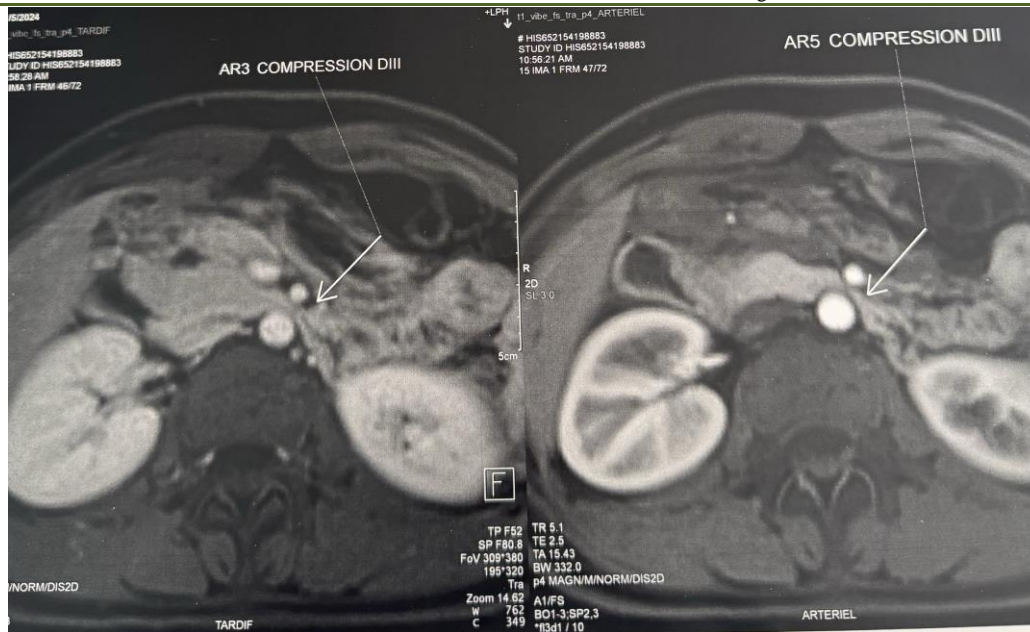


Figure 2: MRI biliary sequence showing compression of the third portion of the duodenum between the superior mesenteric artery and the aortovertebral plane

Final Diagnosis

The coexistence of annular pancreas and superior mesenteric artery syndrome was identified as the cause of recurrent acute pancreatitis and vomiting.

DISCUSSION

Annular Pancreas

Annular pancreas is a rare congenital anomaly caused by incomplete fusion of embryonic pancreatic buds. It may be asymptomatic or cause duodenal obstruction, abdominal pain, or recurrent acute pancreatitis due to duodenal stasis and increased pancreatic ductal pressure [1, 3].

Superior Mesenteric Artery Syndrome

SMA syndrome results from duodenal compression by the superior mesenteric artery, often exacerbated by rapid weight loss [7, 9]. Symptoms include postprandial vomiting, weight loss, and postprandial abdominal pain.

Relationship Between the Two Anomalies

In this case, the coexistence of annular pancreas and SMA syndrome created a vicious cycle [8]:

- Mechanical obstruction exacerbated duodenal stasis, promoting pancreatitis.
- Weight loss secondary to vomiting amplified mesenteric compression.

Management

Initial management focused on medical stabilization:

- Intravenous rehydration.
- Parenteral nutrition to interrupt the vicious cycle of malnutrition and compression.

Surgical intervention was required after conservative treatments failed [11, 12]:

- Duodenojejunostomy was performed to bypass the compression site, with significant clinical and biological improvement. Partial resection of the annular pancreas was not necessary.

CONCLUSION

This case highlights the diagnostic and therapeutic complexity of congenital anatomical anomalies when they coexist. A multidisciplinary approach is crucial to improve prognosis and prevent complications. Advanced imaging and collaborative management are essential in managing rare, complex anatomical anomalies.

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