

# Duodenal carcinoma in a patient with an annular pancreas: rarity personified

## Abstract

It is fascinating how organs live longer than man. This simple fact makes it possible for an organ to be a resource for society, a valuable gift that can be returned once it has fulfilled its role for the current bearer. Facilitation of transference of one organ from its original bearer (donor) to the new recipient has been matured and manicured into a well oiled process over the last half a century (Organ donation, Organ harvest, Organ preservation, Organ allocation and Organ Implant – Five key steps in cadaveric organ transplantation). It all starts with a simple will of a patient or family to donate the organs of their loved ones once they are no more. In the Indian context, cadaveric organ transplantation conceptualized over the past decade and prolifically prospered over the last five years or so in the western India. His in some instances has led to enthusiasm without knowledge where in there has been propaganda to promote organ donation without preparation of facilities to honor this commitment when the patients or the families come forward. As always result these activities get limited to a promise of a donor card! Any organ has to be seen as a resource of the country and drive should be prolific throughout the state. All the institutions with ICU facilities should be approached as stake holders in the process of organ donation and should be designated retrieval centers. The multi organ transplant centers should take the responsibility of organ harvest in these centers irrespective of whether the organ is being used in their centre or not.

**Keywords:** primary duodenal carcinomas, non-specific signs, symptoms, gastrointestinal, endoscopy, dull, non-radiating, furthermore, fever, soft, duodenum, differentiated, pt3, n0, m0, occur at any age, minimal or absent

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**Abbreviations:** MSCT, multi-slice computed tomography; MRI, magnetic resonance imaging; MRCP, magnetic resonance cholangio-pancreatography; ERCP, endoscopic retrograde cholangio-pancreatography

## Introduction

Annular pancreas (AP) is a rare congenital anomaly, which consists of a ring of pancreatic tissue, confluent with the head of the pancreas, partially or completely encircling the second segment of the duodenum. It was first described by Tiedemann<sup>1</sup> and first named “pancreas anulare” by Ecker.<sup>2</sup>

The estimated incidence of AP, with the diagnosis made on autopsy or during surgery, is 3 in 20000<sup>3</sup> However, when the diagnosis is based on modern imaging methods, such as multi-slice computed tomography (MSCT), magnetic resonance imaging (MRI), magnetic resonance cholangio-pancreatography (MRCP) or endoscopic retrograde cholangio-pancreatography (ERCP), the incidence seems to be higher, up to, 1 in 250.<sup>3,4</sup> About 700 cases of AP have been reported in the literature.<sup>5</sup>

Although the small intestine constitutes 75% of the gastrointestinal tract, tumors arising from it are rare. Small intestine tumors account for about 5% of all alimentary tract tumors and the duodenum has a higher proportion of these tumors than the jejunum and ileum.<sup>6</sup> Primary Duodenal carcinomas (PDC) are rare and account for 20%-25% of all small bowel malignancies, whereas sarcomas, carcinoid and lymphomas<sup>7,8</sup> are even more less common. A PDC is difficult to diagnose because of its rarity, non-specific signs and symptoms and it inherently being overlooked during upper gastrointestinal endoscopy.

Association of AP with malignancy is also rare. It has been described 14 times in the medical archives (five pancreatic carcinomas, six ampullary carcinomas, two cholangiocarcinomas, and one in sulinoma).<sup>9-20</sup>

A review of 151 cases of annular pancreas in Japan by Ladd et al.<sup>11</sup> revealed an association with malignant tumors of the duodeno-pancreato-biliary system in 15 patients (five cholangiocarcinomas, four gallbladder carcinomas, four duodenal, and two pancreatic carcinomas). Herein we report a case of AP associated with a PDC, where both the rarities presented in a single patient.

## Case report

A 52year-old woman had a short, (2weeks), history of persistent postprandial nausea and occasional vomiting containing food particles. She also demonstrated loss of appetite and significant weight loss of 7kg (15% of body weight) in a month with a prolonged history (3month-2 years) of epigastric pain that would respond to proton pump inhibitors (PPI).

The pain was dull, non-radiating, had no particular pattern or association with food. Furthermore, she did not have a history of fever, hematemesis/melena, diarrhoea or jaundice.

On clinical examination, the patient appeared alert, hemodynamically stable, but dehydrated (with a low JVP). Her abdomen was soft, non tender with no obvious palpable masses. Laboratory results revealed a mild hypokalemia of 3.2mmol/L, and a metabolic alkalosis with an initial pH of 7.65. Liver function tests, renal function tests and serum amylase were normal.

Gastroscopy revealed a proliferative growth on the lateral wall of the second part of the duodenum with a normal visualized part of ampulla. An endoscopic biopsy was done that was reported as an adenocarcinoma of the duodenum.

Abdominal MSCT confirmed a 3x3cm growth in the lateral wall of the duodenum that was encircled by an AP. There was no evidence of dilatation of the main pancreatic duct or the common bile duct.

Furthermore there was no evidence of significant peripancreatic or interaortocaval lymph nodes, as cists or intraperitoneal deposits. Surgery, confirmed an AP with a hard mass in the lateral wall of the 2nd part the duodenum. There was no evidence of extra duodenal spread or as cists.

Pancreaticoduodenectomy was done as described by Whipple (ref). A feeding jejunostomy was inserted. The tumor was clearly located in the duodenum, No tumor was observed in the head of the pancreas. Pathological examination showed a poorly differentiated, infiltrating adenocarcinoma of the duodenum, surrounded by the incomplete annular pancreas. All 7 lymph nodes were reactive. The tumor stage was pT3, N0, M0.

## Discussion

Clinical manifestation of AP in adults can present with cramping epigastric pain, postprandial fullness, vomiting, and weight loss.<sup>7,8,21</sup> Furthermore they can be associated with pancreatitis, or gastric/duodenal ulcers.<sup>5,7</sup> It can occur at any age, from neonatal period to adulthood however when the duodenal constriction is minimal or absent, AP can remain asymptomatic for lifelong.<sup>22</sup>

The most frequent symptom in adult AP seems to be abdominal pain.<sup>5,21</sup> Most adults become symptomatic between the ages of 20 and 50.<sup>7</sup> The differential diagnosis includes other upper gastrointestinal pathologies, such as peptic ulcer disease, duodenal web, pancreatitis, or pancreatic carcinoma.<sup>6,17</sup> Obstructive jaundice is described as a rare direct result of AP but usually appears in coexisting periampullary malignancies.<sup>17</sup> The most common symptoms of PDC's are also abdominal pain, but in addition they would have significant weight loss.<sup>23</sup> These two signs, should be investigated with a combination of imaging and ERCP.

AP in today's advanced imaging world is increasingly being diagnosed by non-invasive methods like MRCP and CT.<sup>23</sup> However, a recent study reviewed 55 cases with AP in adults,<sup>5</sup> where the diagnosis was made with ERCP in 47%, MSCT in 18%, with MRCP in 16%, and in 13% of the patients, at the time of surgery.

We were able to make the diagnosis of an AP non-invasively using an abdominal MSCT but an upper gastrointestinal tract endoscopy was crucial in clinching the diagnosis of the PDC.<sup>23</sup> The combination of both these modalities of investigations, led us to believe that, the duodenal obstruction, was caused by the PDC rather than AP.

## Conclusion

AP is a rare malformation that manifests itself primarily by signs related to duodenal stenosis. Presence of upper gastrointestinal obstruction with annular pancreas without obstructive jaundice in elderly patients should not distract from possible co-existing PDC. The diagnosis is currently based on a multi-modality approach with abdominal CT scan and upper gastrointestinal endoscopy and biopsy.<sup>24-28</sup>

Treatment is exclusively surgical with a by-pass procedure for AP with benign obstruction. On the other hand, if endoscopy and biopsy confirms a PDC, the only option is a pancreaticoduodenectomy in patients with a resectable tumour. This case highlights the rarity of an AP with coexisting PDC, its diagnostic algorithm and treatment options.

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## Conflict of interest

Author declares that there is no conflict of interest.

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