

CASE REPORT

Ampullary Carcinoma Associated with an Annular Pancreas

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ABSTRACT

Context Annular pancreas is an uncommon congenital abnormality. Co-existence of this condition with a pancreaticobiliary malignancy is an exceptionally rare occurrence.

Case report We present a case report of a 78-year-old woman with jaundice due to an ampullary carcinoma associated with an annular pancreas treated by pancreaticoduodenectomy.

Conclusions A collection of previously reported cases is reviewed together with the relevant literature. Obstructive jaundice is an uncommon feature of annular pancreas; hence the possibility of co-existent pancreaticobiliary malignancy should be excluded.

pancreaticobiliary malignancy is therefore exceptionally rare and hence only few cases have been reported. We report a case of ampullary adenocarcinoma in a patient with an annular pancreas.

CASE REPORT

A 78-year-old woman presented with epigastric pain radiating into her back, made worse on eating. She had no significant comorbidity. Laboratory investigations were unremarkable apart from abnormal liver functions with raised total bilirubin (67 $\mu\text{mol/L}$; reference range: 5-21 $\mu\text{mol/L}$), ALT (42 IU/L; reference range: 0-40 IU/L) and alkaline phosphatases (719 IU/L; reference range: 70-300 IU/L). Tumour markers CEA and CA 19-9 were normal.

INTRODUCTION

Annular pancreas was first described by Tiedemann in 1818 and is due to malrotation of the pancreatic ventral bud during embryological development [1]. Popular theories explaining the pathogenesis include adhesion of the right ventral bud to the anterior duodenal wall [2] and failure of the left ventral bud to involute [3]. This congenital anomaly is a rare occurrence. In 20,000 autopsies only three were discovered [4], while a review of 22,243 surgical cases demonstrated only a single one [5]. Co-existence of annular pancreas with



Figure 1. CT scan image demonstrating significant dilation of the common bile duct (black arrows).



Figure 2. CT scan image revealing a soft tissue mass representing the ampullary carcinoma (white arrows).

ERCP revealed an enlarged and protuberant ampulla of Vater. This was biopsied and reported as tubulo-villous adenoma with high grade dysplasia. An abdominal CT scan demonstrated marked intra- and extrahepatic bile duct dilation. The common bile duct was dilated to 2 cm (Figure 1) down to the level of the ampulla where a 2 cm tumour was noted (Figure 2). There was no evidence of distant disease or contraindications to a curative resection.

The patient underwent a pancreaticoduodenectomy. Findings were of a complete annular pancreas wrapping around the duodenum 5 cm distal to the pyloric sphincter. There was a palpable tumour in the ampullary region. The resection was

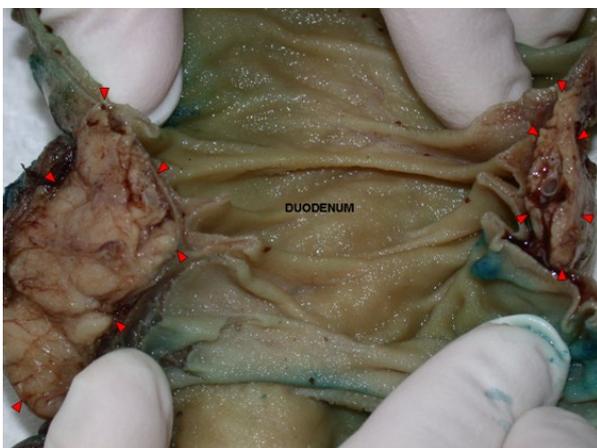


Figure 3. Anterior aspect of duodenum and the annular segment of pancreas (red arrowheads) divided longitudinally.



Figure 4. Cross sectional view showing tumour within the ampulla (white arrowheads). The duodenum is surrounded by the annular (red arrowheads) and non-annular (blue arrowheads) segments of the pancreas.

performed without any complications and after an uneventful recovery the patient was discharged home on the tenth post operative day.

Gross pathological examination confirmed the presence of a complete annular pancreas with pancreatic parenchyma wrapping around and narrowing the duodenum over a length of 2.5 cm (Figure 3). With the exception of minimal pancreatic duct dilatation, histology of the annular pancreas was normal and there was no evidence of inflammation or pancreas divisum. The ampulla of Vater was occupied by a 1.2x3.5 cm polypous tumour (Figure 4). Histology demonstrated a papillary adenoma of intestinal type showing severe dysplasia. In addition, there was a focus of well differentiated adenocarcinoma, which was confined to the fibromuscular layer of the ampulla. The tumour stage was pT1 N0 V0 R0 and hence no adjuvant treatment was indicated.

DISCUSSION

Annular pancreas has a bimodal pattern of presentation, the first peak is in infancy and a later peak occurs in the fourth decade of life. The proportion of patients treated for annular pancreas is split equally between the two age groups. However, both have very different clinical pictures. Paediatric cases usually present with gastrointestinal obstruction or

jaundice. Associated conditions include mongolism, intestinal malrotations, duodenal atresia/stenosis/webs, tracheoesophageal fistulas and cardiac defects [6].

Presentation in adulthood is rare, although annular pancreas is now identified with increasing frequency because of more liberal use of abdominal CT, ERCP and MRCP. Adult annular pancreas is associated with duodenal obstruction (60%) [6], pancreatitis (15-50%) and peptic ulceration (26-48%) [7]. Obstructive jaundice is an uncommon feature and not usually a direct result of the annular pancreas. Morrell and Keynes reported on 15 annular pancreas patients with jaundice and causes included pancreatitis (10 cases), choledocholithiasis (4 cases) and alcoholic liver disease (1 case) [8]. A series of 135 adult annular pancreas cases revealed only one case with biliary obstruction due to the pancreatic annulus [6].

Later work in the Japanese literature started to elude attention to co-existent pancreatico-

biliary malignancy as a cause of the obstructive jaundice or as an association in annular pancreas patients. A review by Yogi *et al.* of 105 annular pancreas patients revealed nine with obstructive jaundice due to ampullary carcinoma (5 cases), pancreatic carcinoma (2 cases) and cholangiocarcinoma (2 cases) [9]. A subsequent collection of 151 cases of annular pancreas by Ogawa *et al.* revealed 15 had pancreaticobiliary malignancy, including 5 cholangiocarcinomas, 4 gallbladder carcinomas, 4 duodenal and 2 pancreatic carcinomas [10]. Hence, clinical presentation of annular pancreas with obstructive jaundice is rare and should alert the clinician to the possibility of another cause such as underlying malignancy. Including ours, there are 14 cases of carcinoma associated with annular pancreas in the English literature (Table 1) with the first published in 1982 [10, 11, 12, 13, 14, 15, 16, 17, 18, 19, 20, 21]. There were eight female and six male patients with ages ranging from

Table 1. Summary of pancreaticobiliary malignancy cases associated with annular pancreas.

Author	Age (years) and sex	Presenting features	Surgical treatment	Pathology
Transveldt <i>et al.</i> , 1982 [11]	80 F	Jaundice, weight loss	PD	Ampullary adenocarcinoma
Matsusue <i>et al.</i> , 1984 [12]	53 F	Weight loss, abdominal pain	TP	Pancreatic head carcinoma
Schlinkert <i>et al.</i> , 1990 [13]	45 F	Recurrent hypoglycaemia	Insulinoma enucleation	Insulinoma
Yasui <i>et al.</i> , 1995 [14]	54 M	Jaundice, vomiting, pancreatitis	PPPD	Pancreatic head adenocarcinoma
Kamisawa <i>et al.</i> , 1995 [15]	79 F	Abdominal pain and mass	None	Pancreatic body adenocarcinoma ^a
Benger, Thompson, 1997 [16]	66 M	Jaundice, weight loss, anorexia	PD	Ampullary adenocarcinoma
Rathnaraj <i>et al.</i> , 1998 [17]	55 M	Jaundice	PD	Ampullary adenocarcinoma
Ogawa <i>et al.</i> , 2000 [10]	48 M	Jaundice, abdominal pain	PPPD	Cholangiocarcinoma
Shan <i>et al.</i> , 2002 [18]	40 F	Jaundice, abdominal pain, distension, vomiting	PD	Ampullary adenocarcinoma
Shan <i>et al.</i> , 2002 [18]	45 M	Jaundice, abdominal pain	PPPD	Ampullary adenocarcinoma
Yamaguchi <i>et al.</i> , 2003 [19]	64 M	Jaundice	PPPD	Cholangiocarcinoma
Ben-David <i>et al.</i> , 2004 [20]	52 F	Abdominal pain	TP	Diffuse pancreatic adenocarcinoma ^a
Cholet <i>et al.</i> , 2004 [21]	88 F	Jaundice, weight loss	None	Pancreatic head carcinoma ^a
Foo <i>et al.</i> , 2007 (Present study)	78 F	Jaundice, abdominal pain	PD	Ampullary adenocarcinoma

^a Denotes pathology occurred in the non-annular/dorsal segment of the pancreas

PD: pancreaticoduodenectomy

PPPD: pylorus preserving pancreaticoduodenectomy

TP: total pancreatectomy

40 to 88 years. Seven of the 14 patients were of oriental ethnicity. The most common presenting symptoms were jaundice (11 cases), upper abdominal pain (7 cases) and weight loss (4 cases).

Biochemical investigations revealed jaundice and deranged liver function tests in 85% of patients. Tumour markers were poor indicators with positive predictive values of only 50% and 57% of the time for CEA and CA 19.9 respectively.

Annular pancreas was diagnosed pre-operatively in 6 cases mostly by means of ERCP. CT was poorly sensitive and failed to detect annular pancreas in 10 out of 13 times performed. Diagnosis of annular pancreas on CT and MRI is obtained by visualisation of pancreatic duct and parenchyma surrounding the duodenum. However, this is difficult without adequate thickness of the pancreatic parenchyma. ERCP is more sensitive, providing a definitive diagnosis by visualisation of the pancreatic duct encircling the duodenum, but this investigation has attributed risks. MRCP and endoscopic ultrasound have been found to be less invasive alternatives which can also diagnose a co-existent malignancy [19].

Amongst the cases reported in the English literature, the most commonly occurring cancer in the annular pancreas was ampullary carcinoma (6 cases). Others included pancreatic head carcinoma (3 cases), cholangiocarcinoma (2 cases), pancreatic body adenocarcinoma (1 case), diffuse pancreatic carcinoma (1 case) and insulinoma (1 case) [10, 11, 12, 13, 14, 15, 16, 17, 18, 19, 20, 21]. The malignancy usually involved the annular (ventral) segment of the pancreas. This has been attributed to chronic inflammatory changes in this area associated with pancreatitis. There were only three reports of cancer occurring in the non-annular (dorsal) segment [15, 20, 21]. Surgical treatment of pancreaticobiliary malignancy in the annular pancreas was similar to that in the non-annular pancreas.

Annular pancreas presenting with obstructive jaundice is a rare occurrence. The possibility of a co-existent underlying malignancy of the

pancreaticobiliary system should always be considered in the management of the patient.

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Keywords Adenocarcinoma; Ampulla of Vater; Jaundice, Obstructive; Pancreas /abnormalities, /embryology; Pancreatic Neoplasms

Abbreviations PD: pancreaticoduodenectomy; PPPD: pylorus preserving pancreaticoduodenectomy, TP: total pancreatectomy

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