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Case Report

Annular Pancreas Presenting with Painless Jaundice: A Case Report and Review of Literature

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ABSTRACT

Annular pancreas (AP) is a rare congenital abnormality in which pancreatic tissue completely surround the duodenum. Presentation of AP in adulthood is very rare and obstructive jaundice is one of the uncommon manifestations of AP in adults. Herein; we reported a 47 years old woman who presented with icterus, mid common bile duct stricture and annular pancreas. She was diagnosed as pancreas cancer three months after surgery. Although AP presenting with jaundice is rare but is indicative of significant relationship with periampullary malignancies and requires a complete investigation of these cancers and close follow up.

Keywords: Annular Pancreas; Obstructive Jaundice; periampullary malignancy

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INTRODUCTION

AP is a rare congenital abnormality in which the ventral bud of the pancreas divides into right and left parts which they enclose completely the duodenum and form AP(1).

The majority of cases were among neonate and children. The most prevalent symptoms in childhood are nausea and vomiting related to gastric outlet obstruction. This abnormality remains asymptomatic most of the time until adulthood. Features of AP in adulthood include; abdominal pain, nausea and vomiting(2,3), pancreatitis, peptic ulceration(4), pancreatic hyperenzymemia, and rarely obstructive jaundice or malignancy(2). Jaundice was a rare manifestation of AP. According to the literature many of these cases had concurrent periampullary malignancies such as carcinoma of vater ampulla or pancreatic adenocarcinoma(2- 4).

We presented here a 47 year old woman who had concomitant AP and pancreatic adenocarcinoma.

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CASE REPORT

A 47years old woman was admitted to the hospital because of recent onset jaundice and pruritus. The patient had been well until two weeks ago when icterus gradually developed. She also reported discoloration of stool and dark urine during these two weeks. She did not have any abdominal pain, nausea, vomiting, abdominal distention, belching or early satiety. She did not report any malaise, weakness or flulike symptoms before the onset of disease. She did not have any significant weight loss. She was married and had two children. She had taken oral contraceptive pills ten years ago for about two years; otherwise her medication history was negative.

Her grandmother died because of gastric cancer. On examination the patient was obese. The vital signs were normal. There was marked conjunctival and skin icterus with no spiderangioma, palmar erythema or telangiectasia.The abdomen was soft without tenderness or organomegaly.
Result of Complete Blood Count were normal
AST: 110IU/L, ALT: 88IU/L, ALP: 587IU/L total Bill: 14.2 mg/dl
Direct Bill:8.8mg/dl,Urea:17mg/dl,Cr:0.7mg/dl, PT: 12sec INR: 1, PTT: 26sec
IGg4:12mg/dl (11-57), CA19-9:40IU/ml (up to 37), CEA: 0.88ng/ml (up to 5.5)

Ultrasonography revealed dilated intrahepatic bile ducts and, the diameter of common hepatic duct (CBD) was14mm.Liver parenchyma was normal; Gall bladder had normal wall thickness without any sludge or stone. A magnetic resonance colangio pancreatography (MRCP) performed and revealed dilation of intra and extra hepatic bile duct and mid common bile duct stricture, Gallbladder was dilated without stone (Figure 1). According to these findings endoscopic retrograde colangio pancreatography (ERCP) was performed for her. After a hard intubation and enfacement of papilla cytology brushing was done and a plastic stent replaced to maintain biliary drainage.

4 hours after ERCP she developed severe abdominal pain and serum amylase raised to 1220 mg/dl. She was diagnosed as post ERCP pancreatitis, she was admitted in the ICU and the day after, a multi slice computed scan(CT) performed and showed signs of acute pancreatitis as enlargement of pancreas and heterogeneous enhancement of pancreas with fat stranding and fluid accumulation in retroperitoneal space with thickening of retroperitoneal fascia. Luminal narrowing of second portion of duodenum and partial dilation of proximal part of duodenum was also noted. Pancreas tissue completely surrounded the second part of the duodenum. Dilation of CBD and intra hepatic bile ducts was also noted (Figure 2).

Radiologic findings were compatible with annular pancreas .She received severe hydration with ringer lactate and she was admitted in ICU for about 3 days. The abdominal pain was completely resolved after seven days. She underwent surgery after recovery from pancreatitis. During laparotomy, pancreatic head had completely surrounded the duodenum with small suspicious lesion in pancreas head, pathology of frozen section revealed inflammatory cells and it was negative for malignancy (Figure3). A hepatico duodenostomy performed, patient's symptoms decreased and she was discharged in good health 10 days after surgery.

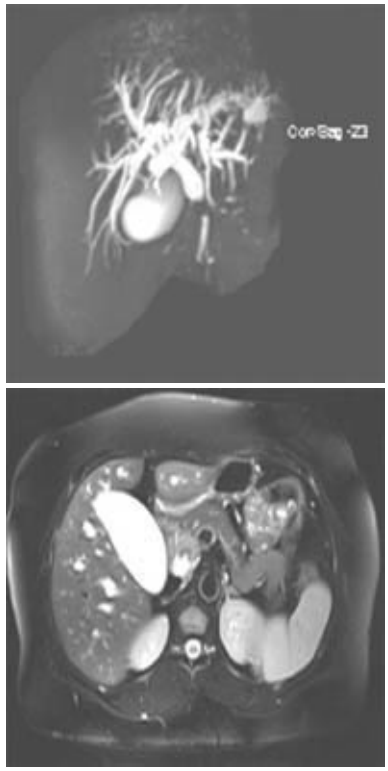

Fig. 1: MRCP showed dilation of intra and extra hepatic bile duct and mid common bile duct stricture

Three months later the patient was again admitted to the hospital with acute pancreatitis and significant weight loss. A multi slice triphasic abdominopelvic CT scan showed multiple metastatic lesions in the liver and a hypo echoic lesion in the pancreatic head (Figure4). Gasteroduodenoscopy and colonoscopy were all normal. Biopsy from liver lesions showed metastatic adenocarcinoma and fine needle aspiration of the pancreas lesion showed adenocarcinoma of pancreas. The patient was inoperable due to multiple liver metastasis therefore she was scheduled for chemotherapy.

DISCUSSION

Annular pancreas was diagnosed in1818 by Tieddman for the first time. (2) In this rare congenital abnormality the pancreatic tissue surround the second part of duodenum. There are many theories about the possible origin of AP. In the most accepted theory, pancreas is made of dorsal bud which forms the body and

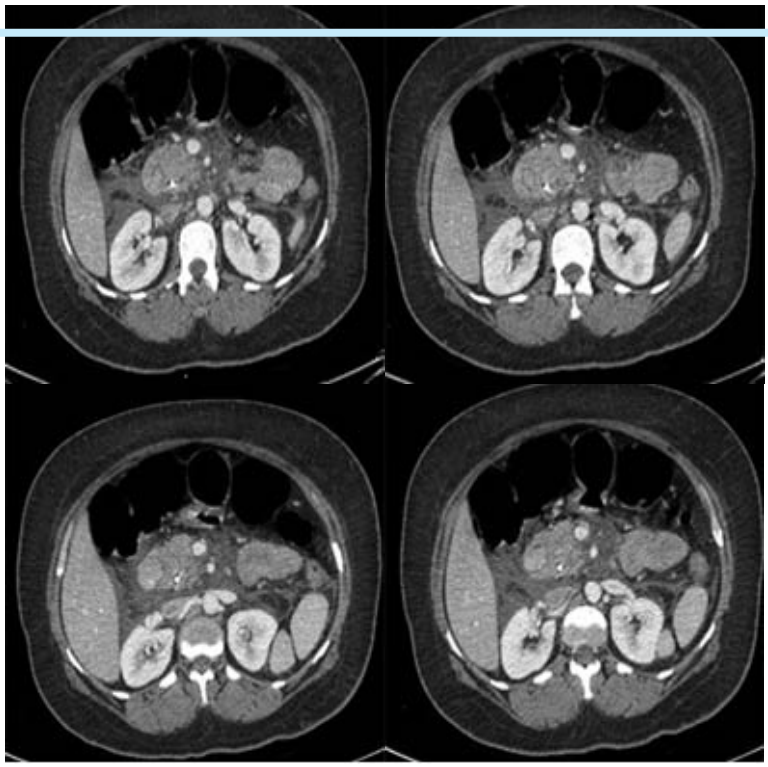

Fig. 2: CT scan revealed Pancreas enlargement with Luminal narrowing of second portion of duodenum and partial dilation of proximal part of duodenum. Pancreas tissue completely surrounded the second part of the duodenum

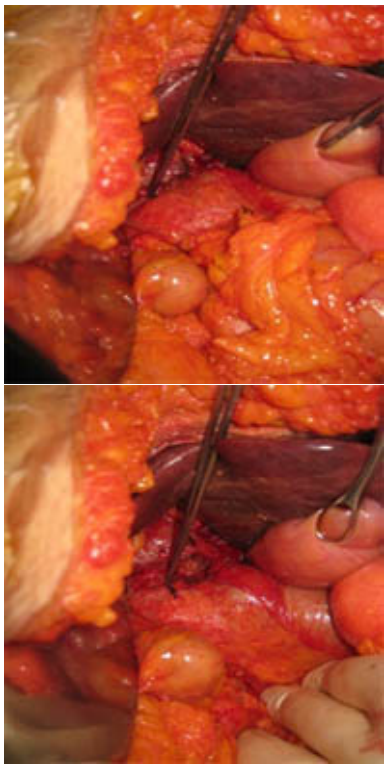

Fig. 3: Hepatico duodenostomy performed at laparoromy

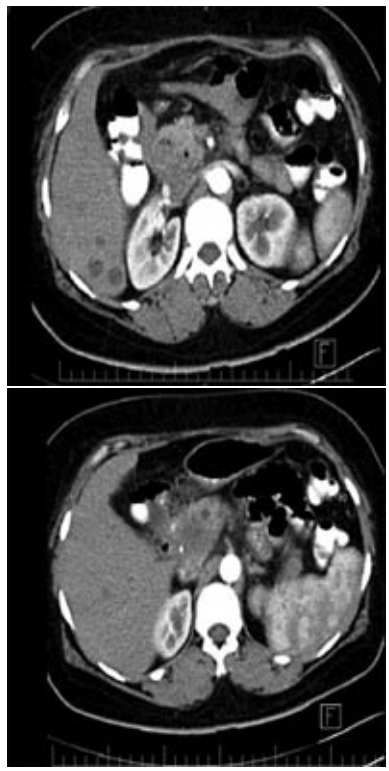

Fig. 4: showed multiple metastasis in liver, a hypo echoic mass is also noted in pancreatic head

tail, and ventral bud, which makes the head and uncinate process. In this congenital abnormality, the ventral bud divides into right and left parts; these two parts enclose completely the duodenum and form AP(1).

The majority of cases are among neonate and children and present with symptoms such as nausea, vomiting and other symptoms of gastric outlet obstruction. This abnormality remains asymptomatic most of the time and rarely become symptomatic in adults(2). The mean age of presentation in adults is between 20 to 50 years of age(5). In a series of adult patient with AP,70%presente with abdominal pain while47%presnt with nausea and vomiting due to duodenal obstruction(2,3). In another series adult annular pancreas was associated with duodenal obstruction (60%) , pancreatitis (15-50%) and peptic ulceration (26-48%)(4).

Other presentations include pancreatic hyperenzymemia, peptic ulcer disease, chronic pancreatitis and rarely obstructive jaundice or malignancy(2).

Surgery is required whenever annular pancreas is symptomatic. In those who present as duodenal obstruction duodenoduodenostomy is the preferred procedure. This is the most physiologic surgery as it results in the shortest blind loop(2).

In certain patients in whom the obstruction is in the distal duodenum, gasterojejunostomy is an alternative. The complications of postoperative ulcer formation and abdominal pain limit the use of this technique in all patients(6).

Obstructive jaundice is one of the rare presentations of adult AP. In 1982 Cang J Sang et al reported a 80 years old woman with obstructive jaundice and AP who had concomitant ampullary carcinoma for the first time(7). Sham et al reported 3cases of AP with obstructive jaundice, in their survey 1 of them had chronic pancreatitis and 2 had concurrent carcinoma of the ampulla of vater(8). Morrell and Keynes reported 15 annular pancreas patients with jaundice. The causes included pancreatitis (10 cases), choledocholithiasis (4 cases) and alcoholic liver disease (1 case)(9). Fung reported only 14 cases of carcinoma associated with annular pancreas in the English literature. The most common presenting symptoms in his report were jaundice.(11 cases) 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).

Yazawa have described a case of annular pancreas associated with carcinoma of the papilla of Vater but without jaundice.(13) Cases of insulinoma, mucinous cystadenocarcinoma of pancreas have been reported(11,12). Recently there have been a report of AP with pancreaticobiliary maljunction; this disorder may induce malignancies in biliary tract such as gallbladder cancer(4). Brönnimann reported a 55 years old woman who had annular pancreas associated with duodenal cancer, which was not diagnosed at the time of first surgery and revealed four weeks after surgery(6).

The case presented here was 47years old, which she was in the range of presentation of AP in adults. Contrary to most of the adult AP, this case was not complaining of any signs and symptoms of duodenal obstruction and painless jaundice was the main problem. At presentation time there was no evidence of malignancy despite complete investigation. Our presented case had had pancreatitis before surgery and distal obstruction of duodenum; consequently hepaticoduodenostomy was performed for her in order to retain biliary drainage. The frozen section of pancreatic lesion just showed inflammatory cells. Hence 3months later the patient came back with a pancreatic mass and liver metastasis. Similar to Brönnimann's report, pancreatic cancer was not diagnosed at the time of first surgery and discovered three months after that.

As it has been shown in litterateur, AP presenting with obstructive jaundice is very rare. This is interesting that most of these cases have association with periampullary malignancies, the most prevalent reported cancer is carcinoma of ampula of vater and rare cases of duodenal and pancreas cancer have also been described.

Presenting this case is of importance to emphasis on the fact that, when AP presents with obstructive jaundice the presence of periampullary carcinoma should be highly considered. The physician must thoroughly evaluate the patient for occult periampullary malignancies, and whenever these findings are negative, close follow up is mandatory for early detection of cancers originating from this area.

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