

case report

Annular pancreas causing extrahepatic biliary obstruction

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Background. Annular pancreas is an uncommon congenital abnormality, consisting of a flat band of pancreatic tissue, which encircles duodenum or extrahepatic biliary duct. We present a case of obstructive jaundice, caused by annular pancreas.

Case report. A 46 years old female was admitted because of a sudden onset of abdominal pain, vomiting and jaundice. For the last six years she occasionally noticed her skin was light yellow, in the last year she felt distension in the upper abdomen, especially after fatty meals.

Conclusions. Two US examinations, the first one six months before the admission, showed dilated hepatic ducts. The reason of dilatation was unclear, even after the endoscopic US examination. At operation an almost complete obstruction of the common hepatic duct was found, caused by a narrow band of pancreatic tissue.

Key words: pancreas abnormalities; bile duct obstruction, extrahepatic; cholangiopancreatography, endoscopic retrograde

Introduction

Annular pancreas is an uncommon congenital abnormality. It consists of a flat band of pancreatic tissue completely encircling the second portion of the duodenum or the extrahepatic bile duct.¹

It may manifest clinically in the neonate (52%) or remain asymptomatic until adulthood (48%).¹ It can represent an incidental finding during an endoscopic retrograde cholangiopancreatography (ERCP).²

In adults, symptoms of annular pancreas usually present at the age of 20-50 years as a duodenal obstruction, rarely as a biliary tract obstruction.²

Jaundice associated with an annular pancreas has been presumed to result from pancreatitis,^{2,3} but some reports suggest that an annulus is, by itself, capable of causing a significant biliary obstruction.^{4,5}

We present a case of obstructive jaundice, caused by annular pancreas, without pancreatitis.

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Case report

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A 46 years old female presented to the emergency room suffering from an acute abdominal pain and vomiting. At physical examina-

tion she was jaundiced. Liver function tests were abnormal (AST 7.14, ALT 8.78, bilirubin 57/42, γ GT 2.92), but levels of lipase and amylase were normal. The US examination showed thickened gallbladder wall, proximally dilated common bile duct and moderate dilatation of the right and the left intrahepatic ducts. No gallstones or masses were seen (Figure 1).

For the last six years the patient occasionally noticed having light yellow coloured skin. In the last year she felt distension in the upper abdomen after meals, especially in the afternoon, after eating fatty food. She never vomited, had no diarrhoea and was afebrile. Difficulties gradually increased and 6 months ago she underwent the first US examination. A concrement, a few mm in size, was found in the infundibulum of the gallbladder. The common bile duct was dilated up to 1 cm

above the ampulla of Vater. The cause of dilatation was not clear.

The patient underwent ERCP.

Duodenum and papilla of Vater were normal. A 2 mm long stricture of the common bile duct with prestenotic dilatation was seen, intrahepatic ducts were also dilated, the cystic duct was not obstructed (Figure 2).

The pancreatic duct opened into the common ampulla and had a normal course. There was no accessory pancreatic duct visible.

The endoscopic US examination confirmed a short, smooth stenosis of the choledochal duct and a dilatation of the hepatic duct, looking benign. Some small concretions were found in the gallbladder. There were no signs of tumour like masses (Figure 3).

A congenital anomaly was suspected and with the patient in a good condition, despite a long history of symptoms, the surgeon de-



Figure 1. Ultrasound: dilatation of hepatic ducts.

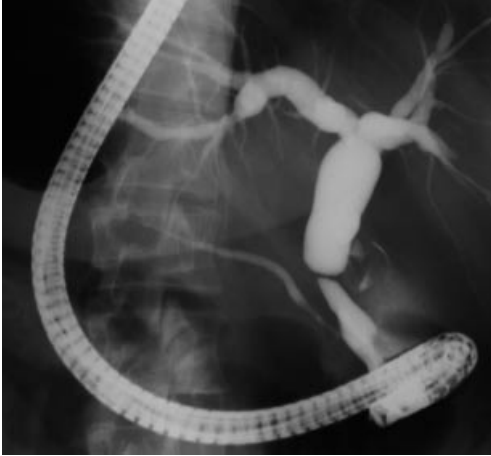


Figure 2. ERCP: stenosis of common hepatic duct proximally.



Figure 3. Endoscopic ultrasound: stenosis.

cided to perform abdominal laparotomy without any additional imaging procedures.

At intraoperative cholangiography, an almost complete obstruction of the common bile duct was found (Figure 4). It was caused by a narrow band of pancreatic tissue encircling the common bile duct. A cholecystectomy and a hepaticojejunal anastomosis were done.

Discussion

Annular pancreas is a congenital anomaly of rotation and fusion of primitive intestine and



Figure 4. Intraoperative cholangiography: stenosis causing complete obstruction of common hepatic duct.

its associated structures, which may cause duodenal obstruction in the neonatal period or remain silent throughout life.² At post-mortem, the incidence has been quoted as between 5-15/100.000. ERCP has led to a much more frequent diagnosis of annular pancreas in as many as one in 150 patients undergoing this procedure.⁴

There are three theories concerning the formation of the annular pancreas:

- hypertrophy of both, the ventral and the dorsal ducts, resulting in a complete ring,
- adherence of the ventral duct to the duodenum before rotation and
- hypertrophy or adherence of the left bud of a paired ventral primordium.¹

There is a clear distinction between the annular pancreas encircling duodenum presenting in children and that in adults, probably due to different severity of stenosis.⁶

In neonates, the symptom of severe duodenal obstruction is vomiting on the first day of life. There is often previous history of polyhydramnion. A number of other anomalies such as intestinal malrotation, duodenal atresias, and cardiac anomalies are often present as well.^{1,6}

In older children and adults, the onset is more insidious, with intermittent clinical problems. Nausea, vomiting, and epigastric

pain are the main complaints due to the duodenal obstruction.^{2,4} Pancreatic stenosis of the duodenum is usually not sufficient to cause symptoms until there the provoking factors - peptic ulcer disease or pancreatitis - supervene.

The incidence of associated gastric and duodenal ulcers ranges from 26 to 48%, and pancreatitis develops in 15 to 30% of patients.^{1,2}

It has been suggested that annular pancreas may be the cause of extrahepatic biliary obstruction because of pancreatitis in the parenchyma, encircling the duodenum and bile duct, although jaundice is rare.³

In our case, no laboratory or clinical signs of pancreatitis were present. Laboratory data (AST 7.14, ALT 8.78, bilirubin 57/42, γ GT 2.92, normal lipase and amylase) were suggestive of biliary obstruction with the abnormal liver function.

The US examination is the first in patients with abdominal symptoms. ERCP is the procedure of choice in patients with biliary obstruction, especially if the findings at US are equivocal. A diagnosis of annular pancreas encircling biliary tract is not always made preoperatively.^{2,7} ERCP fails to make the diagnosis of annular pancreas if there is the obstruction of the duct leading to annular pancreas, if the duct of the annular pancreas does not empty into the main pancreatic duct or if there is a high grade of duodenal obstruction.²

In our case, no anomaly of the pancreatic duct was shown at ERCP.

In reviewing literature, not many cases of annular pancreas, causing biliary obstruction, without pancreatitis or duodenal obstruction, were found.

Adamo and co-authors reported it as being very interesting because of obstructive jaundice being the first symptom of this anomaly.⁸ Another article presents three cases of annular pancreas with the obstructive jaundice, but the obstruction was due to carcinoma of ampulla of Vater.^{4,9}

In another case annular pancreas was causing a dilatation of biliary and pancreatic ducts, without obstructive symptoms and with normal liver tests. This patient underwent CT because of the other suspected disease and the dilatation was an accidental finding.¹⁰

Conclusions

The prevalence of annular pancreas in Slovenia is not known. In spite of such congenital anomaly being rare and much rarely being the cause of obstruction of the extrahepatic biliary tract we should keep it in mind as a differential diagnostic possibility. But, as other authors suggest, an annular pancreas should not be considered a satisfactory explanation for the obstructive jaundice until other possibilities have been fully excluded.⁴

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