Carcinoma of Papilla Vateri presenting as recurrent acute pancreatitis

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Abstract

Tumors of the Papilla of Vater can cause several clinical symptoms, the most prominent being jaundice, weight loss, anorexia, fever, abdominal pain and itching (1). Acute pancreatitis as a presenting symptom of ampullary carcinoma is rare. Few previous cases have been described in the literature (2,3).

The prognosis of patients with jaundice is unfavorable in comparison with non icteric patients at the time of diagnosis (4) due to different staging, hence more complications, but not due to different histology.

We report here a case of recurrent pancreatitis that was the only presentation of Vater ampullary carcinoma diagnosed by endoscopic ultrasound followed by duodenoscopy with guided biopsy.

Recurrent pancreatitis without identifiable cause, particularly in elderly patients, could suggest tumor of the head of pancreas or the periampullary region among other causes such as intraductal papillary mucinous tumor, microlithiasis etc. An endoscopic ultrasound can allow earlier diagnosis and mandates biopsy in these cases. (Acta gastroenterol. belg., **2004**, 67, **309-310**).

Key words : Papilla of Vater, papillary carcinoma, acute pancreatitis.

Case report

A 75-year-old man without chronic diseases or alcohol abuse was admitted to our department with a two days history of upper abdominal pain radiating to his back without fever, vomiting, diarrhea, weight loss or jaundice. Two months earlier he had been hospitalized in our department for acute pancreatitis for the first time, ultrasound and computed tomography of the abdomen then, showed signs of acute pancreatitis only.

Physical examination was normal except for diffuse tenderness in the upper abdomen.

Laboratary tests : serum amylase 5074 (normal 0-200 u/l). Serum bilirubin, transaminases, alkaline phosphatase, gamma glutamyl transpeptidase , triglycerides and calcium were normal. An ultrasound of the abdomen showed edema and enlargement of pancreas, normal-sized liver, without cholelithiasis or dilatation of bile ducts. A computed tomography of the abdomen showed decreased density of the pancreatic body, with strands in the peripancreatic fat and a small amount of peripancreatic fluid. Spleen, liver, gallbladder, bile ducts and pancreatic duct within normal limits.

An endoscopic ultrasound was done, showing multilobular mass of the papilla of Vater with two separate ducts. No ductal enlargement was found at the site of the papilla, the papilla was pathologically hypoechoic and appeared to be separable from the head of the pancreas. Duodenoscopy with biopsy was then performed and specimens demonstrated anaplastic carcinoma of papilla.

The patient underwent Whipple's resection, the tumor was localized at the ampulla of Vater without evidence of metastatic disease. The surgical specimens confirmed the diagnosis of biopsy, two separate open, not dilated ducts, pancreatic and choledochus, entering the papilla were found. The patient did well and was symptom free for the next 12 months.

Discussion

The ampulla of Vater may be the site of sarcoma, carcinoid tumor, adenocarcinoma or benign tumors. The most common clinical manifestations of papillary tumors are obstructive jaundice (84%), weight loss (75%), abdominal pain (59%), fever due to cholangitis (52%) and in a rare cases acute pancreatitis (1,2,3). Patients with ampullary carcinoma usually have icterus at the time of diagnosis, and the clinical outcome remains unfavorable (4). A few patients with ampullary carcinoma are not icteric at the time of diagnosis as was in our case and in most non-icteric cases, an elevation of the serum alkaline phosphatase levels and dilatation of the biliary tree were clues to the diagnosis of an ampullary tumor (4).

We found our case to be of special interest due to the rarity of clinical presentation, normal primary imaging and laboratory data at the time of diagnosis. Perhaps two separate ducts, pancreatic and choledochus, before entering the papilla may explain the normal laboratory data in our case.

There are many causes of unexplained idiopathic acute pancreatitis including occult microlithiasis (in approximately 30% of patients with idiopathic pancreatitis) (6,7), choledochocele, foreign bodies, pancreas divisum, ampullary or pancreatic tumors (8), abnormalities of sphincter of Oddi (stenotic or high-pressure sphincter of Oddi), mutations of the cystic fibrosis transmembrane conductance regulatory (CFTR) gene (9) and high index of suspicion should be maintained, especially in elderly patients with pancreatitis.

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The importance of recognizing this association is that a potentially curable malignancy may be diagnosed and treated promptly and successfully.

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