

creatic duct communicated with an inferior branch of the ventral duct at the uncinate process, draining through the minor papilla (Figures 1, 2). Nevertheless, ansa pancreatica was not considered as a predisposing factor in any of them.

When there is an obliteration in the embryological course of the dorsal pancreatic duct at the confluence level with the ventral duct, the proximal portion of the dorsal duct communicates with the inferior branch of the ventral duct, and a new accessory duct shaped like a reverse 'S' character, terminating in the minor papilla, is formed (1,4). Despite the other side branches of the ventral duct, in the ansa pancreatica type, the accessory duct communicates with the ventral duct at an ob-

lique angle. Although it can be hypothesized that an accessory dorsal duct draining through the minor papilla may prevent pancreatitis by decreasing the pressure in the main duct, in the ansa pancreatica anatomic variation, the drainage is not that matured (1, 3).

'Ansa pancreatica' can be considered as a predisposing factor in patients with idiopathic pancreatitis, can accompany other risk factors such as alcoholism, and can also take part in the etiology of postoperative pancreatitis following pancreaticobiliary surgery (2). As MRCP is being widely used, it is important to recognize and clearly define the pancreas ductal variations, especially in the preoperative period, as they may be clinically significant.

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Carcinoid tumor of the ampulla of Vater presenting as recurrent acute acalculous cholecystitis attacks

Tekrarlayan akut akalkuloz kolesistit atakları ile görülen ampulla Vateri'nin karsinoid tümörü

To the Editor

Carcinoid tumors are rare neuroendocrine neoplasms that comprise only 1.2% to 1.5% of all gas-

trointestinal tumors. The ampulla of Vater is an extremely rare location for carcinoid tumors (less

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Manuscript received: 10.01.2008 Accepted: 13.03.2008

than 0.3% of all GIS carcinoids) (1).

A 63-year-old woman was admitted with nausea, pain in the upper abdominal region and jaundice. She had upper abdominal discomfort over the last two years and was treated for acute cholecystitis three times. Blood biochemistry evaluation revealed a total serum bilirubin of 3.66 mg/dl, direct bilirubin 2.75 mg/dl, aspartate aminotransferase 320 IU/L, alanine aminotransferase 263 IU/L, alkaline phosphatase 154 IU/L, and gamma glutamyl transferase 428 IU/L. Radiologic examination revealed no stones in gall bladder, thickness in the wall, dilated common bile duct (10 mm) and periportal, peripancreatic, and mesenteric lymphadenopathies. An endoscopic retrograde cholangiopancreatogram (ERCP) was performed and a swollen, ulcerated papilla was encountered, and papilla biopsy was performed (Figure 1). The immunohistochemical examination of the specimen showed the mass on the ampulla of Vater to be a carcinoid tumor.

Pancreatoduodenectomy with extended lymph node dissection was performed. The pathological examination confirmed the carcinoid tumor, 1 x 1.7 cm in diameter. Surgical margins were tumor-free and all lymph nodes dissected were negative for tumor infiltration. No chemotherapy was planned and the patient remains disease-free three years after the operation.

The characteristics of carcinoid of the ampulla of Vater (CAV) are unclear. The most common presenting symptoms of CAV are according to its mass effect, which are jaundice (59%) and epigastric pain (37%). The occurrence of carcinoid syndrome is rare [2], and this makes the diagnosis more difficult.

Detection of the tumor is not easy due to its size and mucosal sparing. ERCP and duodenoscopy are more helpful in the diagnosis of CAV and differential diagnosis for other types of periampullary tumors, papillary stenosis, or impacted stones, which may produce similar findings (2). Large and deep biopsies are needed to verify the tumor.

Our patient presented with jaundice and acute acalculous cholecystitis. Her medical history revealed recurrent acalculous cholecystitis attacks before jaundice was observed. ERCP was the ma-

jor helpful test for diagnosis in our patient.

Since the review of patients with ampullary carcinoid revealed that 25% of those patients have von Recklinghausen's disease, the clinical features of the disease such as skin lesions (café au lait spots) should be informative for the clinician regarding the location of the lesion that causes jaundice (3).

Carcinoid of the ampulla of Vater can be accepted as a benign disease considering the fact that five-year survival is over 90%. However, recent studies agree on the fact that achieving a tumor-free area is the key factor for good prognosis. Metastasis, rather than size of the tumor, has a direct relationship with the prognosis (4). In our case, computerized tomography scan suggested local lymph node involvement. We then performed a pancreaticoduodenectomy deciding that resection with larger tumor-free margins and resection of the lymph nodes in the neighborhood of the tumor would be the optimal approach in the patient. The patient is still well with no signs of a recurrence.

In conclusion, all patients with relapsing acalculous cholecystitis attacks should be examined for CAV, since early detection of this kind of tumor may improve the postoperative survival.

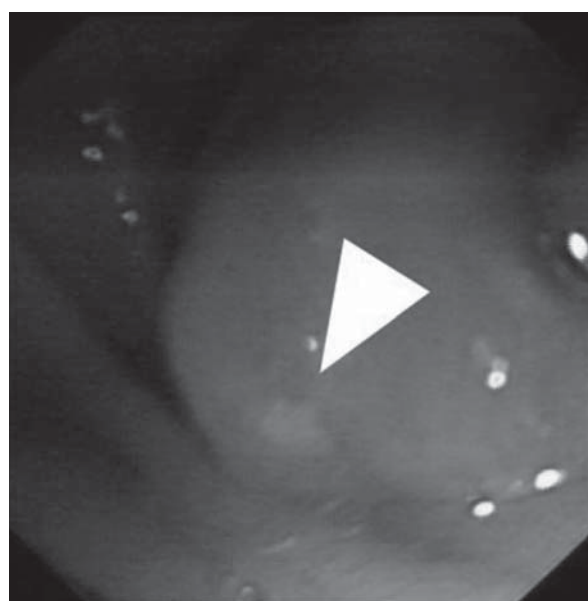


Figure 1. ERCP demonstrates the enlarged ampulla of Vater.

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