Chronic periaortitis presenting as common bile duct obstruction

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Abstract
The case of a 67 year old woman is reported who presented with cholestatic jaundice and was found to have, in addition, an inflammatory abdominal aortic aneurysm. Only at necropsy did histopathology show chronic periaortitis as the aetiology of a pancreatic head mass which, during life, mimicked a pancreatic neoplasm obstructing the bile and pancreatic ducts.

The association of chronic periaortitis (peri-aneurysmal retroperitoneal fibrosis) with duodenal, pleural, and lung fibrosis, chronic coronary periarteritis, and ureteric entrapment have previously been documented. Although there have been reports of common bile duct compression by an abdominal aortic aneurysm and retroperitoneal fibrosis simulating carcinoma of the pancreas in an anicteric patient, we believe that this is the first report of a case of chronic periaortitis presenting with cholestatic jaundice and obstruction of the common bile duct.

Case report
A 67 year old woman was referred with jaundice, vomiting, anorexia, weight loss, and upper abdominal and back pain. She was also found to have a pulsatile mass in the epigastrium.

Her erythrocyte sedimentation rate was 47 mm in the first hour and 'liver function tests' were abnormal with a serum bilirubin concentration of 479 μmol/l (normal range 10–20), an alkaline phosphatase activity of 2230 IU/l (normal range 80–300), and transaminases raised fivefold. The ultrasound scan of her abdomen showed intrasplenic duct dilatation but no evidence of a stone or mass in the head of the pancreas. There was also a 4 cm fusiform abdominal aortic aneurysm.

Endoscopic retrograde cholangiopancreatography (ERCP) showed strictures in the distal common bile duct and the distal pancreatic duct with presstenotic duct dilatation of both. Using the criteria proposed by Nix et al., a tumour of the pancreatic head or ampulla of Vater would have been likely. A biliary endoprosthesis was inserted at ERCP to drain the
biliary tree. Coeliac axis and superior mesenteric intra-arterial digital subtraction angiography showed no 'tumour circulation.' At the time it was thought that further imaging was unlikely to change management.

At laparotomy, surprisingly, no tumour mass was identified around the pancreatic head or common bile duct. In the absence of a tissue or surgical macroscopic diagnosis of malignancy, plans for a Whipple's procedure were abandoned. The large inflammatory aortic aneurysm with surrounding retroperitoneal fibrosis was resected and replaced with a Dacron prosthesis. Histology of the resected specimen showed severe atherosclerosis, medial breaches, and typical adventitial inflammation and fibrosis seen in chronic periarteritis (Fig 1).

With the biliary stent in situ the patient's jaundice faded and the erythrocyte sedimentation rate fell to 25 mm in the first hour. Follow up ultrasound scans done four and seven months postoperatively showed a mass posterior to the stent and to the left of the coeliac axis. At the time it was thought that this mass represented a pancreatic neoplasm and the patient was managed accordingly. She remained in good health until a few months before her final admission. Despite pancreatic enzyme supplements she continued to lose weight and eventually died with perinephric sepsis 19 months after presentation.

Necropsy showed a ruptured right perihepatic abscess and acute peritonitis, which was clearly the immediate cause of death. In addition, there was generalised atheroma in the major arteries. A firm 4 × 3 × 2 cm mass was present in the pancreatic head severely constricting the pancreatic duct and infiltrating part of the wall of the common bile duct adjacent to the ampulla but not extending to the ureters. Macroscopically this appeared to be a primary pancreatic carcinoma. The biliary stent had remained in correct position and the common bile duct was not dilated. Histology of the pancreatic mass (Fig 2) showed extensive dense fibrosis in the peripancreatic tissues adjacent to the site of the previously resected inflammatory aneurysm. Despite careful examination of multiple sections of this pancreatic mass there was no evidence of malignancy. The histological appearances were similar to those in the aneurysmal adventitial tissues.

Discussion

Chronic periarteritis, as defined,17 describes the active chronic inflammation around the lower abdominal aorta accompanied by progressive scarring with venous obliteration and contraction seen, typically, in middle aged and elderly patients, especially men. The erythrocyte sedimentation rate usually exceeds 45 mm in the first hour. Parums et al suggested an autoimmune mechanism incriminating the insoluble lipid, ceroid, within atheroma.4 Corticosteroid treatment has been preferred8 and shown by computed tomography in one case9 to result in shrinkage of periaortic tissue. Although inflammation may rarely subside spontaneously, it usually does so after treatment with corticosteroids or, curiously, after surgery and even biopsy alone. In this case an endobiliary stent to bypass the stenosing lesion combined with an aneurysmectomy resulted in a moderate fall in the erythrocyte sedimentation rate from 47 to 25 mm in the first hour.

Clearly, periarteritis affecting the pancreatic head may mimic pancreatic malignancy. A readily available serological diagnostic test, possibly involving ceroid,7 would prove useful. In the absence of controlled trials we can only speculate if corticosteroids may have helped in this case.

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