

Answer

Papillary Low-Grade Neuroendocrine Tumor of the Pancreas With Pancreatic Duct Dilation

Figure 1. Axial magnetic resonance image of the abdomen demonstrating large pancreatic ductal dilation without obvious mass.

Figure 2: Intraoperative photograph demonstrating an enlarged main pancreatic duct (black arrows) and enlarged peripheral pancreatic ducts (white arrows).

Prominent pancreatic duct dilation with a mass lesion within the head of the pancreas has been found to represent a primary adenocarcinoma of the pancreas in most patients. These patients are also found to have concomitant biliary dilation. Isolated significant (>2-cm) pancreatic ductal dilation has been demonstrated to occur more commonly in mucin-producing lesions of the pancreas and primarily in intraductal papillary mucinous tumors of the pancreas.¹

Neuroendocrine lesions of the pancreas represent 0.5% of all pancreatic tumors.² Approximately one third of neuroendocrine tumors are hormonally inactive and account for about 20% of all endocrine tumors of the pancreas.³ Histochemically, they are identified as insulin (50% of cases), pancreatic polypeptide (40%), glucagon (30%), and somatostatin (13%) cells. Nonfunctioning pancreatic tumors are usually unifocal, except when associated with multiple endocrine neoplasia type 1 syndrome.⁴ These lesions usually occur during the fourth or fifth decade of life, with an even sex distribution.⁵

Because these tumors remain clinically silent during their growth, they may attain great size without causing apparent clinical findings and commonly present in an advanced stage. Other forms of nonfunctioning neuroendocrine tumors, when originating in critical locations, can be found with elevated liver function test results or, as in this case, as a dilated pancreatic duct.

Patients who present with significant pancreatic ductal dilation should be examined to rule out proximal mass lesions. Most of these patients will ultimately undergo

pancreaticoduodenectomy, which establishes the final diagnosis.

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REFERENCES

1. Sohn TA, Yeo CJ, Cameron JL, Iacobuzio-Donahue CA, Hruban RH, Lillemoe KD. Intraductal papillary mucinous neoplasms of the pancreas: an increasingly recognized clinicopathologic entity. *Ann Surg.* 2001;234:313-321.
2. Phan GQ, Yeo CJ, Hruban RH, Lillemoe KD, Pitt HA, Cameron JL. Surgical experience with pancreatic and peripancreatic neuroendocrine tumors: review of 125 patients. *J Gastrointest Surg.* 1998;2:472-482.
3. Hochwald SN, Zee S, Conlon KC, et al. Prognostic factors in pancreatic endocrine neoplasms: an analysis of 136 cases with a proposal for low-grade and intermediate-grade groups. *J Clin Oncol.* 2002;20:2633-2642.
4. Norton JA, Alexander HR, Fraker DL, Venzon DJ, Gibril F, Jensen RT. Comparison of surgical results in patients with advanced and limited disease with multiple endocrine neoplasia type 1 and Zollinger-Ellison syndrome. *Ann Surg.* 2001;234:495-505.
5. Eriksson B, Oberg K. Pipomas and nonfunctioning endocrine pancreatic tumors: clinical presentation, diagnosis, and advances in management. In: Mignon M, Jensen RT, eds. *Endocrine Tumors of the Pancreas: Recent Advances in Research and Management.* Basel, Switzerland: S Karger AG; 1995:208-222.

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