

Periampullary carcinoid of the ampulla of Vater presenting as an intraductal papillary mucinous tumour of the pancreas: a sheep in wolf's clothing

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Carcinoids are uncommon, with an estimated incidence at all anatomic sites of 1 in 12 000.¹ Although 67% of these lesions are found within the gastrointestinal tract, fewer than 1% are located at or near the ampulla of Vater.² These periampullary tumours have characteristics that set them apart from carcinoids at other sites, including strong immunohistochemical positivity for somatostatin and presence of psammoma bodies.^{3,4} Although extremely rare in the general population, periampullary carcinoids have a well-described association with neurofibromatosis type 1 (NF1),^{3,4} a genetic disorder with an incidence of 1 in 2500–3000.²

We describe the case of a woman who had a periampullary carcinoid that presented as an intraductal papillary mucinous tumour of the pancreas.

Case report

A 49-year-old woman with a history of NF1 had presented 9 years previously with vague abdominal discomfort. Endoscopy revealed a lesion in the duodenum, measuring 1.0 cm in diameter, 1.0 cm proximal to the ampulla of Vater. Biopsy specimens showed no abnormality. Imaging showed marked dilatation of the entire pancreatic duct, up to 1.8 cm in diameter. The duct could be traced all the way to the ampulla; however, the

mass seen on endoscopy was not demonstrated, and no cause for the dilatation was found.

The patient was lost to follow-up for several years; then, endoscopic ultrasonography and computed tomography (CT) revealed cystic changes in her pancreas, with dilatation of the pancreatic duct, up to 2.4 cm in diameter (Fig. 1). A diagnosis of intraductal papillary mucinous tumour was made on the basis of these changes and the normal ampullary appearance. She underwent total pancreatectomy with duodenectomy, distal gastrectomy, segmental jejunectomy and splenectomy.

On gross pathological examination of the tumour, the submucosal mass in the duodenal wall measured 2.0 × 1.5 × 1.2 cm. The pancreatic duct was dilated to a maximum circumference of 6.0 cm, and the pancreatic parenchyma was diffusely fibrotic. No papillae were apparent on the duct epithelium, and no mucin was identified. The orifice of the pancreatic duct was separate from the common bile duct, corresponding to the region of the submucosal mass (Fig. 2).

Histologically, the mass was composed of uniform cells arranged in cords and nests with granular cytoplasm, and central round nuclei with coarse chromatin. Frequent psammoma bodies and rare mitotic figures were seen. Immunohistochemical staining showed tumour

cells that were strongly positive for neuroendocrine markers and somatostatin. The morphology and immunohistochemical profile of this tumour were consistent with those of a well-differentiated neuroendocrine (carcinoid) tumour.

Discussion

The case we present is interesting, not only for the rarity of the lesion but also for its unusual presentation. Most patients having periampullary carcinoids present with jaundice or epigastric pain,³ and although our patient had discomfort, she never had obstructive jaundice caused by the variant anatomy of her ampullary region. Obstruction of the pancreatic duct outlet led to duct dilatation, chronic pan-



FIG. 1. A CT scan showing cystic dilatation of the pancreatic duct (arrowhead).

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Accepted for publication Sept. 22, 2007

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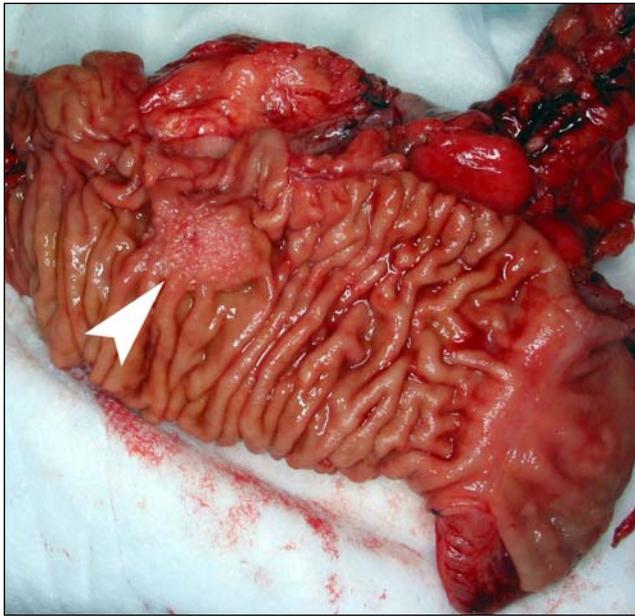


FIG. 2. The opened duodenum shows the submucosal mass (arrowhead), measuring 2.0 cm in dimension. The orifice of the pancreatic duct was separate from the common bile duct, corresponding to the region of the submucosal mass.

creatitis and eventual parenchymal fibrosis. Imaging performed later in her clinical course led to a presumptive diagnosis of intraductal papillary mucinous tumour, and the decision was made to perform a total pancreatectomy.

No specific guidelines exist for optimal management of periampullary carcinoids, but surgical excision is the treatment of choice. The Kausch–Whipple procedure is favoured over local resection when tumour dimension is greater than

2.0 cm¹; however, unlike carcinoids at other locations, the size of periampullary carcinoids does not predict metastatic disease, and spread to lymph nodes or liver has been detected with a primary tumour less than 1 cm in size.⁴ For this reason, many surgeons recommend pancreaticoduodenectomy for all periampullary carcinoids, regardless of tumour size.^{2,5}

The prognosis for these tumours is good even in the face of metastases, with reported postoperative 5-year survival rates of nearly 90%.

Competing interests: None declared.

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