

competitive inhibitor of KIT and platelet derived growth factor receptor α (PDGFR α) and the results have been encouraging in patients having unresectable or metastatic GISTs.⁸

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Spleen preserving distal pancreatectomy for a large papillary

cystic and solid tumor of the pancreas

Introduction

Most pancreatic tumors are malignant and have a bad prognosis. However, papillary cystic and solid tumour of the pancreas (PCSTP) is an unusual low-grade malignancy that rarely metastasizes.¹ Surgical resection is generally curative and the prognosis is excellent. It mostly affects young females with a mean age of 25 years.² It has also been referred to as a solid-cystic epithelial tumour, solid-pseudopapillary tumour or papillary-cystic tumour.³ It makes up 0.2–2.7% of all pancreatic cancers.⁴ Since the original description by Frantz in 1959, the incidence of PCSTP has been increasing,⁵ although it may be that it is increasingly being diagnosed. Very few cases of PCSTP have been reported from India.

Case report

A 29-year-old female presented with chronic epigastric and left hypochondriac discomfort since 1 year. It was associated with postprandial fullness and occasional vomiting. On examination she was overweight and there was epigastric tenderness, vague mass was palpable in the left hypochondrium which moved with respiration. All haematological and biochemical parameters were within normal limits. Abdominal ultrasonography demonstrated partially cystic mass of 18 x 12 cm in distal pancreas with a possibility of a neoplasm. Contrast enhanced computed tomography (CT) scan confirmed 18 cm mass in the tail of pancreas without any metastases.

The patient underwent staging laparoscopy which showed large mass arising from the tail of the pancreas without any metastases. An en-block spleen preserving distal pancreatectomy, including the pancreatic mass, was performed (**Figure 1**). The patient made an uneventful recovery.

On gross examination the pancreatic tumour was oval, 18 cm in diameter, and was surrounded by a fibrous pseudocapsule. Its cut surface showed solid and cystic spaces. On microscopy, the solid portion of the tumour revealed sheets of uniform polygonal cells as well as non-cohesive papillae arranged around fine fibrovascular cores. The cyst wall was composed of dense acellular fibrous tissue within which the tumour cells were arranged as cords and trabeculae set within a mucinous background. No invasion of the tumour into

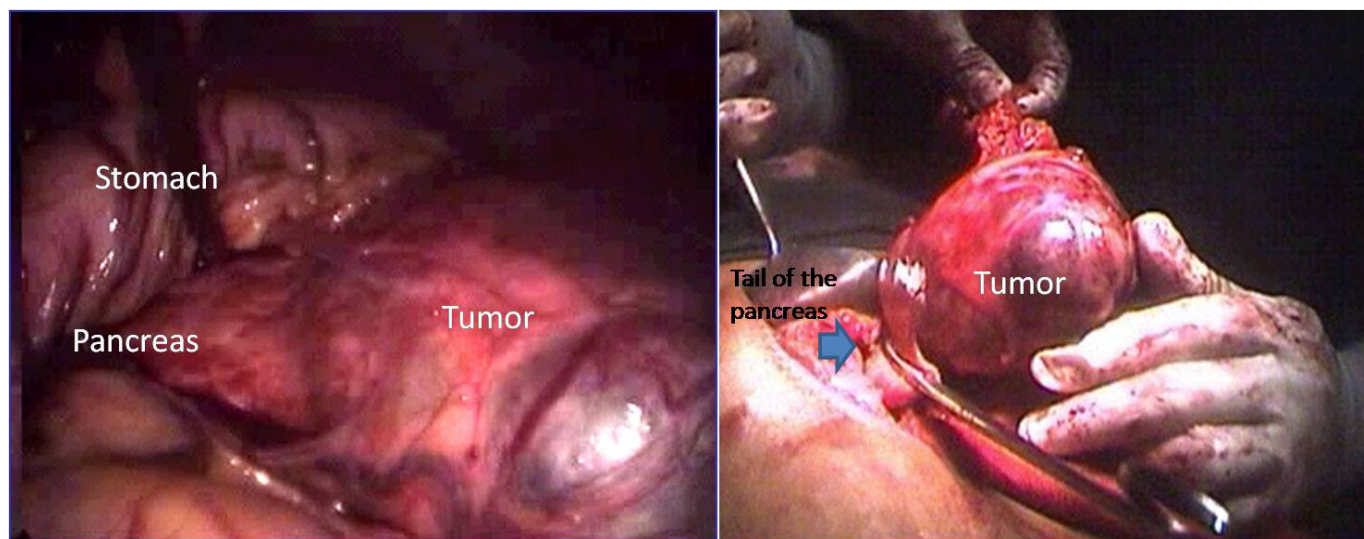


Figure 1: Laparoscopic view of the tumor and distal pancreatectomy procedure

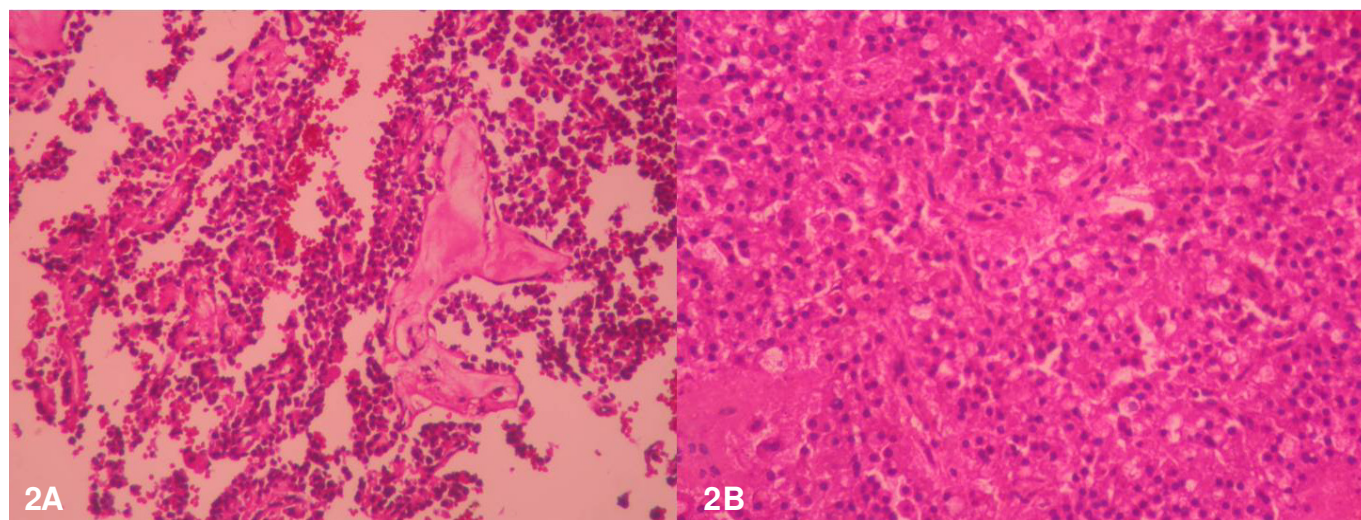


Figure 2: Photomicrograph showing papillary structures having a delicate fibrovascular core lined by bland round to oval tumor cells H and E 200X

surrounding normal tissue was present (**Figure 2a & 2b**). The patient is well after a follow up for the last two year.

Discussion

The papillary cystic and solid tumour of the pancreas (PCSTP) is an unusual low-malignant epithelial tumour, which mostly affects young females with a mean age of 25 years.¹ It was first, described in 1959 by Frantz, since then approximately 452 cases have been reported in the world literature.⁵ They constitute 0.13-2.77% of all the pancreatic tumors and 70% of tumors occur in the first three decades of life.¹ The tumor is rare in men, accounting for 7% of the cases with male: female ratio of 1:13. Clinically patient may be asymptomatic or present with vague abdominal discomfort. Tumor is localized in head, body and tail of the pancreas in the ratio of 4:2:4. Grossly, it is

usually a well circumscribed tumor ranging in size from 2-25 cm diameter. It has a variegated appearance with solid, cystic and papillary areas with foci of necrosis and hemorrhages. These degenerative changes are probably related to vascular ischemia. The clinical differential diagnosis includes all cystic and solid lesions of the pancreas, like inflammatory pseudocyst, mucinous cystic tumors, microcystic adenoma and mucinous cystadenocarcinoma. However, the histological features of this tumor are characteristic and diagnostic. Histological parameters predicting the tumor's aggressive behavior include: capsule thickness of more than 2 mm, high nuclear grade, prominent necrobiotic nests, capsular invasion into the surrounding normal pancreatic tissue and other tissues, vascular invasion and metastasis.⁶

The origin of this tumor remains an enigma. Immunophenotype is not specific: it displays positivity for vimentin,

alpha-1-antitrypsin and neuron specific enolase. This diversity of immuno-staining emphasizes its exocrine and endocrine differentiation.⁵

Surgical management has been tailored to the slow-growing, non-invasive nature of this tumour. Depending on the location of the PCSTP, the surgical option is chosen. Size of the tumor does not decide the resectability. With tumour involvement of the head of pancreas, a pylorus-preserving pancreaticoduodenectomy is recommended. PCSTPs involving the neck or body of the pancreas are resected by central pancreatectomy and reimplantation of the pancreatic remnant into the stomach, with theoretical benefit of preserving pancreatic parenchyma and spleen.⁷ When the tumour was located at the pancreatic tail, tail and body, or body of pancreas, distal pancreatectomy with splenectomy was employed in many cases.⁸ But the presence of the spleen may have a favorable effect on recurrence control and survival. The hypothesis that the spleen suppresses tumor growth from minimal residual disease in the critical early postsurgical period and reduces the risk of recurrent disease was tested. Now many authors recommend spleen conservation following distal pancreatectomy whenever possible.^{7,9}

Laparoscopic pancreatic resection can be applied for small tumor. Baker et al¹⁰ and Nakamura et al¹¹ studied 27 and 21 patients, respectively, who underwent laparoscopic distal pancreatectomies. They concluded that the procedure is a safe, effective modality for managing neoplasms of the pancreatic body and tail, providing a morbidity rate comparable to that of the open procedure and a substantially shorter length of stay.

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Asymptomatic heterotopic pancreas in the mesentery

Introduction

Heterotopic pancreas (HP) is an uncommon entity. It is usually detected incidentally and may or may not be associated with complications related to the pathologic conditions of the pancreas itself but may present with complications like inflammation, bleeding, obstruction or malignant transformation.^{1,2} HP in the mesentery of the small intestine is very rare. A brief review of the HP is discussed along with the case.