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Case Report
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Solid Pseudopapillary Neoplasm of the Pancreas

*Yi-Wen Liao[1], Vincent Chong[1], Neil Wylie[1], Neville Angelo[2] and Jonathan Koea[1]**

[1]Department of Surgery, North Shore Hospital, New Zealand

[2]Department Pathology North Shore Hospital, New Zealand

Abstract

A 35 year old presented with a 6 month history of an epigastric mass and upper abdominal discomfort. Clinical examination confirmed a firm mass in the epigastrium but no signs of metastatic disease. Investigations showed that tumour markers (carcinoembryonic antigen, Ca 19-9, Ca 125, chromogranin A) were within the normal limits. A CT scan of the abdomen showed a large well-encapsulated complex cystic mass with internal calcification, measuring 16 x 17 x 11.6 cm arising in the tail of pancreas with a small focus of calcification. Following multidisciplinary review she proceeded to a distal pancreatectomy and a splenectomy with the operative findings of a firm mass arising in the pancreatic tail involving the splenic artery and vein. She was discharged on day 6 and remains well 18 months post-operation.

Histological and immunohistochemical examination showed features consistent with solid pseudopapillary neoplasm, with areas of necrosis and haemorrhage with tumour cells arranged as discohesive nests and nuclear and cytoplasmic staining for vimentin and B-catenin. Solid pseudopapillary neoplasm is a rare primary pancreatic tumour that usually presents in young women with an abdominal mass and, while the primary tumours are often large, resection is usually feasible and long term survival expected.

Keywords: Solid pseudopapillary neoplasm; Pancreatectomy

Introduction

Solid tumours of the exocrine pancreas are usually characterised by aggressive biology with local invasion and metastatic disease common findings at presentation making effective treatment difficult and resulting in poor survival [1,2]. Solid pseudopapillary neoplasm (SPN) is an unusual tumour that develops from the exocrine pancreas and tumours are often over 10 cm in diameter at presentation. However metastatic disease is rare and complete resection is usually possible with excellent long term survival [3].

Case Presentation

A 35 year-old Samoan woman was referred with a six-month history of a painless epigastric mass and weight loss. She had no significant previous medical or surgical history and physical examination was unremarkable except for a firm mass in the epigastrium. Full blood count, biochemistry and tumour markers (carcinoembryonic antigen, Ca 19-9, Ca 125, chromogranin A) were all within the normal limits. A CT scan of the abdomen showed a large well- encapsulated complex cystic mass with internal calcification, measuring 16 x 17 x11.6 cm arising in the tail of pancreas (Figure 1). There were no signs of distant spread or regional lymphadenopathy.

Following multidisciplinary review she proceeded to a distal pancreatectomy and a splenectomy with the operative findings of a firm mass arising in the pancreatic tail involving the splenic artery and vein. No metastatic disease was seen and her post-operative course was uncomplicated.

Histological and immunohistochemical examination showed features consistent with SPN, with areas of necrosis and haemorrhage with tumour cells arranged as discohesive nests accompanied by small blood vessels imparting a papillary configuration (Figure 2). There was a nuclear and cytoplasmic staining for vimentin and B-catenin (Figure 3). The patient remains alive and well 18 months post-operation and under regular follow up.

Discussion

Tumours of the exocrine pancreas include adenocarcinoma, acinar cell carcinoma, intraductal papillary mucinous neoplasm and mucinous cystadenocarcinoma. They are generally biologically

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Correspondence:
Jonathan Koea, Department of Surgery, North Shore Hospital, New Zealand, Tel: 649 486 8900; Fax: 649 488 4621, E-mail: jonathan.koea@waitematadhb.govt.nz

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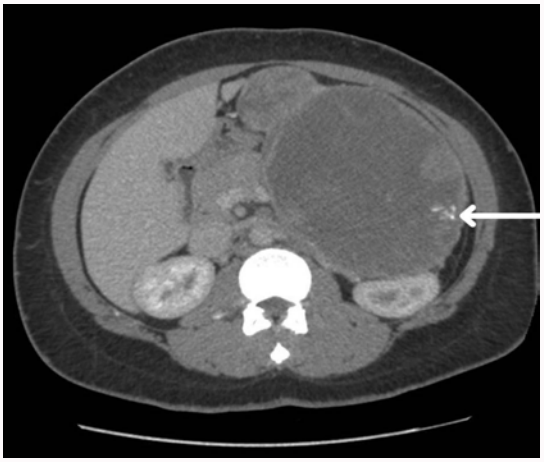

Figure 1: Axial CT scan demonstrating a large, heterogeneous soft tissue mass arising from the pancreas and containing a small area of calcification (arrow).

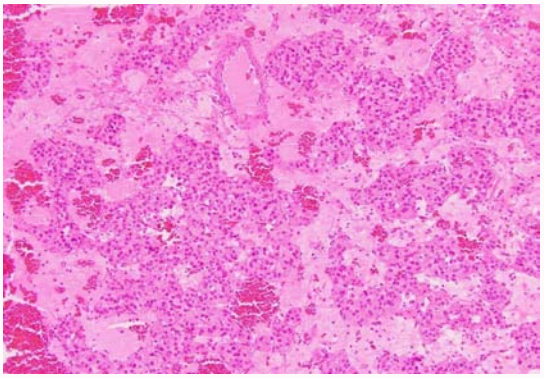

Figure 2: High power microscopy showing areas of necrosis and haemorrhage with tumour cells arranged in a papillary configuration associated with small blood vessels (haematoxylllin and eosin, 500 x magnification).

aggressive and associated with poor survival. Solid pseudopapillary neoplasm develops from the exocrine part of the pancreas [1] but is an indolent tumour. It was first described by Virginia Frantz [2] and historically it accounts for 1% of all the pancreatic neoplasms, 5% of the cystic neoplasms and presents most commonly in women in their 30s but can occur between the ages of 13 and 60 years [3]. The tumours are usually large at presentation and presenting symptoms are related to the presence of a mass and early satiety [3]. The diagnosis is confirmed with typical imaging features demonstrating large encapsulated tumours with solid and cystic spaces [4]. Although pseudopapillary neoplasms are often large they are generally localised although between 9-15% may present with metastases or signs of local invasion [1-4]. Routine preoperative biopsy is not recommended as it is of limited utility and may result in dissemination [4]. SPN can

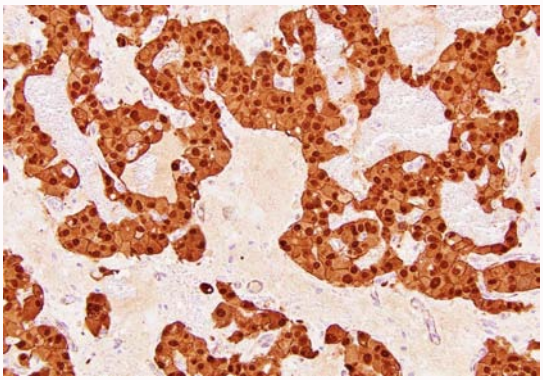

Figure 3: Immunohistochemical staining for β-catenin demonstrating nuclear uptake (500 x magnification).

occur throughout the pancreas but up to 60% develop in the tail or body. Treatment is surgical resection and the outcome following complete resection is excellent with overall 5 year survival of over 95% [5].

SPN are an increasingly important pancreatic lesion. A recent systematic review has shown that 90% of all cases have been reported in the 12 years and, of these 90% are now initially diagnosed as an incidental finding on cross-sectional imaging. Consistent with more early diagnosis is the observation that the mean size at presentation has decreased from 10 cm to 8 cm in the last decade [6]. These findings suggest that SPN is more common than initially thought and will become a significant part of the caseload for the pancreatic surgeon.

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