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## Pancreatic pseudopapillary tumour: A rare misdiagnosed entity

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## ABSTRACT

**INTRODUCTION:** Solid pseudo papillary pancreatic tumour is a rare entity. The atypical presentation causes a delayed or misdiagnosis of these pathology. It commonly affects the female population in the 2nd and 3rd decade of life. The presentation varies from non-specific abdominal pain to incidental findings in asymptomatic patients. It is a low-grade premalignant condition that is curable by excision of the tumour.

**PRESENTATION OF CASE:** This paper presents a 17-year-old girl with intra-abdominal mass diagnosed with solid pseudo papillary tumour that underwent Whipple's procedure.

**DISCUSSION:** We discuss the presentations, diagnosis and pathology findings of this rare pathology.

**CONCLUSION:** The diagnosis remains an enigma in view of the nature and location of the tumour. Resection is still the best choice remains for this condition.

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## 1. Introduction

Solid pseudopapillary pancreatic tumour is rare pathology. The scarce cases throughout the world make the diagnosis difficult and often missed. It affects mostly the female population. They present in early adult life especially in the 2nd and 3rd decades. It is a tumour with good prognosis and rarely metastases. Nevertheless, it has been shown to be premalignant in nature. It is difficult to diagnose as the presentation varies from non-specific abdominal pain to incidental findings in asymptomatic patients. It can also mimic the presentation of an adenocarcinoma. This makes it unusually difficult to manage especially when it involves an extensive surgical intervention. Diagnoses are mainly based on radiological evidence and occasionally tissue biopsy. The common location in the pancreatic head and tail often necessitates surgical intervention.

## 2. Case report

A 17-year-old girl came to us with non-remittance right-sided abdominal pain for 3 years duration. The pain was persistent and progressive, non-radiating with no relieving factors. No fever was documented. Systematic reviews were unremarkable. There were no constitutional symptoms identified. On physical examination, she was pink with no jaundice. Her abdomen was soft with

evident of mass at the right hypochondrium palpable. The mass was vague with no definable margin. It was immobile and appears not fixed to the skin. It was enhanced with stooping forward. Only mild tenderness was elicited. No rebound tenderness felt. Bowel sounds were audible with no other significant findings. Her routine blood investigations were normal with no obvious raised inflammatory marker. Liver function test and body fluid amylase were within normal range. Tumour markers were negative.

Computed tomography scan revealed a cystic lesion in the head of pancreas. No solid or multi-septated lesion seen. It appears resectable with no distant metastases. A decision for resection was made. No preoperative biopsy was performed in view of the possible nature of the lesion and to prevent any seedlings that might be introduced. A 6 cm × 5 cm × 7 cm mass were removed intra-operative. The mass evidently arises from the head of pancreas with presence of lymph nodes at the peripancreatic region. An R0 resection was performed with lymphadenectomy done for staging purpose. Liver and peritoneum were clear of metastases. Post-operatively were uneventful. Oral intakes were recommenced on day 2 onwards with drains removed on day 5. She was ambulating well by day 3. There were no delay in postoperative recovery and was sent home on day 7 with no complication.

Histological review of the tumour revealed a well-circumscribed mass with capsule invasion. Cut section shows a heterogenous, necrotic and haemorrhagic surface with central cavitation. Fortunately, the margins were clear of tumour. Microscopically, the presence of pseudo papillary structure confirms solid pseudo papillary tumour. There were no pleomorphism or mitotic activity seen. Immunohistochemistry test were positive for vimentin and CD10 and negative for chromogranin A.

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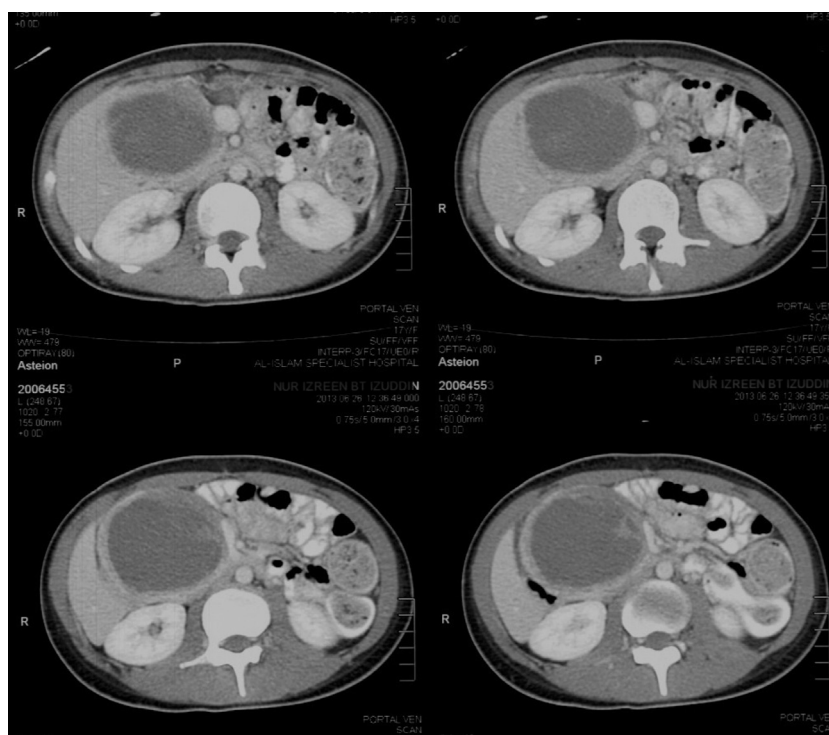


Fig. 1. CT shows a large cystic mass arising from head of pancreas compressing the stomach and abutting the liver.

### 3. Discussion

Solid pseudopapillary tumour (SPT) is an uncommon entity. Frantz first described it in 1959 as its own entity 'benign' or 'malignant'.<sup>1</sup> Among other names associated with it includes solid and papillary epithelial neoplasm, papillary cystic neoplasm, papillary epithelial neoplasm and solid and cystic acinar cell tumour to name a few. In 1996, WHO has reclassified it as SPT of the pancreas.<sup>2</sup>

This lesion usually occurs in young females although there are reported incidents among the male and elderly. It is a tumour with low malignant potential and fair better even after metastases, which occurs in 15% of cases.<sup>3</sup> Most common sites were at the tail of pancreas (35.9%) followed by the head (34%) and other parts of pancreas.<sup>4</sup> Patient presents with variable clinical presentations, which include abdominal discomfort, abdominal pain, intra-abdominal mass and arrays of constitutional symptoms. Jaundice has been reported but it is a rare presentation of SPT and usually a sign in a more sinister diagnosis. Diagnosis of this entity remains an enigma with adenocarcinoma a more common finding associated with pancreatic tumour. Although adenocarcinoma predominantly involves elderly, but the occurrence in the younger population has been documented.<sup>5,6</sup> Histories in both pathological entities are usually similar with or without abdominal pain. They may present incidentally or came with variation of symptoms. Adenocarcinoma is commonly associated with constitutional symptoms, but in early stages of the disease, it may not be apparent hence the difficulty of determining the nature of tumour. Physical examination may reveal variable findings from a simple malnutrition, mass, and even unremarkable. This unfortunate young lady presents with a prolonged and devastating abdominal pain. She denies the presence of other possible related symptoms. No constitutional symptoms and signs were found on history or physical examinations. However, a possible retroperitoneal mass was found during the physical examination. The presence of palpable retroperitoneal mass was perturbing in view of possibility of an advanced pancreatic tumour.

We proceeded with imaging for her immediately after all routine blood and tumour markers were taken. Previous literatures were clear with regards to the association between SPT's tumour and blood investigations. Tumour markers were nearly always negative and no inflammatory markers have been proven to have any direct association with this condition.<sup>4</sup> Nevertheless, the routine blood and tumour markers may point us to other possible sinister lesion such as adenocarcinoma, metastatic and inflammatory lesions.

The choices of imaging are usually straightforward. Ultrasonography has limited role especially in retroperitoneal tumour. It may assist in isolating the type of lesion and possible of origin and also identifying liver or other solid organ metastases but does not give extra information for surgery preparation. Ultrasonographically, SPTs tumours may appears as well-defined hypoechoic solid masses, solid in cystic lesion, cystic mass, and calcified lesions.<sup>7,8</sup>

We chose CT scan as mode of imaging. Literatures have shown that computed tomography and magnetic resonance imaging remains the most specific in the diagnosis preoperatively. CT usually demonstrates a well-encapsulated mass with mixture of solid and cystic components of variable degree and enhanced solid portions in its periphery after contrast administration.<sup>7–9</sup> This is useful in planning of her surgery to maintain a R0 resection. It helps in identifying the size, depth of penetration; define the pancreatic anatomy and also invasion to surrounding structures. CT of this patient shows a cystic lesion over the head of the pancreas causing compression to the stomach and abutting the liver. The rest of the pancreas appears clear from this lesion (Fig. 1).

We deterred from pursuing any biopsies based on several factors; there is high chance of a potential premalignant lesion and prevention of any tumour seedlings during the procedures. This is imperative in maintaining the curative intention. However, biopsies were performed as a routine in few literatures.<sup>10,11</sup> Others chose surgery as the initial intervention upon diagnosis with radiological imaging.<sup>12,13</sup> Nevertheless, most of these papers were case series at best. No randomized trial has been done with regards to

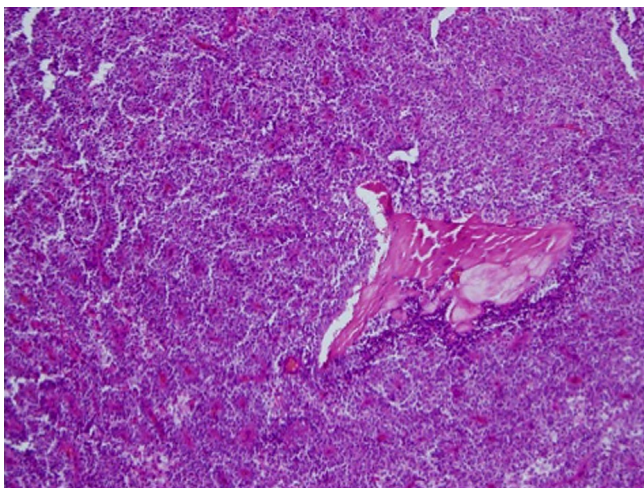


Fig. 2. Solid area within the cystic lesion.

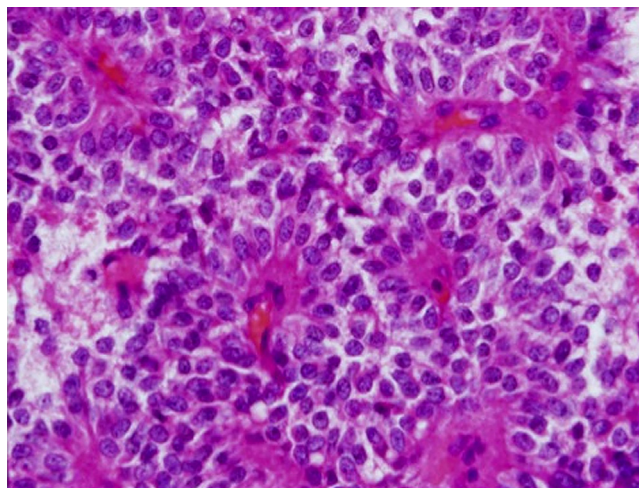


Fig. 4. Monomorphic with uniform, round to oval nuclei, inconspicuous nucleoli and eosinophilia or clear vacuolated cytoplasm.

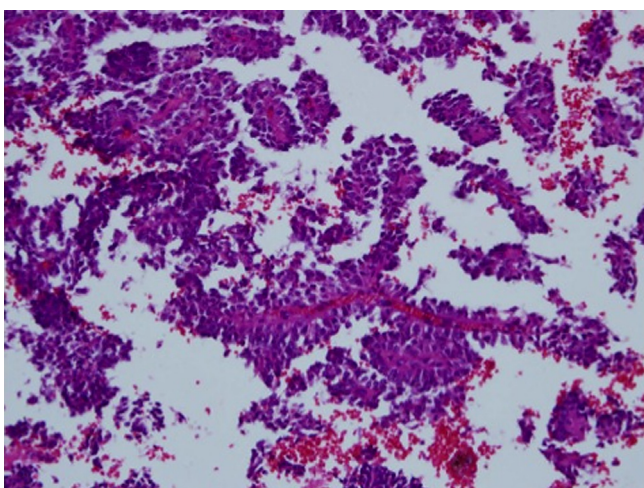


Fig. 3. Pseudo papillary pattern.

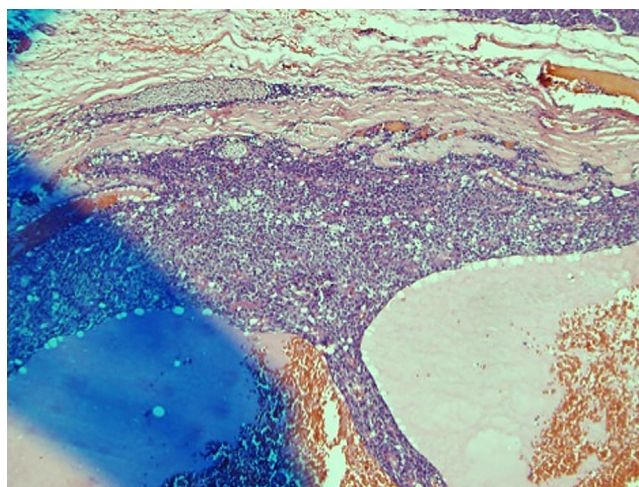


Fig. 5. Capsular invasion (high power magnification).

this matter hence the conflicting role of biopsies for diagnosis. A review article of 718 patients was performed which highlight these issues. The results showed only 52 patients were diagnosed with biopsy.<sup>4</sup> The remaining patients were advised for surgery based on history, physicals and radiological findings alone. In our patient, a preoperative biopsy probably would not give any extra information as a decision for surgery has been made in view of the radiological appearance and possible nature of this premalignant tumour.

On gross histopathology examination, SPTs is usually well circumscribed with a well-defined margin while adenocarcinomas are poorly defined and irregular. However, this is not definitive in view of the similar location and appearance. A microscopic evaluation and staining is required for definite diagnosis.

The tumour was well circumscribed with cross section revealed a heterogeneous mixture area of necrosis and haemorrhages with central cavitation. The microscopic examination demonstrates a mass surrounded by thick fibrous capsule composed of neoplastic cells arranged in solid and pseudo papillary structures with endypmal rosette formation (Figs. 2–4).

Individual cells shows monomorphic with uniform, round to oval nuclei, inconspicuous nucleoli and eosinophilia or clear vacuolated cytoplasm. Capsular invasion and peri-neural involvement are evident in this specimen raised a suspicion of possibility of malignant transformation. Fortunately for her, the margins were free of tumour (Fig. 5). Immunohistochemically, the neoplastic cells

are positive for vimentin and CD10, which are similar findings in most of the literatures. Nonetheless, this positive finding is not pathognomonic of SPTs.

Complete resection of tumour remains the mainstay for curative treatment. Resections control the symptoms and progression of this rare premalignant condition. Local recurrence and metastases are rarely recorded; a review article revealed a recurrence rate of 6.63% for 10 years follow up with the liver and lymph nodes as the preference sites of metastases.<sup>5</sup> Nevertheless, all patient requires long-term surveillance of late metastases or recurrence.

#### 4. Conclusion

Solid pseudopapillary tumour of pancreas is a rare but curable disease. Its rarity posed a difficulty in diagnosis and exclusion of a malignant or benign disease. A definitive diagnosis can only be arrived upon histopathology and immunohistochemical examination. Resection remains the mainstay in treating this disease in view of its premalignant potential.

#### Conflict of interest

None declared.

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**Ethical approval**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

**Author contributions**

Affirul Chairil Ariffin: writing and correction; Nik Qjsti Fathi: patient identification and selection; Zamri Zuhdi: data and case history collection; Azlanuddin Azman: histopathology and radiology data collection; Hairul Azrin Othman: discussion and interpretation; Razman Jarmin: final review and correction.

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