Solid-pseudopapillary tumour of the pancreas: A report of a rare case

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ABSTRACT

The solid pseudopapillary tumour (SPT) of the pancreas is an uncommon cystic pancreatic neoplasm mostly seen in young females. We present a large SPT in a 16-year-old female who presented with acute onset upper- and mid-quadrant abdominal pain associated with nausea and vomiting for 4 days. The lesion was demonstrated by abdominal ultrasound and by computed tomography, which was successfully removed surgically.

Key words: Pancreas, pancreatic neoplasm, solid pseudopapillary tumour

INTRODUCTION

Solid pseudopapillary tumour (SPT) of the pancreas is a rare cystic pancreatic neoplasm that accounts for only 1–2 % of all exocrine pancreatic tumours.[1] This tumour was first described in 1959 by Frantz. Synonyms of this tumour include papillary epithelial neoplasm, papillary cystic neoplasm, solid papillary neoplasm, solid cystic neoplasm and low-grade papillary neoplasm of the pancreas.[2] SPT occurs predominantly in the young female. We report a 16-year-old female with a large SPT demonstrated by computed tomography (CT), which was successfully removed surgically.

CASE REPORT

A 16-year-old female presented with constipation and vomiting for 4 days. She had acute onset upper- and mid-quadrant abdominal pain associated with nausea and vomiting for 4 days. The pain was intermittent in nature, with an acute onset and sometimes radiating to the back. She also complained of vomiting containing food particles without any evidence of blood or bile staining. There was no history of fever, jaundice, per rectal bleeding or urinary symptoms. She denied any history of trauma and there was no history of loss of weight or appetite.

On examination, a poorly nourished young girl with mild pallor was noted. There was no jaundice or lymphadenopathy. Her vitals were stable. Abdominal examination revealed a tendered mass, firm to hard in consistency, noted on the right side of the umbilicus occupying the epigastric and the right hypochondriac region. The mass had free horizontal and restricted vertical mobility. Blood investigation revealed haemoglobin of 10 gm/dl (reference range- 12–18 gm/dl) and serum amylase of 1297 U/L (reference range- up to 50 U/L). Serum lipase, liver function tests and pancreatic tumour markers were within normal limits. Abdominal ultrasound revealed a mixed echotexture mass [Figure 1] in the subhepatic region anterior to the right kidney within the C-loop of the duodenum showing peripheral vascularity. Plain and contrast-enhanced CT of the abdomen revealed a well-circumscribed, rounded, cystic lesion measuring 6.5X5.6 cm within the C-loop of the duodenum causing minimal narrowing of the lumen of the second part of the duodenum [Figures 2 and 3]. There was no evidence of calcification in the lesion; however, there was peripheral rim enhancement of the lesion on postcontrast study. Based on the CT and the ultrasound findings, differential diagnosis of duplication cyst and cystic neoplasm of the pancreas were considered.

Laparotomy was performed using supraumbilical right transverse incision. Duodenum and pancreas preserving complete enucleation of the cystic mass [Figure 4] was performed. Her postoperative recovery was excellent. The histopathological examination of the resected mass revealed an SPT of the pancreas [Figure 5].

DISCUSSION

SPT of the pancreas accounts for less than 4% of the pancreatic cystic tumours.[3] The rare SPT occurs more frequently in females of age between 15 and 35 years.

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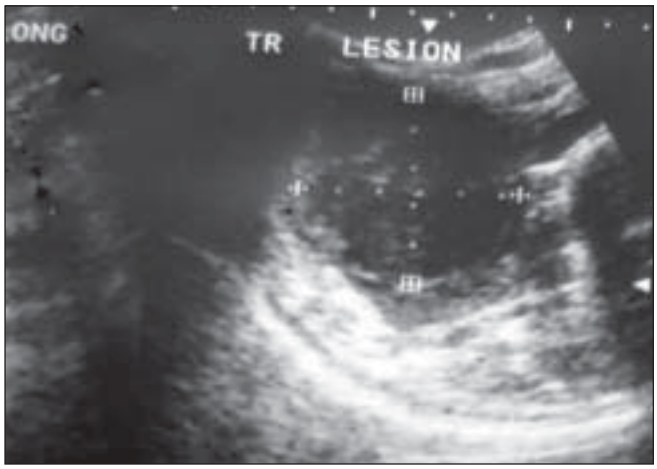


Figure 1: Ultrasound image showing a well-defined, mixed echotexture mass in the subhepatic region anterior to the right kidney within the C-loop of the duodenum.

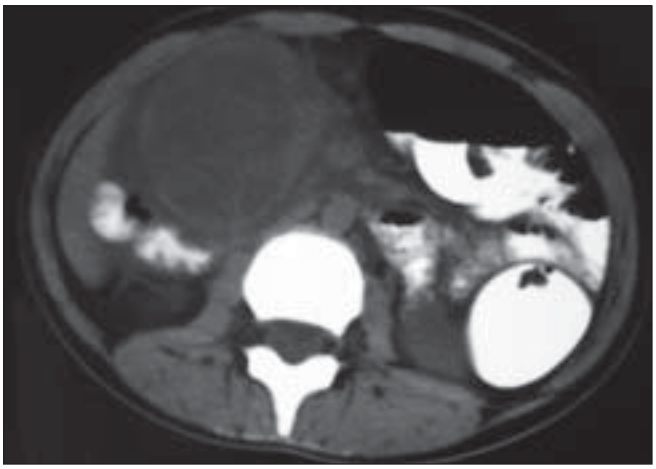


Figure 2: Unenhanced axial computed tomography image shows a well-defined mass within the C- loop of the duodenum causing minimal narrowing of the lumen of the second part of the duodenum

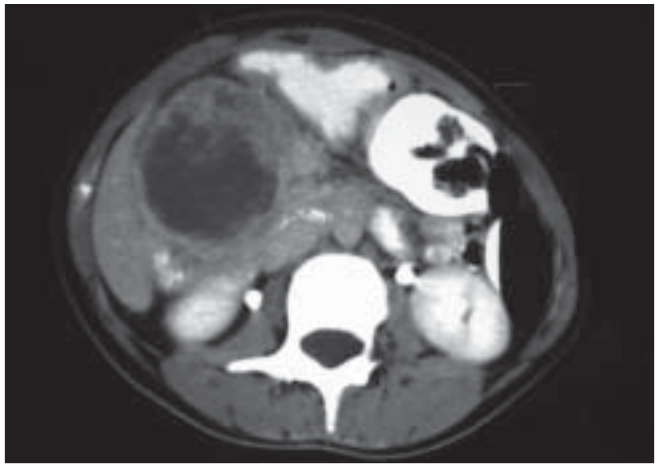


Figure 3: Contrast-enhanced axial computed tomography image shows a well-defined mass with cystic and enhancing solid components within the C-loop of the duodenum

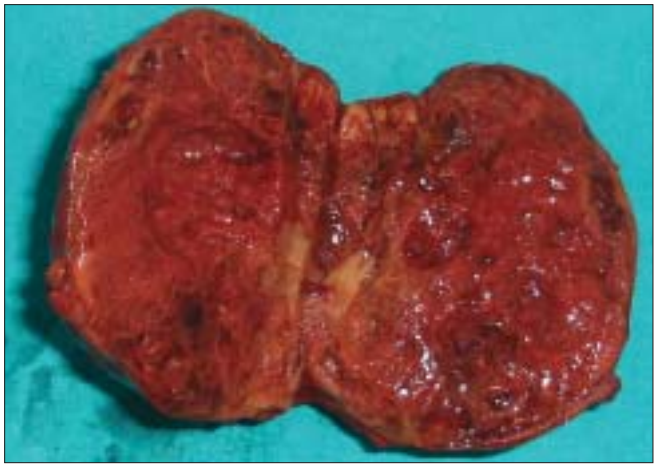


Figure 4: The cut surface of the resected tumour shows a well-circumscribed solid mass with central necrosis

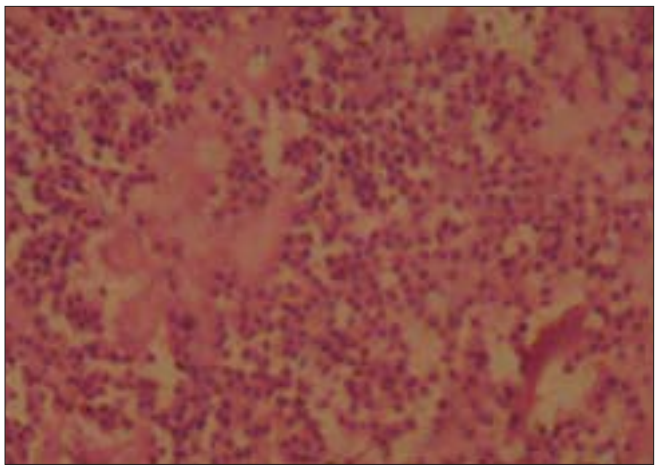


Figure 5: Microscopic features showing a pseudopapillary pattern with cores of stroma. Small uniform tumour cells with round nuclei and eosinophilic cytoplasm are noted

The mass may occur anywhere in the pancreas but is most frequently found in the head or the tail of the pancreas.[4]

Lack of specific clinical symptoms is typical of this tumour. Many tumours are detected incidentally, but may occasionally present with sudden pain due to bleeding within the tumour or with symptoms related to the compression of adjacent organs.[3] They are frequently diagnosed during investigation of gastrointestinal complaints such as abdominal pain, weight loss, nauseas, vomiting or abdominal mass. Our patient was presented with acute onset upper- and mid-quadrant abdominal pain, vomiting and constipation.

In ultrasound, well-encapsulated cystic and solid masses are typically seen, but sometimes masses are pure solid looking or have internal septa or calcification.[4] CT demonstrates a well-marginated lesion with solid and cystic components. Magnetic resonance imaging typically demonstrates a well- defined lesion with heterogenous signal intensity on T1 and T2 weighted images, which reflects the complex nature of the mass.[4] The diagnosis of SPT is often made only at laparotomy

because of the lack of specific laboratory tests and imaging studies.[5] Common differential diagnosis include pancreatoblastoma, pancreatic pseudocyst or duplication cyst. SPT posses a malignant potential risk of 5-10% and must therefore be resected completely and aggressively as there are no prognostic factors to distinguish between SPT with or without a malignant potential.[6] Radical resection, where technically feasible, should be considered the therapy of choice as it is a safe and effective control of the disease.[7] Unlike pancreatic ductal adenocarcinoma, surgical resection often results in cure and long disease-free periods even in patients who have recurrence of metastases.[3] The various surgical procedures are whipple's operation, pylorus preserving pancreaticoduodenectomy, distal pancreatectomy with or without splenectomy, enucleation and excision.[7] In our case, complete excision of the tumour was performed.

Macroscopically, they are well-encapsulated tumours that contain solid and cystic areas consisting of haemorrhagic and central necrotic areas. SPT contain both solid and pseudopapillary patterns, which give rise to the designation of solid pseudopapillary pancreatic tumour.[5] SPT of the pancreas show strong cellular immunoreactivity for vilamentin and focal weak keratin reactivity.[3]

In conclusion, SPT is a rare low-grade slow-growing malignant tumour of the pancreas, which usually occurs in the young female. Surgical resection is the treatment of choice and offers excellent result even in the case of metastases.

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Source of Support: Nil, **Conflict of Interest:** None declared.

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