J HK Coll Radiol 2002;5:50-53

CASE REPORT

Solid-cystic Pancreatic Tumour (Frantz's Tumour) — An Unusual Spleno-renal Angle Tumour in a Teenager

PSF Lee, ASC Ching, YL Chan

Department of Diagnostic Radiology and Organ Imaging, Prince of Wales Hospital, Hong Kong

ABSTRACT

We report a rare case of solid-cystic pancreatic tumour presenting as a spleno-renal angle mass in a 14-year-old girl. Transabdominal ultrasound showed a well-capsulated mass with irregular heterogeneous echogenic foci intermixed with small hypoechoic components in a bizarre pattern. The interactive roles of ultrasound in lesion localisation, characterisation, and guidance for percutaneous needle biopsy are described. Differentiating this rare, low grade malignant paediatric tumour from other lesions in the spleno-renal angle is essential for both surgical planning and prognosis.

Key words: *Neoplasms, Pancreas, Ultrasonography*

INTRODUCTION

Pancreatic tumours are relatively rare in paediatric patients,[1,2] and solid-cystic pancreatic tumours are among the rarest of all pancreatic tumours. First described in 1959 by Frantz et al,[3,4] solid-cystic pancreatic tumour is more prevalent in girls. Moreover, despite histological signs of malignancy, they have a favourable clinical course. Ultrasound (US) has been reported to be useful in the evaluation of pancreatic pathology in childhood.[5] This report illustrates a case of solid-cystic pancreatic tumour and the value of US in reaching a final diagnosis of this rare entity. The diagnostic role of percutaneous needle biopsy is also discussed.

CASE REPORT

A 14-year-old girl with a past history of good health was admitted to our institution on account of acute, severe abdominal pain and bile-stained vomiting. On clinical examination, a mass was palpated in the left subcostal region, which was thought to be splenomegaly. The results of haematological and biochemical studies were unremarkable apart from a slightly elevated white cell count. An abdominal radiograph revealed a relative paucity of gas shadow at the left upper quadrant. No calcification or dilated bowel loop was observed.

A water-soluble contrast meal and follow through was subsequently performed to rule out upper gastrointestinal tract obstruction. It showed a soft tissue mass indenting on the greater curvature of the stomach, pushing it anteriorly, and displacing the transverse colon caudally (Figure 1). No definite intraluminal defect or mucosal irregularity was noted.

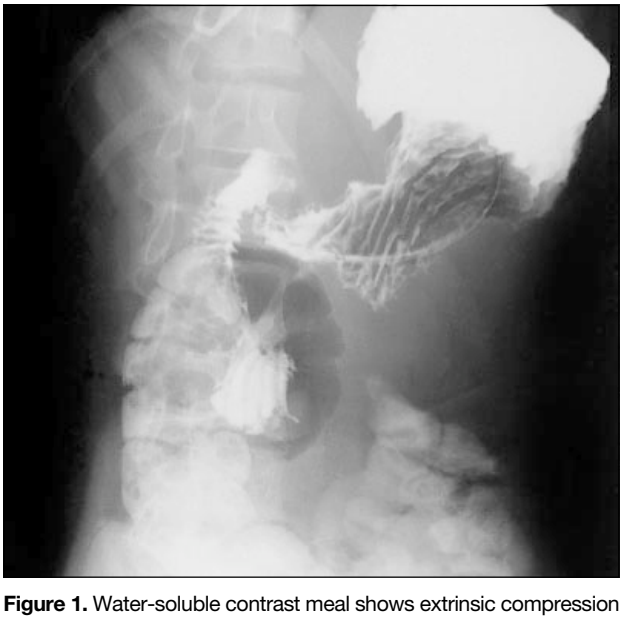

Figure 1. Water-soluble contrast meal shows extrinsic compression along the greater curve of the stomach.

Correspondence: *Dr. PSF Lee, Department of Diagnostic Radiology and Organ Imaging, Prince of Wales Hospital, 30-32 Ngan Shing Street, Shatin, New Territories, Hong Kong.*
Tel: (852) 2632-2277; Fax: (852) 2648-4122;
E-mail: leesfp@hotmail.com

Submitted: 31 October 2001; Accepted: 17 December 2001.

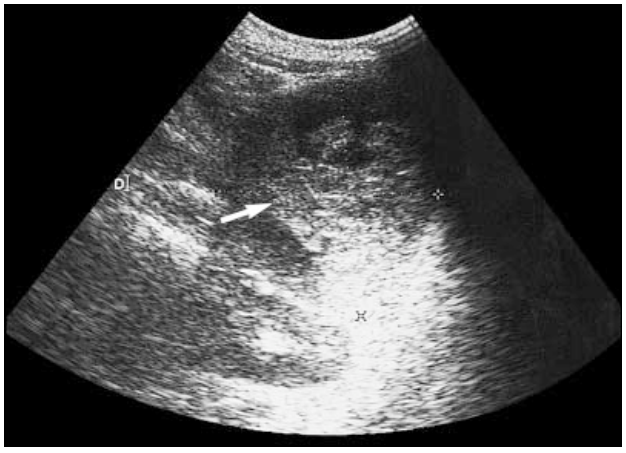

Figure 2. Ultrasound of the left upper abdomen in an oblique coronal plane shows a circumscribed complex cystic mass arising from the left spleno-renal angle.

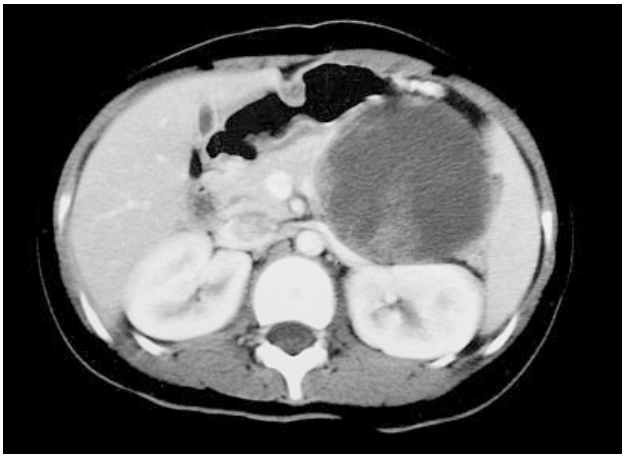

Figure 3. Contrast axial CT scan of the upper abdomen reveals a huge cystic mass with a solid element arising from the pancreatic tail. No vascular encasement or retroperitoneal lymphadenopathy is detectable.

US examination was then carried out. Transabdominal US (ATL HDI 3000, Bothell, USA) using a 3.5 MHz sector probe revealed a complex cystic mass measuring 7 x 8 x 9 cm at the spleno-renal angle. Its contents displayed a bizarre pattern, with heterogeneous echogenic foci intermingled with small hypoechoic areas (Figure 2). The mass had a well-defined margin in close proximity to the pancreatic tail, and the left adrenal gland could not be visualised. A clear plane separating the mass and the adjacent spleen was present. The left kidney was displaced caudally. The mass did not appear hypervascular on colour Doppler.

In view of these findings, the origin of the mass seemed to be either the pancreatic tail or the left adrenal gland. The preliminary sonographic diagnosis, therefore, was a cystic tumour arising from the pancreatic tail or left adrenal gland. The pancreatic origin of this tumour was later proven on contrast-enhanced CT. It appeared as a well-circumscribed mass and was predominantly low attenuated. The mass was separate from the adjacent stomach, spleen, and left kidney, although it elevated the splenic vessels and compressed the left renal vein (Figure 3). The adrenal glands appeared normal. No lymphadenopathy or other pathology was seen in the abdomen or retroperitoneum. Although the lesion appeared non-aggressive in nature on radiology, malignant growth could not be excluded.

US-guided percutaneous biopsy of the lesion was performed under local analgesia. To avoid repeated percutaneous punctures of the lesion, and for better haemostasis control, a 19G coaxial needle was advanced into the centre of the mass from where 80 mLs of altered blood was aspirated. Then, a preloaded 20G coaxial (Temno) needle was inserted into the solid element of the mass for multiple tissue core biopsies. No obvious haemorrhage was encountered, and overall the procedure was uneventful and well tolerated. The retrieved tissue core specimens were fixed in formalin and sent for study. Histological examination of the same revealed the presence of epithelial neoplasm, suggesting a solid-cystic papillary tumour of the pancreas.

One week later, an elective laparotomy was performed. A 9 cm cystic mass was identified at the tail of the pancreas, with no evidence of local infiltration. Distal pancreatectomy was performed. Macroscopically, the pancreatic tumour was enveloped by a fibrous capsule and a thin rim of pancreatic tissue. Tumour incision revealed a solid-cystic lesion containing haemorrhagic and necrotic debris. The diagnosis of solid-cystic pancreatic tumour was confirmed on histopathological and immunochemical examinations.

The patient's postoperative course was uneventful. Twelve months later, she was well and had no evidence of disease on either clinical examination or US.

DISCUSSION

Solid-cystic pancreatic tumours account for only 0.3% of pancreatic tumours.[6] This type of tumour occurs predominantly in girls and young women, but sporadic cases in men and the elderly have also been reported.[4,7,8] Eighty four percent of patients diagnosed with this tumour are less than 35 years of age.

Solid-cystic pancreatic tumours are controversial with respect to histogenesis and malignant potential.

This uncertainty is partly reflected in the many synonyms used to describe them, such as solid and papillary epithelial neoplasms, solid and cystic acinar cell tumours, and solid-cystic-papillary neoplasms.[4] The ease with which solid-cystic pancreatic tumours can be misdiagnosed as nonfunctioning islet cell tumours, or as cystadenomas or cystadenocarcinomas, is well recognised.

Solid-cystic pancreatic tumours are most often located in the tail of the pancreas (in 58% of cases), followed by the head (38%), and the body (2%).[6] They present as a slowly growing asymptomatic mass (65%), which frequently accounts for the delay in diagnosis. Macroscopically, the mean size of the tumour at presentation is 10 cm. It has a thick fibrous capsule with solid and cystic components intermixed with foci of haemorrhage and necrosis in various proportions. The key histological hallmarks are solid and pseudopapillary or pseudo-glandular proliferation with no mitoses, or atypism of the tumour cells in the solid portion. This characteristic polymorphic differentiation suggests that solid-cystic pancreatic tumours derive from multipotential primordial cells rather than acinar cells as was previously thought.[9,10] Although the tumour shows signs of malignancy on histology, local invasion and distant metastasis have rarely been reported.[7,11] Indeed, solid-cystic pancreatic tumours usually run a benign course with a good prognosis after complete surgical excision.

From an imaging viewpoint, it is not uncommon for a solid-cystic pancreatic tumour to present as a solitary mass sitting along the lienorenal ligament between the spleen and left kidney. It is well known that lesions in this region can be easily misinterpreted with regard to both origin and nature.[12] Tumours in the spleno-renal region can arise from the spleen, kidney, adrenal gland, splenic vessels, gastric wall, and splenic flexure, as well as the pancreatic tail. Disease entities such as protuding splenic abscess, haemorrhage in an adrenal cyst, pancreatic mass complex, pedunculated renal tumour, leiomyoma or leiomyosarcoma from the stomach, and (rarely) mesenteric cyst complicated with haemorrhage, may present as a solitary heterogeneous mass at the spleno-renal angle. Real time dynamic US can serve as an effective means to narrow the differential diagnosis list. Displacement of constant anatomic landmarks, such as the splenic angle and the left kidney, already narrow the differentials and point to a lesion in the pancreatic tail.

The variable appearance of solid-cystic pancreatic tumours on US depends on the balance between the intrinsic regulation of tumoural integrity and the extent of spontaneous retrogression.[13] The two most common patterns are a well-defined hyperechoic mass and a well-defined, hyperechoic mass with echopoor areas. Lesions with mixed anechoic and hypoechoic areas, hypoechoic mass with calcification, or isoechoic mass are infrequent findings.[14,15]

The differential diagnosis of pancreatic lesions with similar sonographic features includes: exocrine tumours, such as pancreatoblastoma, mucinous cystic neoplasm of the pancreas, and serous marcocystadenoma; endocrine tumours, such as islet cell tumour with cystic transformation; and miscellaneous lesions, such as cystic lymphangioma and pseudocyst. Pancreatoblastoma is a relatively common paediatric pancreatic tumour, and its salient features of diffuse infiltrative hypoechoic nature with calcific foci and vessel encasement are unlike those of solid-cystic pancreatic tumour.[16] Pancreatic cystic tumours including the remaining exocrine tumours mentioned above, i.e. mucinous cystic pancreatic neoplasm, serous marcocystadenoma, and cystic degeneration of islet cell tumours, are rarely encountered in children. They usually present as unilocular or paucilobular masses with echopoor areas and may or may not be associated with hyperechoic areas in older patients. Cystic lymphangioma rarely occurs in the pancreatic tail and is characteristically multi-loculated. A pseudocyst may have a complex hypoechoic and hyperechoic appearance if complicated by haemorrhage or infection. For this lesion, the diagnosis is usually suspected clinically from a history of pancreatitis.

Our patient's tumour had a comparable appearance on US to those described for solid-cystic pancreatic tumours in the published radiological literature. The heterogeneously echogenic areas with scattered echopoor foci probably represent solid and haemorrhagic/necrotic components. Cystic and haemorrhagic necrosis, which are characteristic of this tumour, appear on US as areas of high or low echogenicity. The solid component and well-defined margin seen on US correlate with, respectively, the partially solid component and well-formed capsule on pathology.

In previously reported cases,[4,8,11] open surgery or exploratory laparotomy were required to establish the diagnosis. The role of percutaneous needle biopsy has thus far not been emphasised. However, as illustrated

in this case, preoperative percutaneous needle biopsy with ultrasound guidance can be done safely under simple local analgesia. A histologically proven diagnosis of solid-cystic pancreatic tumour is useful for surgical planning and can be obtained preoperatively using US-guided percutaneous needle biopsy.

CONCLUSION

Although rare, the possibility of solid-cystic pancreatic tumour should be considered in children presenting with a solitary, heterogeneous, pancreatic mass. US imaging and guided biopsy provide prompt and appropriate information for further management and surgical treatment.

REFERENCES

1. Jaksic T, Yaman M, Thorner P, Wesson DK, Filler RM, Shandling B. A 20-year review of pediatric pancreatic tumours. J Pediatr Surg 1992;27:1315-1317.
2. Grosfeld JL, Vane DW, Rescoria FJ, McGuire W, West KW. Pancreatic tumors in childhood: Analysis of 13 cases. J Pediatr Surg 1990;25:1057-1062.
3. Todani T, Shimada K, Watanabe Y, Toki A, Fujii T, Urushihara N. Frantz's tumour: A papillary and cystic tumor of the pancreas in girls. J Pediatr Surg 1988;23:116-121.
4. Herskovits M, Cohen I, Loberant N, Szavalb S. Papillary cystic neoplasm of the pancreas in a teenage boy. Eur Radiol 1999;9: 1354-1356.
5. McCain AH, Berkman WA, Bernardino ME. Pancreatic sonography: past and present. J Clin Ultrasound 1984;12:325-332.
6. Sclafani LM, Reuter VE, Coit DG, Brennan MF. The malignant nature of papillary and cystic neoplasm of the pancreas. Cancer 1991:68:153-158.
7. Klöppel G, Maurer R, Hofmann E, et al. Solid-cystic (papillary-cystic) tumours within and outside the pancreas in men: report of two patients. Virchows Arch A Pathol Anat Histopathol 1991; 418:179-183.
8. Takahashi H, Hashimoto K, Hayakawa H, et al. Solid cystic tumour of the pancreas in elderly men: report of a case. Surg Today 1999;29:1264-1267.
9. Matsunou H, Konishi F. Papillary-cystic neoplasm of the pancreas: A clinicopathologic study concerning the tumor aging and malignancy of nine cases. Cancer 1990;65:283-291.
10. Mao C, Guvendi M, Domenico DR, Kim K, Thomford NR, Howard JM. Papillary cystic and solid tumors of the pancreas: a pancreatic embryonic tumor? Studies of three cases and cumulative review of the world's literature. Surgery 1995;118: 821-828.
11. Bombi JA, Milla A, Badal JM, Piulachs J, Estape J, Cardesa A. Papillary-cystic neoplasm of the pancreas: report of two cases and review of the literature. Cancer 1984;54:780-784.
12. Meyers MA. Dynamic radiology of the abdomen (5th edition). Heidelberg: Springer Verlag;2000:154-158.
13. Buetow PC, Buck JL, Pantongrag-Brown L, Beck KG, Ros PR, Adir CF. Solid and papillary epithelial neoplasm of the pancreas: Imaging-pathologic correlation in 56 cases. Radiology 1996; 199:707-711.
14. Friedman AC, Lichtenstein JH, Fishman EK, Oertel JE, Dachman AH, Siegelman SS. Solid and papillary epithelial neoplasm of the pancreas. Radiology 1985;154:333-337.
15. Yamaguchi K, Hirakata R, Kitamura K. Papillary cystic neoplasm of the pancreas: Radiological and pathological characteristics in 11 cases. Br J Surg 1990;77:1000-1003.
16. Gupta AK, Mitra DK, Berry M, Dinda AK, Bhatnagar V. Sonography and CT of pancreatoblastoma in children. AJR Am J Roentgenol 2000;174:1639-1641.