J Hong Kong Col Radiol. 2010;13:158-60

CASE REPORT

Pancreatic Neoplasm in a Patient with Situs Inversus Totalis

BK Paunipagar[1], DD Rasalkar[1], P Lonikar[2], WCW Chu[1]

[1]Department of Diagnostic Radiology and Organ Imaging, The Chinese University of Hong Kong, Prince of Wales Hospital, Shatin, Hong Kong, and [2]Shri Tirupati Diagnostic Center, Aurangabad, India

ABSTRACT

We reported on a patient with a cystic neoplasm of the pancreas with situs inversus totalis that showed the typical imaging features of serous cystadenoma in an anatomically inverted pancreas. The possible co-existence of malformations of transposed organs and anatomo-surgical situations require particular attention in diagnosis and preoperative evaluation.

Key Words: *Digestive system abnormalities; Pancreatic neoplasms; Situs inversus; Tomography, X-ray computed; Ultrasonography*

中文摘要

一位完全性內臟異位病人的胰腺腫瘤

BK Paunipagar、DD Rasalkar、P Lonikar、朱昭穎

本文報告一名有完全性內臟異位的病人患有胰腺囊性腺瘤。病人的影像顯示其「倒轉」了的胰腺出現漿液性囊腺瘤的典型特徵。如要為有內臟異位的病人進行手術，必須小心診斷並特別注意術前評估。

CASE REPORT

A 46-year-old female with congenital heart disease (dextrocardia, univentricle with eisenmenger's syndrome) and corrective shunt surgery performed 30 years earlier, presented with a recent history of anorexia, a dull aching abdominal pain, and a palpable abdominal mass in November 2009. The biochemical parameters were within normal limits. An ultrasound revealed an encapsulated multilocular cystic mass in the uncinate process of pancreas, measuring up to 2.5 cm in diameter (Figure 1). Additional features of situs inversus with liver and spleen located vice versa at the left and right upper quadrant were also noted. Further computed tomography of the abdomen substantiated the sonographic features showing a multi-septated cystic mass with measurable locules at the anatomically inverted pancreas (Figure 2a). Dextrocardia was also noted along with other organ transpositions (Figure 2b and 2c). Imaging features were compatible with a serous cystic neoplasm of pancreas in a patient with situs inversus.

DISCUSSION

Situs inversus totalis is a congenital anomaly, with an incidence of 1 in 10,000-50,000 live births.[1] The aetiology for the transposed viscera is obscure, but apparently this condition does not influence normal health or life expectancy. Associations with situs inversus include primary ciliary dyskinesia (also known as Kartagener syndrome).[2] About 58% of patients

Correspondence: *Dr DD Rasalkar, Department of Diagnostic Radiology and Organ Imaging, The Chinese University of Hong Kong, Prince of Wales Hospital, Shatin, New Territories, Hong Kong.*
Tel: (852) 2632 1189; Fax: (852) 2636 0012; Email: darshana@cuhk.edu.hk

Submitted: 9 Feb 2010; Accepted: 7 Apr 2010.

with this developmental abnormality have other malformations, such as duodenal atresia, biliary atresia, malrotation, and tracheoesophageal fistula.[3]

Very few case reports of pancreatic tumour with situs inversus have been published in the English literature,[4-6] all being in non-radiology journals and to our knowledge none reported any radiology. Imaging is an excellent non-invasive tool to demonstrate anatomy and pathology. Moreover, the imaging features of pancreatic tumours are often diagnostic.

Serous cystic neoplasm is a benign slowly growing pancreatic tumour arising from acinar cells, composed of innumerable small cysts containing proteinaceous fluid separated by connective tissue septae.[7,8] Usually it occurs in elderly women in their 5th to 7th decade, and can be encountered in any part of pancreas; 30% are in the pancreatic head.[7] Cysts range in size from 5 to 20 cm in maximum diameter and consist of innumerable small cysts (1-20 mm) within the cystadenoma. The so-called characteristic central sunburst calcifications are uncommon. In a study by Johnson et al,[9] a central scar was found in only two (13%) of 16 patients with microcystic adenomas.

Unlike mucinous macrocystic adenomas, microcystic adenomas are not premalignant and have characteristic imaging that can differentiate them from potentially malignant cystic tumours, such as mucinous cystic tumours and intraductal papillary mucinous neoplasms.[10,11] Furthermore, if asymptomatic, they do not require surgical removal.[8,12,13] Thus, making a

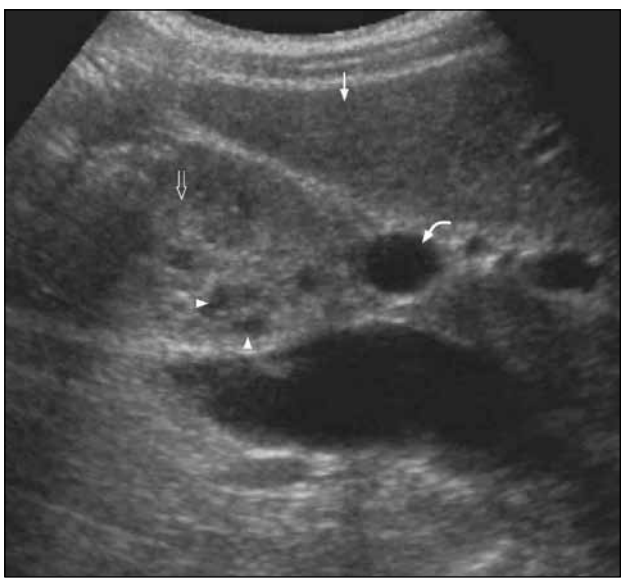

Figure 1. A grey-scale transverse transabdominal ultrasound of the patient showing an ill-defined hypoechoic mass in the pancreatic head (open arrow), which contains small microcysts (white arrowheads). Note part of the liver (white arrow) and part of the portal vein (curved arrow).

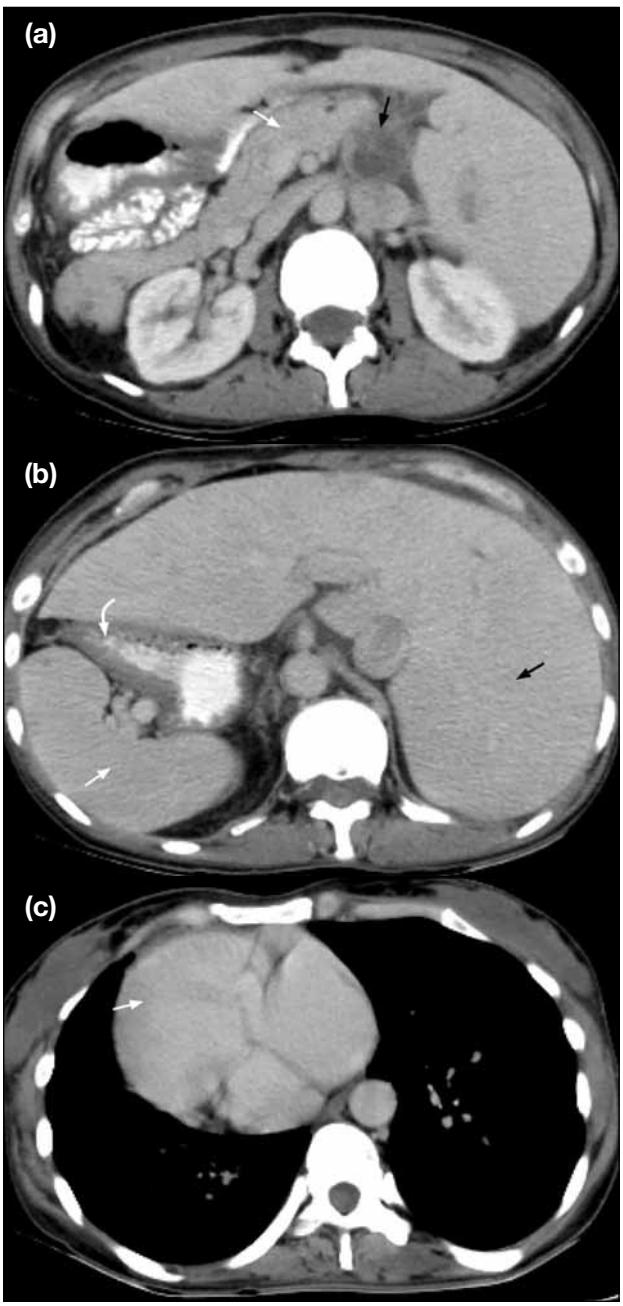

Figure 2. (a) A contrast-enhanced axial computed tomographic (CT) image shows an inverted pancreas (white arrow) with an encapsulated multi-septated cystic mass located at its head (black arrow). (b) A contrast-enhanced axial CT image of the abdomen showing visceral transposition in a known case of situs inversus. Note the right-sided spleen (white arrow) and stomach (white curved arrow). The liver is located on the left (black arrow). (c) A contrast-enhanced axial CT image of the thorax showing dextrocardia. Note the monoventricle (white arrow).

definitive diagnosis of microcystic adenoma is very important.

Serous cystadenomas have 3 morphological patterns: polycystic, honeycomb, and oligocystic.[14] In 70% of cases, they are characterised by a polycystic pattern of multiple cysts measuring 2 cm or less in diameter.

Typically, ultrasonography shows a cluster-of-grapes pattern. External lobulation may also be seen. However, when the cysts are small, the mass can be echogenic (owing to the large number of acoustic interfaces), and may appear solid. This finding can suggest the presence of an adenocarcinoma. The presence of increased through transmission of ultrasound beams, even if the mass is fairly echogenic as in our patient, should suggest the diagnosis of serous cystadenomas.[15]

Computed tomographic examinations reveal an encapsulated mass. The cystic spaces are separated by fibrous septa that can coalesce into a central scar that may calcify.[16] Occasionally, delayed imaging may be a helpful means of revealing the central calcified scar.[17]

In conclusion, the association of pancreatic neoplasm with situs inversus is unusual. Reverse visceral and vascular anatomy in situs inversus may challenge surgery and can predispose to multiple post-surgery complications. Imaging plays a crucial role in revealing the detailed anatomy and characteristics of the tumour. In this case a benign serous cystadenoma was followed up with static imaging features, thus helping to avoid a difficult operation and the potential complications of such surgery.

REFERENCES

1. Blegen HM. Surgery in situs inversus. Ann Surg. 1948;129:244-59.
2. Holzmann D, Ott PM, Felix H. Diagnostic approach to primary ciliary dyskinesia: a review. Eur J Pediatr. 2000;159:95-8.
3. Lee SE, Kim HY, Jung SE, Lee SC, Park KW, Kim WK. Situs anomalies and gastrointestinal abnormalities. J Pediatr Surg. 2006;41:1237-42.
4. Bilimoria MM, Parsons WG, Small W Jr, Talamonti MS. Pancreaticoduodenectomy in a patient with ampullary carcinoma and situs inversus. Surgery. 2001;130:521-4.
5. Srivastava DN, Gujral RB. Pancreatic carcinoma associated with situs inversus in a young adult. Am J Gastroenterol. 1993;88:156-7.
6. Ibrarullah M, Awasthi S, Choudhuri G, Kapoor VK. Periampullary carcinoma in a young female with situs inversus. Indian J Gastroenterol. 1992;11:91.
7. Carlson SK, Johnson CD, Brandt KR, Batts KP, Salomao DR. Pancreatic cystic neoplasms: the role and sensitivity of needle aspiration and biopsy. Abdom Imaging. 1998;23:387-93.
8. Wolfman NT, Ramquist NA, Karstaedt N, Hopkins MB. Cystic neoplasms of the pancreas: CT and sonography. AJR Am J Roentgenol. 1982;138:37-41.
9. Johnson CD, Stephens DH, Charboneau JW, Carpenter HA, Welch TJ. Cystic pancreatic tumors: CT and sonographic assessment. AJR Am J Roentgenol. 1988;151:1133-8.
10. Colonna J, Plaza JA, Frankel WL, Yearsley M, Bloomston M, Marsh WL. Serous cystadenoma of the pancreas: clinical and pathological features in 33 patients. Pancreatology. 2008;8:135-41.
11. Kim HJ, Lee DH, Ko YT, Lim JW, Kim HC, Kim KW. CT of serous cystadenoma of the pancreas and mimicking masses. AJR Am J Roentgenol. 2008;190:406-12.
12. Compagno J, Oertel JE. Mucinous cystic neoplasms of the patient with overt and latent malignancy (cystadenocarcinoma and cystadenoma). A clinicopathologic study of 41 cases. Am J Clin Pathol. 1978;69:573-80.
13. Hyde GL, Davis JB Jr, McMillin RD, McMillin M. Mucinous cystic neoplasm of the pancreas with latent malignancy. Am Surg. 1984;50:225-9.
14. Sarr MG, Murr M, Smyrk TC, et al. Primary cystic neoplasms of the pancreas. Neoplastic disorders of emerging importance—current state-of-the-art and unanswered questions. J Gastrointest Surg. 2003;7:417-28.
15. Starkov IuG, Solodinina EN, Shishin KV, Plotnikova LS. Endoscopic ultrasonography in diagnosis of surgical treatment of pancreas [in Russian]. Khirurgiia (Mosk). 2008;(1):47-52.
16. Federle MP, McGrath KM. Cystic neoplasms of the pancreas. Gastroenterol Clin North Am. 2007;36:365-76,ix.
17. Pamuklar E, Semelka RC. MR imaging of the pancreas. Magn Reson Imaging Clin N Am. 2005;13:313-30.