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Case Report

A Rapidly Growing Epidermoid Cyst in an Intrapancreatic Accessory Spleen Treated by Laparoscopic Spleen-Preserving Distal Pancreatectomy: Case Report

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Epidermoid cysts arising in an intrapancreatic accessory spleen are exceedingly rare; furthermore, the natural course of them is hardly known. We report a case correctly diagnosed with epidermoid cyst in an intrapancreatic accessory spleen, followed by 1 year of observation, that underwent surgical treatment. The patient presented with diarrhea. Contrast-enhanced computed tomography (CT) revealed a pancreatic cyst 20 mm in diameter, surrounded by a solid component showing the same enhancement as the spleen, suggesting the presence of an epidermoid cyst in an intrapancreatic accessory spleen. One year later, back discomfort developed and a CT scan revealed that the cyst had grown to 38 mm in diameter. To obtain a definitive diagnosis, we performed a laparoscopic spleen-preserving distal pancreatectomy. The histopathological diagnosis was compatible with an epidermoid cyst in an intrapancreatic accessory spleen, which is benign. The postoperative course was uneventful. This case demonstrates that an epidermoid cyst arising in an intrapancreatic accessory spleen can rapidly grow, even if it is benign. Laparoscopic spleen-preserving distal pancreatectomy can be a useful procedure, with the advantages of low invasiveness and organ preservation, for the treatment of benign or low-grade malignant tumors located in the pancreatic body or tail.

Key words: Epidermoid cyst – Intrapancreatic accessory spleen – Laparoscopic pancreatic resection – Spleen-preserving distal pancreatectomy

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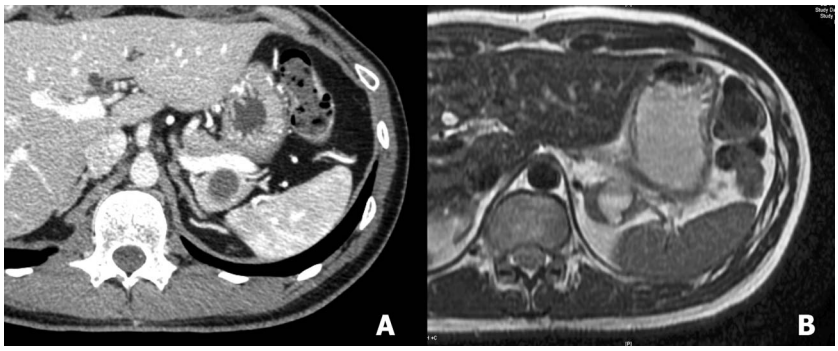

Fig. 1 A pancreatic cyst 20 mm in diameter, surrounded by a solid component showing the same attenuation as the spleen, was detected on contrast-enhanced abdominal CT (A) and SPIO-MRI (B).

Epidermoid cysts arising in an intrapancreatic accessory spleen are exceedingly rare. To our knowledge, only 39 cases have been previously reported in the English-language literature.[1–31] Most previous cases were misdiagnosed preoperatively and were treated by distal pancreatectomy with splenectomy for a suspected diagnosis of malignancy.

Itano *et al* reported that a correct diagnosis should be feasible on careful assessment of imaging findings.[14] If epidermoid cysts are diagnosed correctly, they can be observed[14,25,27] because such cysts are known to have no malignant potential. However, the natural course of epidermoid cyst arising in an intrapancreatic accessory spleen has not been reported so far.

We describe a patient in whom an epidermoid cyst arising in an intrapancreatic accessory spleen was correctly diagnosed, followed by the development of back discomfort after 1 year due to rapid growth of the cyst. The patient was successfully treated by laparoscopic spleen-preserving distal pancreatectomy.

Case Report

A 39-year-old man consulted a local physician because of diarrhea, and blood tests were performed. Serum chemistry testing showed no abnormalities, but the carbohydrate antigen 19-9 (CA19-9) level was slightly elevated to 287 U/mL. The patient was referred to our hospital for further evaluation. Contrast-enhanced computed tomography (CT) revealed a pancreatic cyst 19 × 20 mm in diameter, surrounded by a crescent-like solid component showing the same enhancement as the spleen at every phases such as arterial, portal, and equilibrium phase. The presence of an epidermoid cyst in an intrapancreatic accessory spleen was suspected (Fig. 1A). Superparamagnetic iron oxide (SPIO)–magnetic resonance imaging (MRI) showed typical findings of an intrapancreatic accessory spleen. For example, the solid component surrounding the cyst showed characteristic uptake of SPIO and revealed the same attenuation as the spleen (Fig. 1B). As endoscopic ultrasonography did not detect any findings suspicious of malignancy such as nodular lesions or cystic wall thickness, a fine-needle aspiration (FNA) was not performed. Based on the above mentioned results, our diagnosis was an epidermoid cyst in an intrapancreatic accessory spleen. The patient was followed up on an outpatient basis.

Follow-up visits were performed every 6 months. Six months later, CA19-9 level was 266 U/mL, and the pancreatic cyst was larger (22 × 26 mm), but no sign of malignancy was detected by MRI. As he had no symptom at all, he wanted to be observed. The next CT and SPIO-MRI, performed 1 year after the first examinations, showed that the epidermoid cyst had grown to 31 × 38 mm in diameter (Fig. 2A, 2B). The levels of CA19-9 level were still slightly elevated (286 U/mL). Because the patient occasionally had back discomfort, we decided to perform a laparoscopic spleen-preserving distal pancreatectomy.

Operative Technique

The patient was placed in the semilateral position with the left side up and in a reverse Trendelenburg position. The first 12-mm trocar for laparoscopy was placed in the umbilicus. After performing pneumoperitoneum at 8 mmHg, three 5-mm trocars were inserted along the left subcostal line, and one 15-mm trocar was inserted in the left lower position (Fig. 3). First, we dissected the gastrocolic membrane and opened the bursa. Then the inferior border of the pancreas was dissected, and the pancreas was completely detached from the retroperitoneum at the center of the body of the pancreas. The splenic

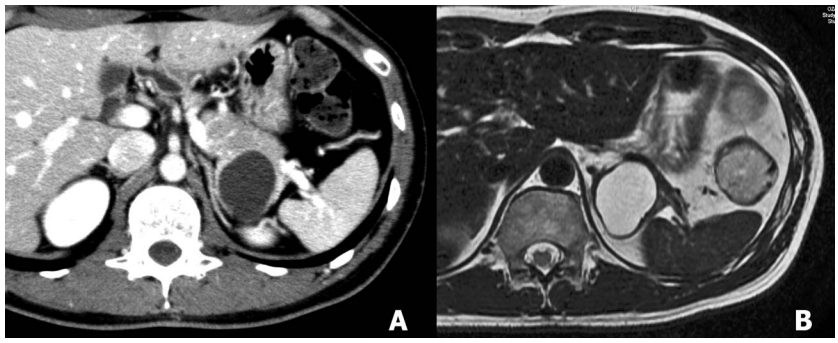

Fig. 2 The pancreatic cyst had grown to 38 mm in diameter on a CT scan (A) and SPIO-MRI (B) obtained after 1 year.

vein was identified on the posterior wall of the pancreas and was bluntly and carefully dissected away from the posterior pancreatic wall and encircled by vessel tape. The splenic artery was identified at the superior border of the pancreas by careful blunt dissection. The pancreas was isolated from the splenic vessels and was transected with the use of a linear stapler (Endo GIA with Tri-staple, Covidien, New Haven, Connecticut), which was introduced through the trocar placed in the left-lower position. Transection was performed by the slow compression technique. The dissection between the splenic vessels and pancreatic tail was performed using an ultrasonically activated scalpel (Harmonic Scalpel, Ethicon Endo-Surgery, Cincinnati, Ohio), which could control bleeding from small vessels. Although moderate inflammatory adherence existed around the cystic wall, spleen-preserving pancreatic tail resection was successfully performed laparoscopically (Fig. 4). The specimen was removed through the umbilical incision, enlarged to 3 cm. The operation time was 6 hours and 15 minutes, with minimal blood loss.

Fig. 3 A 12-mm trocar for laparoscopy was placed in the umbilicus. Three 5-mm trocars were placed along the left subcostal line, and one 15-mm trocar was inserted in the left-lower position. The pancreatic tail was removed through the umbilical incision, extended to 3 cm.

Pathological Findings

Macroscopically, the lesion in the tail of the pancreas involved 2 different components: one was a cystic mass measuring $4.0 \times 3.2 \times 2.0$ cm and the other was a brown solid mass surrounding the cyst. The cystic mass was a well-demarcated, unilocular mass, containing colorless serous fluid. The cystic wall was grayish-white and the inner surface was smooth and no nodular lesion was seen. The brown component looked like normal spleen (Fig. 5). Microscopically, the cyst was lined by nonkeratinizing stratified squamous epithelium and a few layers of atrophic squamous epithelium. The outside cystic wall was composed of relatively thick fibrous connective tissues. The brown solid component was composed of both red and white pulp, locating in the pancreatic parenchyma (Fig. 6).

Immunohistochemically, the lining squamous epithelium was positive for CA19-9 (Fig. 7). The lesion was compatible with an epidermoid cyst originating from an accessory spleen in the pancreas. There was no sign of malignancy.

Postoperative Course

The postoperative course was uneventful. There was no pancreatic fistula, and the drain was removed on postoperative day 5. The patient was discharged on

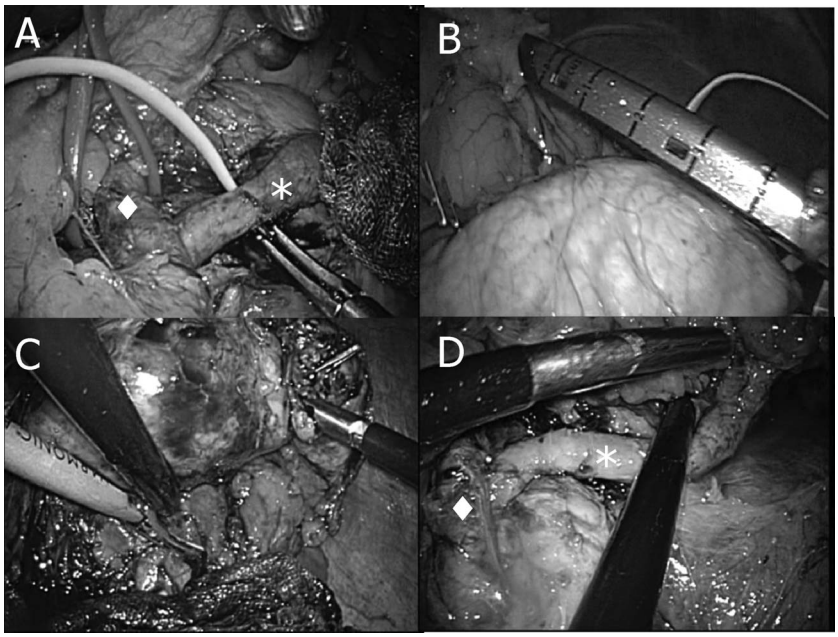

Fig. 4 (A) The splenic vein was separated from pancreas body and encircled by a vessel tape (*). The splenic artery was already encircled (◆). (B) The pancreas body was transected by stapler. (C) The distal pancreas was removed from the retroperitoneum. (D) The spleen and vessels-preserving distal pancreatectomy was done. The splenic artery (◆) and splenic vein (*) were well preserved.

postoperative day 12. The CA19-9 level entered the normal range after operation. A follow-up CT scan, obtained 2 months after operation, revealed that the splenic artery and vein were patent, and the preserved spleen showed no sign of infarction.

Discussion

Reports concerning epidermoid cysts in an intrapancreatic accessory spleen are extremely rare. Since Davidson[1] reported the first case of epidermoid cyst of the intrapancreatic accessory spleen in 1980, 39 cases of English literature have been reported. Most cases of them were preoperatively suspected to be malignancies such as mucinous cystic neoplasm and treated by distal pancreatectomy with splenectomy.

Currently available improved imaging techniques, such as multidetector row-computed tomography (MDCT) and SPIO-MRI have facilitated the diagnosis of epidermoid cyst in an intrapancreatic accessory as compared with previously. Hu *et al*[27] noted that an accessory spleen surrounding the cyst was a key component for correct diagnosis, there-

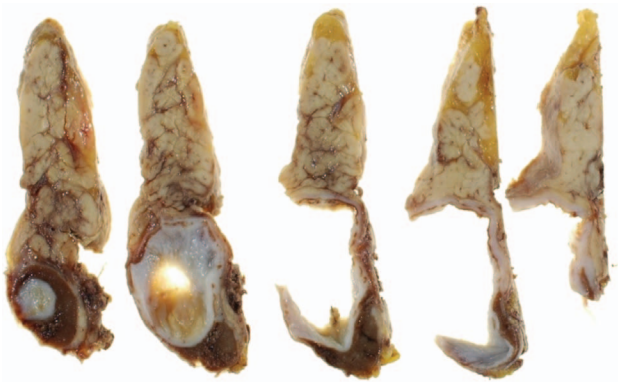

Fig. 5 Gross appearance of the cystic mass located at the pancreas tail in a cut section. The unilocular cyst measuring 4.0 × 3.2 × 2.0 cm was surrounded by a brown solid component.

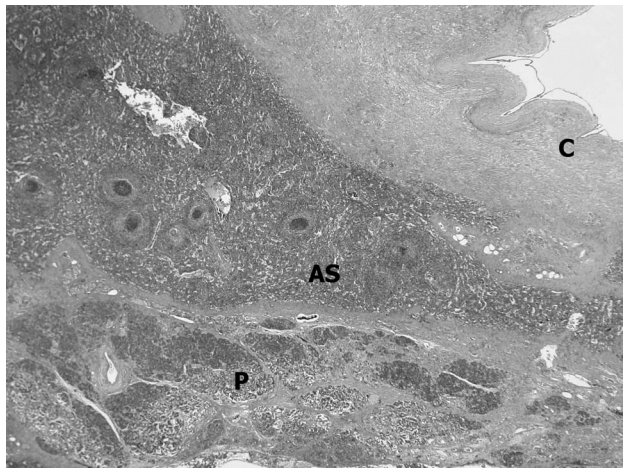

Fig. 6 Histological features of the pancreatic cyst. The cyst (C) is surrounded by fibrous tissue and a thin layer of splenic tissue (AS) containing a germinal center, adjacent to the normal pancreatic tissue (P); hematoxylin and eosin stain.

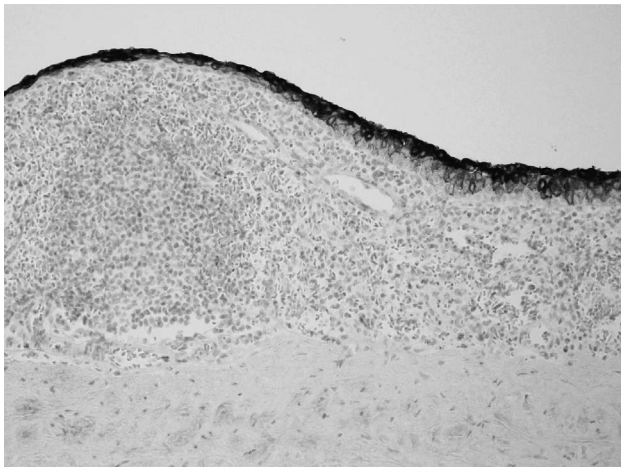

Fig. 7 Immunohistochemistry of the cyst. The cells lining the accessory spleen cyst were positive for CA19-9.

fore, the relationship of enhancement between the splenic parenchyma and the parenchymal component of the lesion for the differential diagnosis of a cystic mass in the pancreatic tail was important.

Endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) was also introduced as a new technique to facilitate the definitive diagnosis of epidermal cysts of an intrapancreatic accessory spleen and several cases have been reported.[23,24] Tatsas *et al*[32] reported that 3 out of 5 cases of intrapancreatic accessory spleen could be diagnosed accurately, but 1 case of epidermoid cyst in an intrapancreatic accessory spleen failed to get accurate diagnosis using EUS-FNA. They speculated that the solid splenic tissue surrounding the epidermoid cyst was sometimes not obtained by aspiration if the amount of splenic tissue was small. The cystic fluid obtained by EUS-FNA was also conducted for measurement of tumor markers to distinguish between benign and malignancy. Zhang and Wang[24] reported that high levels of CEA indicated that mucinous neoplasm existed. To the contrary, Reiss *et al*[23] reported that a benign epidermoid cyst of an intrapancreatic accessory spleen showed high level of CEA in the cystic fluid.

The serum CA19-9 level was relatively high in our patient. Hu *et al*[27] reported that nearly 40% of epidermoid cysts arising in an intrapancreatic accessory spleen show high levels of CA19-9, and the CA19-9 level is not a reliable marker for distinguishing benign from malignant tumors.[29] In the cases of epidermoid cyst in an intrapancreatic accessory spleen, the squamous epithelial lining of the epidermoid cyst seemed to account for the elevated serum CA19-9 levels.[2,3]

An epidermoid cyst arising in an intrapancreatic accessory spleen was correctly diagnosed in our patient, and surgical treatment was considered unnecessary at initial interview. After 1 year, however, the size of the cyst had nearly doubled, and the patient occasionally had back discomfort. The natural course of epidermoid cysts remains unclear. Although epidermoid cysts were reported not to have malignant potential, rapid growth of the cyst and a high CA19-9 level encouraged us to perform surgical treatment.

Recently, spleen-preserving distal pancreatectomy has been used to treat benign or low-grade malignant tumors arising in the pancreatic tail, because the functions of the spleen are immunologically important even in adults. A high incidence of sepsis has been reported after splenectomy.[33]

On the other hand, significant progress has been made in laparoscopic surgery during the past decade, and it is now used to treat diseases of the pancreas. Benign or low-grade malignant tumors of the pancreas tail are good indications for laparoscopic distal pancreatic resection. However, laparoscopic distal pancreatic resection was always accompanied by splenectomy because the simple procedure was easy to perform by laparoscopic surgeons. Formerly, therefore, it was difficult to decide which procedure should be selected, open spleen-preserving distal pancreatectomy or laparoscopic distal pancreatectomy with splenectomy.

In 2002, Fernandez-Cruz *et al*[34] reported the successful performance of laparoscopic distal pancreatectomy with preservation of the splenic vessels and spleen in patients with pancreatitis. Subsequently, this procedure has been used to treat other benign or low-grade malignant tumors of the pancreas. The magnified clear view afforded by laparoscopy and reliable hemostasis enabled by improved energy devices have assured the safety of this procedure.

In conclusion, to our knowledge this is the first paper to document the rapid growth of a benign epidermoid cyst arising in an intrapancreatic accessory spleen. Given the difficulty in definitively diagnosing a cystic tumor located in the body or tail of the pancreas, laparoscopic spleen-preserving pancreatic tail resection is considered a useful procedure that is minimally invasive and allows organ preservation. We believe that laparoscopic spleen-preserving pancreatic tail resection is likely to become a standard procedure for the manage-

ment of benign or low-grade malignant tumors located in the pancreatic body or tail.

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References

1. Davidson ED, Campbell WG, Hersh T. Epidermiod splenic cyst occurring in an intrapancreatic accessory spleen. *Dig Dis Sci* 1980;**25**(12):964–967
2. Hwang HS, Lee SS, Kim SC, Seo DW, Kim J. Intrapancreatic accessory spleen: clinicopathologic analysis of 12 cases. *Pancreas* 2011;**40**(6):956–965
3. Higaki K, Jimi A, Watanabe J, Kusaba A, Kojiro M. Epidermoid cyst of the spleen with CA19-9 or carcinoembryonic antigen productions: report of three cases. *Am J Surg Pathol* 1998;**22**(6):704–708
4. Morohoshi T, Hamamoto T, Kunimura T, Yoshida E, Kanda M, Funo K *et al*. Epidermoid cyst derived from an accessory spleen in the pancreas. A case report with literature survey. *Acta Pathol Jpn* 1991;**41**(12):916–921
5. Tang X, Tanaka T, Tsutsumi Y. Epithelial inclusion cysts in an intrapancreatic accessory spleen. *Pathol Int* 1994;**44**(8):652–654
6. Tateyama H, Tada T, Murase T, Fujitake S, Eimoto T. Lymphoepithelial cyst and epidermoid cyst of the accessory spleen in the pancreas. *Mod Pathol* 1998;**11**(12):1171–1177
7. Sasou S, Nakamura S, Inomata M. Epithelial splenic cysts in an intrapancreatic accessory spleen and spleen. *Pathol Int* 1999;**49**(12):1078–1083
8. Horibe Y, Murakami M, Yamao K, Imaeda Y, Tashiro K, Kasahara M. Epithelial inclusion cyst (epidermoid cyst) formation with epithelioid cell granuloma in an intrapancreatic accessory spleen. *Pathol Int* 2001;**51**(1):50–54
9. Kadota K, Kushida Y, Miyai Y, Katsuki N, Hayashi T, Bando K *et al*. Epidermoid cyst in an intrapancreatic accessory spleen: three case reports and review of the literatures. *Pathol Oncol Res* 2010;**16**(3):435–442
10. Furukawa H, Kosuge T, Kanai Y, Mukai K. Epidermoid cyst in an intrapancreatic accessory spleen: CT and pathologic findings. *Am J Roentgenol* 1998;**171**(1):271
11. Fink AM, Kulkarni S, Crowley P, Crameri JA. Epidermoid cyst in a pancreatic accessory spleen mimicking an infected abdominal cyst in a child. *Am J Roentgenol* 2002;**179**(1):206–208
12. Sonomura T, Kataoka S, Chikugo T, Hirooka T, Makimoto S, Nakamoto T *et al*. Epidermiod cyst originating from an intrapancreatic accessory spleen. *Abdom Imaging* 2002;**27**(5):560–562
13. Yokomizo H, Hifumi M, Yamane T, Hirata T, Terakura H, Murata K *et al*. Epidermiod cyst of an accessory spleen at the pancreatic tail: diagnostic value of MRI. *Abdom Imaging* 2002;**27**(5):557–559
14. Motosugi U, Yamaguchi H, Ichikawa T, Sano K, Araki T, Takayama Y *et al*. Epidermiod cyst in intrapancreatic accessory spleen: radiological findings including superparamagnetic iron oxide-enhanced magnetic resonance imaging. *J Comput Assist Tomogr* 2010;**34**(2):217–222
15. Nakae Y, Hayakawa T, Kondo T, Shibata T, Kitagawa M, Sakai Y *et al*. Epidermoid cyst occurring in a pancreatic accessory spleen. *J Clin Gastroenterol* 1991;**13**(3):362–364
16. Choi SK, Ahn SI, Hong KC, Kim SJ, Kim TS, Woo ZH *et al*. A case of epidermoid cyst of the intrapancreatic accessory spleen. *J Korean Med Sci* 2000;**15**(5):589–592
17. Tsutsumi S, Kojima T, Fukai Y, Kanoh K, Shimura T, Mochiki E *et al*. Epidermoid cyst of an intrapancreaticaccessory spleen: a case report. *Hepatogastroenterology* 2000;**47**(35):1462–1464
18. Kanazawa H, Kamiya J, Nagino M, Uesaka K, Yuasa N, Oda K *et al*. Epidermoid cyst in an intrapancreatic accessory spleen: a case report. *J Hepatobiliary Pancreat Surg* 2004;**11**(1):61–63
19. Ru K, Kalra A, Ucci A. Epidermoid cyst of intrapancreatic accessory spleen. *Dig Dis Sci* 2007;**52**(5):1229–1232
20. Gleeson FC, Kendrick ML, Chari ST, Zhang L, Levy MJ. Epidermiod accessory splenic cyst masquerading as a pancreatic mucinous cystic neoplasm. *Endoscopy* 2008;**40**(suppl 2):E141–E142
21. Itano O, Shiraga N, Kouta E, Iri H, Tanaka K, Hattori H *et al*. Epidermoid cyst originating from an intrapancreatic accessory spleen. *J Hepatobiliary Pancreat Surg* 2008;**15**(4):436–439
22. Servais EL, Sarkaria IS, Solomon GJ, Gumpeni P, Lieberman MD. Giant epidermoid cyst within an intrapancreatic accessory spleen mimicking a cystic neoplasm of the pancreas: case report and review of the literature. *Pancreas* 2008;**36**(1):98–100
23. Reiss G, Sickel JZ, See-Tho K, Ramrakhiani S. Intrapancreatic splenic cyst mimicking pancreatic cystic neoplasm diagnosed by EUS-FNA. *Gastrointest Endosc* 2009;**70**(3):557–558
24. Zhang Z, Wang JC. An epithelial splenic cyst in an intrapancreatic accessory spleen. A case report. *JOP* 2009;**10**(6):664–666
25. Itano O, Chiba N, Wada T, Yuasa Y, Sato T, Ishikawa H *et al*. Laparoscopic resection of an epidermoid cyst originating from an intrapancreatic accessory spleen: report of a case. *Surg Today* 2010;**40**(1):72–75
26. Khashab MA, Canto ML, Singh VK, Hruban RH, Makary MA, Giday S. Endosonographic and elastographic features of a rare epidermoid cyst of an intrapancreatic accessory spleen. Endoscopy 2011;**43**(suppl 2):E193–E194
27. Hu S, Zhu L, Song Q, Chen K. Epidermoid cyst in intrapancreatic accessory spleen: computed tomography

findings and clinical manifestation. *Abdominal Imaging* 2012; **37**(5):828–833

28. Yamanashi H, Kumagi T, Yokota T, Koizumi M, Azemoto N, Watanabe J *et al*. Epithelial cyst arising in an intrapancreatic accessory spleen: a diagnostic dilemma. *Intern Med* 2011;**50**: 1947–1952
29. Horn AJ, Lele SM. Epidermoid cyst occurring within an intrapancreatic accessory spleen. A case report and review of the literature. *J Pancreas* 2011;**12**(3):279–282
30. Urakami A, Yoshida K, Hirabayashi Y, Kubota H, Yamashita K, Hirai T *et al*. Laparoscopy-assisted spleen-preserving pancreatic resection for epidermoid cyst in an intrapancreatic accessory spleen. *Asian J Endosc Surg* 2011;**4**(4):185–188
31. Harris AC, Chaudry MA, Menzies D, Conn PC. Laparoscopic resection of epidermoid cyst within an intrapancreatic accessory spleen: a case report and review article. *Surg Laparosc Endosc Percutan Tech* 2012;**22**(4):e246–e249
32. Tatsas AM, Owens CL, Siddiqui MT, Hruban RH, Ali SZ. Fine-needle aspiration of intrapancreatic accessory spleen: cytomorphologic features and differential diagnosis. *Cancer Cytopathol* 2012;**120**:261–268
33. Di Sabatino A, Carsetti R, Corazza GR. Post-splenectomy and hyposplenic states. *Lancet* 2011;**378**:86–97
34. Fernandez-Cruz L, Saenz A, Astudillio E, Pantoja JP, Uzcategui E, Navarro S. Laparoscopic pancreatic surgery in patients with chronic pancreatitis. *Surg Endosc* 2002;**16**:996–1003