Malaysian J Pathol 2017; 39(3) : 327 – 330

CASE REPORT

Solid pseudopapillary neoplasm of the pancreas with liver metastasis initially misinterpreted as benign haemorrhagic cyst

Min Jung JUNG *MD*, Hee Kyung KIM *MD*, Seo-Youn CHOI* *MD*, Sang Gyune KIM** *MD* and So-Young JIN*** *MD*

*Department of Pathology, Soonchunhyang University Bucheon Hospital, Soonchunhyang University College of Medicine, Bucheon, Republic of Korea, *Department of Radiology, Soonchunhyang University Bucheon Hospital, Soonchunhyang University College of Medicine, Bucheon, Republic of Korea, **Digestive Disease Center and Research Institute, Department of Internal Medicine, Soonchunhyang University Bucheon Hospital, Soonchunhyang University College of Medicine, Bucheon, Republic of Korea and ***Department of Pathology, Soonchunhyang University Seoul Hospital, Soonchunhyang University College of Medicine, Seoul, Republic of Korea*

Abstract

Solid pseudopapillary neoplasm (SPN) of the pancreas is considered a low-malignant neoplasm with a good prognosis. However, 5% to 15% of patients with SPNs develop metastatic disease, most commonly in the liver. Metastatic hepatic malignancies that show pseudocystic features are rare. Here we describe the case of a middle-aged female with a cystic liver metastasis from SPN. To the best of our knowledge, SPN with a single cystic liver metastasis has not been described, although these tumours frequently undergo haemorrhagic-cystic degeneration. Thus, in these patients the marked cystic change could be misinterpreted as a benign lesion.

Keywords: liver, metastasis, pancreas, solid pseudopapillary neoplasm

INTRODUCTION

Solid pseudopapillary neoplasm (SPN) of the pancreas is a rare malignant tumour with low malignant potential. Although 5% to 15% of SPNs metastasise or recur after complete excision, most patients with metastatic SPNs have long-term disease-free survival.[1-4] Because SPN is rare and considered a low-grade malignant neoplasm with few proven predictors of aggressiveness, the possibility of metastasis is often clinically ignored.[4-7] Here, we describe a case in which a patient developed SPN with a liver metastasis that was initially misinterpreted as a benign cystic lesion. To the best of our knowledge, this is the first report of cystic metastasis of SPN.

CASE REPORT

A 49-year-old female was referred to our hospital due to abdominal discomfort. On initial abdominal computed tomography (CT) and magnetic resonance imaging (MRI), a 4.8 cm mainly cystic mass containing calcifications and haemorrhage was demonstrated in the tail of the pancreas. A 3.6 cm cystic mass with haemorrhage was simultaneously observed in the liver (Figs. 1A–F). On a subsequent positron emission tomography (PET)-CT scan, the hepatic mass showed no demonstrable hypermetabolism, whereas the mass in the pancreatic tail had hypermetabolic features (standardized uptake value [SUV] of 4.1). Accordingly, the pancreatic mass was diagnosed as SPN, based on the preoperative images, while the hepatic mass was considered a separate disease entity, possibly a haemorrhagic cyst or a primary cystic tumour of the liver. The patient underwent a distal pancreatectomy with splenectomy.

Pathology

A 6.0 cm × 5.0 cm, cystic and solid mass surrounded by a pseudocapsule was noted in the tail of the pancreas (Fig. 2A). The cystic areas contained haemorrhagic and necrotic material and the solid portion, at the periphery of the tumour, had a white-gray, friable surface.

Address for correspondence: Hee Kyung Kim, MD, PhD, Department of Pathology, Soonchunhyang University Bucheon Hospital, Soonchunhyang University College of Medicine, 170 Jomaru-ro, Wonmi-gu, Bucheon 14584, Republic of Korea. Tel: +82-32-621-5975; Fax: +82-32-621-5961. E-mail: heekyung@schmc.ac.kr

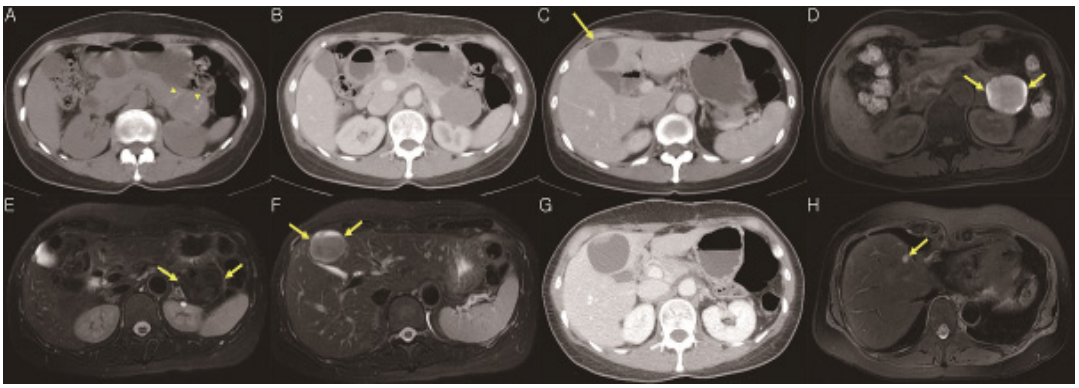

FIG. 1: (A-C) Initial abdominal computed tomography (CT) scans show a 4.8 cm mass in the pancreatic tail with scattered calcifications (arrowheads) on the unenhanced image (A) and poor enhancement after contrast enhancement (B). A 3.6 cm low-density lesion with enhancing foci is seen in the peripheral portion of the liver (arrow in C). The pancreatic mass (arrows) shows high signal intensity (SI) on unenhanced T1-weighted magnetic resonance imaging (MRI) (D) and a low SI on fat-saturated T2-weighted MRI (E), indicative of haemorrhage. A fluid-fluid level with different SIs in the hepatic lesion (arrows in F) is consistent with haemorrhage. On follow-up CT scans obtained 1 year after distal pancreatectomy, the enlarged hepatic mass was treated surgically and diagnosed as metastasis (G). On follow-up MRI 3 months after resection of the hepatic metastasis, an apparently newly developed 0.5 cm cystic lesion (arrow) was detected near the hepatic resection margin, strongly suggesting a new hepatic metastasis (H)

Microscopically, the solid area showed a proliferation of uniform tumour cells with solid and pseudopapillary growth patterns and degenerative change (Fig. 2B). The tumour cells were monomorphous with oval nuclei and moderate amount of eosinophilic or clear cytoplasm. Mitoses were rare and evidence of perineural or lymphovascular invasion or tumour infiltration to the peripancreatic soft tissue was absent. Beta-catenin was expressed within the nuclei. The tumour cells expressed CD10 and vimentin but were negative for chromogranin-A. The expression level of Ki-67 was low, but in focal areas of the tumour the Ki-67 index was elevated to about 8% (Fig. 2C). Neither the four examined lymph nodes nor the spleen contained metastatic tumour cells. Complete resection of the tumour was achieved.

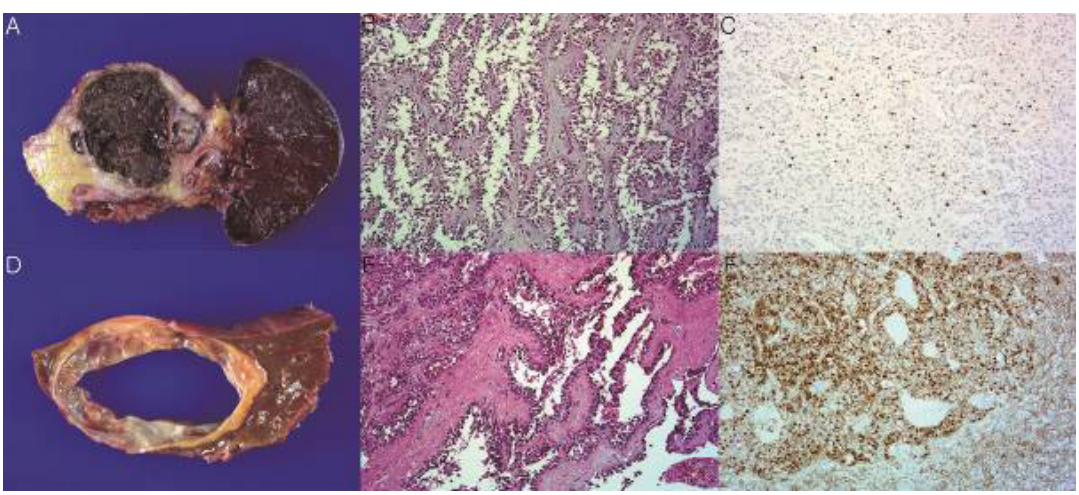

FIG. 2: (A) A well-defined cystic and solid mass with extensive haemorrhagic degeneration is seen in the pancreas. (B) Histology shows a solid pseudopapillary neoplasm (SPN) with characteristic pseudopapillary architecture (H&E, 100x), and (C) increased Ki-67 expression in the tumour hotspot (Ki-67, 100x). (D) The well-defined cystic mass in the liver shows identical histological features (H&E, 100x) and (E) immunohistochemical staining for beta-catenin (β-catenin, 100x)

Follow-up

The pancreatic mass was pathologically confirmed as SPN, and the hepatic mass was closely monitored. On follow-up CT examination 1 year after resection of the pancreatic mass, the size of the hepatic mass had increased, to 4.8 cm (Fig. 1G). An extended left hemihepatectomy was performed based on our suspicion that the mass was malignant. Left hepatectomy resulted in the detection of a cystic mass measuring 8.0 cm × 5.0 cm and filled with a clear yellowish fluid. The cyst wall was irregularly thickened (Fig. 2D). The histological features (Fig. 2E) and immunohistochemical staining results (Fig. 2F) of the liver tumour were identical to those of the resection specimen from the pancreas, supporting the diagnosis of metastatic SPN.

After liver resection, a small, presumably newly developed cystic lesion was identified in the liver during a follow-up examination (Fig. 1H). The lesion consistently increased in size and was hypermetabolic (SUV_{max} = 4.5) on a PET-CT scan. Based on these imaging findings, the newly developed hepatic lesion was considered a metastasis. The patient who refused surgery is alive although the liver metastasis had been left untreated for 35 months. At the 36 months after operation, the lesion was treated by radiofrequency ablation.

DISCUSSION

For SPNs with an aggressive clinical course, despite efforts to identify histopathological features useful for predicting malignant potential, there are still no unequivocal predictors of outcome. Among the clinicopathological features proposed as candidate predictors to date are patient age, tumour size, tumour metastasis at the first operation and pathological features such as diffuse growth pattern, nuclear atypia, a high mitotic rate, and extensive necrosis.[4-7] Recently, Yang *et al*[8] reported that patients with SPNs characterized by a Ki-67 index ≥4% had poorer recurrence-free. Our patient was older than the typical SPN patient, and the initial examination revealed a synchronous hepatic metastasis. Histopathologically, except for the slightly increased Ki-67 labeling index (up to 8% in hotspots), she had no suspicious findings. Nevertheless, after distal pancreatectomy, the liver cystic mass increased in size during 13 months of follow-up. Following hepatectomy performed to treat the metastasis, another metastatic lesion was detected and its size increased slightly thereafter.

Severe degenerative changes with extensive necrosis, haemorrhage, and hyalinization are common histological features of SPN. They may also be features of metastases, which can present as cystic lesions. A female patient with characteristic CT and MRI findings suggests a preoperative diagnosis of pancreatic SPN. However, a marked cystic change in another organ might resemble a pseudocyst and thus lead to a misdiagnosis. In our patient, predictive findings such as older age and the increased Ki-67 labeling index of the tumour were helpful markers that led us to suspect metastasis. However, in most cases an accurate preoperative diagnosis is difficult. Recent advances in the molecular study of cystic fluid have improved the diagnosis of metastatic SPN. In Springer *et al*,[9] SPNs were identified with 100% sensitivity and 100% specificity by the presence of a *CTNNB1* mutation and the absence of *KRAS*, *GNAS*, or *RNF43* mutations or a loss of heterozygosity in chromosome 18. An analysis of the cystic fluid for molecular markers could improve the differentiation of metastatic SPNs from other neoplasms and thus allow correct treatment.

To the best of our knowledge, this is the first report of a patient with a SPN in the pancreatic tail and a single cystic liver metastasis. While most patients described in the previous reports had multiple hepatic metastases, the metastatic hepatic mass in our case presented as a single cystic lesion, which was initially misinterpreted as a benign haemorrhagic cyst. A mass containing only a cystic area in the extrapancreatic tissue is not characteristic for metastatic SPN. However, in patients in whom these lesions are detected, metastatic SPN should be considered, particularly if the mass is present in the liver and the patient is an older female.

ACKNOWLEDGEMENT

This work was supported by the Soonchunhyang University Research Fund. The authors declare no conflict of interest in the conduct of this study.

REFERENCES

1. Papavramidis T, Papavramidis S. Solid pseudopapillary tumors of the pancreas: review of 718 patients reported in English literature. J Am Coll Surg. 2005; 200: 965-72.
2. Klimstra DS, Wenig BM, Heffess CS. Solid-pseudopapillary tumor of the pancreas: a typically cystic carcinoma of low malignant potential. Semin Diagn Pathol. 2000; 17: 66-80.
3. Kim CW, Han DJ, Kim J, Kim YH, Park JB, Kim

SC. Solid pseudopapillary tumor of the pancreas: can malignancy be predicted? Surgery. 2011; 149: 625-34.
4. Bosman FT, Carneiro F, Hruban RH, Theise ND, editors. WHO classification of tumours of the digestive system. 4th ed. Lyon, France: International Agency for Research on Cancer; 2010.
5. Tang LH, Aydin H, Brennan MF, Klimstra DS. Clinically aggressive solid pseudopapillary tumors of the pancreas: a report of two cases with components of undifferentiated carcinoma and a comparative clinicopathologic analysis of 34 conventional cases. Am J Surg Pathol. 2005; 29: 512-9.
6. Kang CM, Kim KS, Choi JS, Kim H, Lee WJ, Kim BR. Solid pseudopapillary tumor of the pancreas suggesting malignant potential. Pancreas. 2006; 32: 276-80.
7. Machado MC, Machado MA, Bacchella T, Jukemura J, Almeida JL, Cunha JE. Solid pseudopapillary neoplasm of the pancreas: distinct patterns of onset, diagnosis, and prognosis for male versus female patients. Surgery. 2008; 143: 29-34.
8. Yang F, Yu X, Bao Y, Du Z, Jin C, Fu D. Prognostic value of Ki-67 in solid pseudopapillary tumor of the pancreas: Huashan experience and systematic review of the literature. Surgery. 2016; 159: 1023-31.
9. Springer S, Wang Y, Dal Molin M*, et al.* A combination of molecular markers and clinical features improve the classification of pancreatic cysts. Gastroenterology. 2015; 149: 1501-10.