

## Well-Differentiated Pancreatic Neuroendocrine Tumor with Solitary Hepatic Metastasis Presenting as a Benign Cystic Mass: A Case Report

양성낭성종괴로 오인된 고분화췌장신경내분비종양의 단발성 간전이 1예:  
증례 보고

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Pancreatic neuroendocrine tumors and their hepatic metastases have an inconsistent appearance with only a small percentage of lesions appearing as cystic masses in computed tomography (CT) and magnetic resonance imaging (MRI). Therefore, they can be mistaken as benign or infectious lesions, which can lead to a false diagnosis with delayed or inadequate treatment. We reported a patient with upper abdominal pain that lasted for several months, caused by a huge cystic neuroendocrine carcinoma of the liver. This was mistakenly interpreted as a complicated or hydatid cyst, and the findings in the CT and MRI was presented.

### Index terms

Pancreatic Neuroendocrine Tumor  
Hepatic Metastasis  
Cystic Mass

## INTRODUCTION

Pancreatic neuroendocrine tumors and their hepatic metastases have an inconsistent appearance with only a small percentage of lesions appearing as cystic masses in computed tomography (CT) and magnetic resonance imaging (MRI). They can therefore be mistaken for benign or infectious lesions, leading to a false diagnosis with delayed or inadequate treatments. We reported a patient with an upper abdominal pain that lasted for several months, caused by a huge cystic neuroendocrine carcinoma of the liver that was mistakenly interpreted as a complicated or hydatid cyst. We presented the findings of the initial CT and the subsequently performed MRI under the application of hepatocyte specific contrast material, which led to distinct differential diagnoses that had a major impact on the therapeutic strategy.

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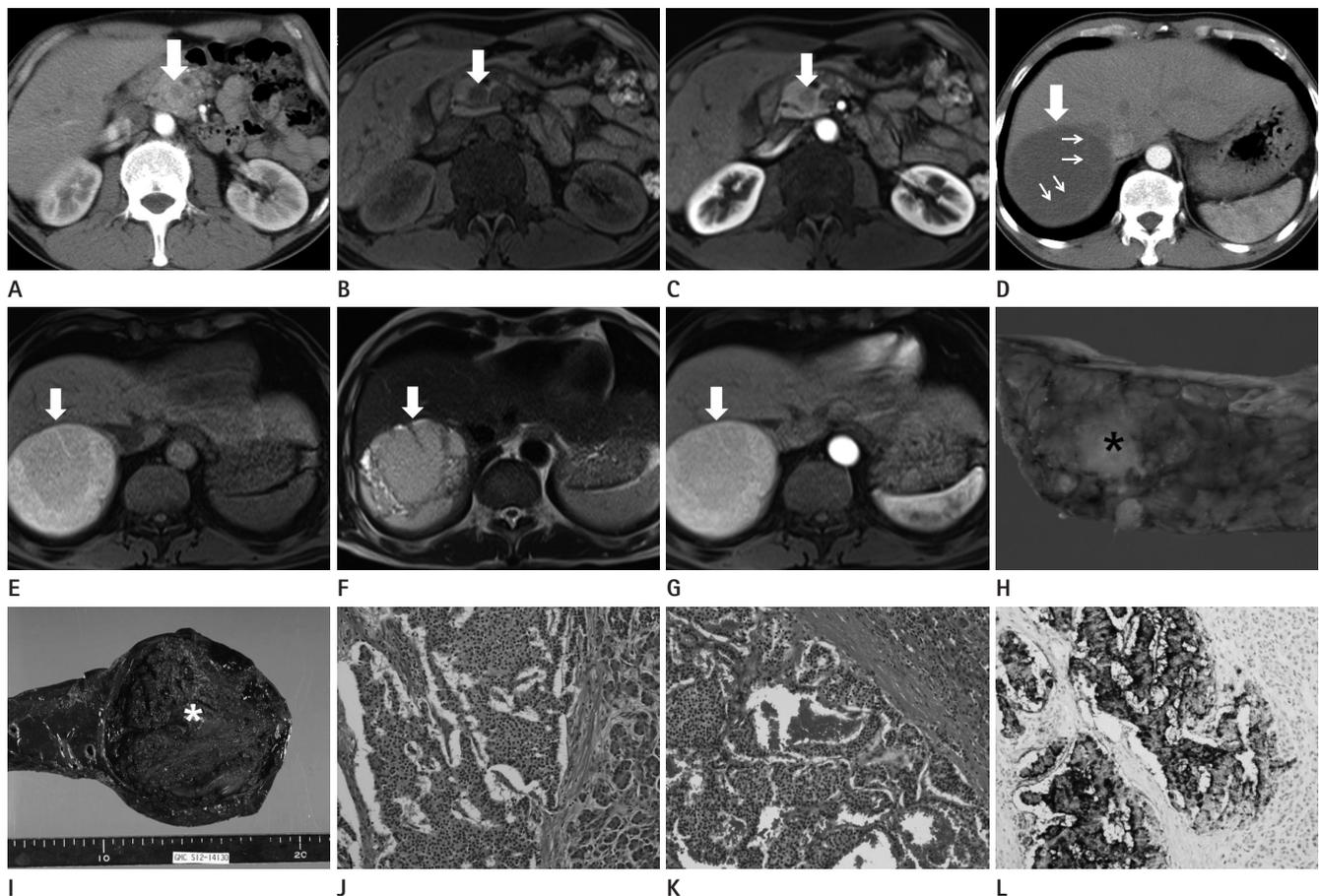
## CASE REPORT

A 50-year-old male patient complained of an epigastric pain that he was having for a week. Hematology laboratory data were within the normal range. Contrast enhanced CT from an external institution showed a 15 mm hypodense mass in the head of the pancreas when compared with the uninvolved pancreas (Fig. 1A). The pancreatic mass was interpreted as ductal adenocarcinoma or neuroendocrine tumor. Additionally, a CT scan showed a 10 cm hypodense mass in the right hepatic lobe with internal thin septa (Fig. 1D), and this was interpreted as a simple cyst of the right hepatic lobe. The patient was referred to our center for surgical therapy. At our center, for further preoperative evaluation, MRI of the liver was performed with unenhanced T1- and T2-weighted images with fat suppression, contrast enhanced sequences using Gd-EOB-DTPA (Primovist, Bayer Schering Phar-

ma, Berlin, Germany) with early dynamic acquisition, and delayed imaging in the hepatobiliary phase after 20 minutes (1.5 T; AVANTO; Siemens Medical Systems, Erlangen, Germany). On T1-weighted MR image, pancreatic mass showed hypointensity compared with the uninvolved pancreas (Fig. 1B). T1-weighted image after application of Gd-EOB-DTPA shows subtle enhancement of the mass in the arterial phase (Fig. 1C).

On MRI, the lesion in the right liver lobe, measuring 10.0 × 6.0 cm, showed a thin wall and septa. On T1-weighted trans-

verse gradient echo sequences, the wall and septa appeared hypointense, while the liquid content showed subtle hyperintensity (Fig. 1E). On T2-weighted transverse turbo spin echo sequences, the wall and septa were hyperintense, while the liquid content showed subtle hyperintensity (Fig. 1F). On T1-weighted image with contrast enhancement, no Gd-EOB-DTPA uptake is present in wall and septations of the mass, while the cyst content remained hypointense (Fig. 1G). Based on these findings, a complicated or hydatid cystic tumor was preferred as the differ-



**Fig. 1.** A 50-year-old man presented with a week's history of epigastric pain.  
**A.** Transverse CT scan shows a 15 mm hypodense mass in head of pancreas compared with the normal pancreas parenchyma (arrow).  
**B.** T1-weighted MR image shows a 15 mm hypointense mass in head of pancreas compared with the normal pancreas parenchyma (arrow).  
**C.** T1-weighted image after application of Gd-EOB-DTPA shows subtle enhancement of the mass in the arterial phase (arrow).  
**D.** Transverse CT scan shows a 10 cm hypodense mass (thick arrow) in the right hepatic lobe with suspicious internal thin septa (thin arrows).  
**E.** T1-weighted MR image shows a 10 cm hyperintense mass in the right hepatic lobe with suspicious internal thin septa (arrow).  
**F.** T2-weighted MR image shows hyperintense mass including internal multiple septations (arrow).  
**G.** On T1-weighted image after contrast media injection, no Gd-EOB-DTPA uptake of the mass is present (arrow).  
**H.** A photograph of the resected pancreatic specimen shows the 23 mm irregular margined yellowish mass (asterisk) in head of pancreas.  
**I.** A photograph of the resected hepatic specimen shows the large liver mass (asterisk) possessing a capsule, containing hemorrhages.  
**J.** Histology of the neuroendocrine carcinoma in the pancreas. Tumor cells featuring abundant cytoplasm, prominent nucleoli; frequent mitotic rate shows the organoid, nesting, trabecular and palisading pattern (H&E, × 200).  
**K, L.** Histology of the neuroendocrine carcinoma in the liver. Tumor cells featuring abundant cytoplasm, prominent nucleoli; frequent mitotic rate shows the organoid, nesting, trabecular and palisading pattern (H&E, × 200) (**K**). Positive immunohistochemical stain for synaptophysin (synaptophysin, × 200) (**L**).

ential diagnosis.

Pancreatotomy and a right hemihepatectomy (Segments VI–VII) were performed. Resected pancreatic specimen shows irregular, margined, and yellowish tumor in the head of the pancreas, measuring  $2.3 \times 1.5 \times 1.2$  cm, without common bile duct invasion (Fig. 1H). Resected hepatic specimen shows the large liver mass possessing a capsule, containing hemorrhages (Fig. 1I). Histology of the pancreatic tumor reveals tumor cells featuring abundant cytoplasm and prominent nucleoli; frequent mitotic rate shows the organoid, nesting, trabecular, and palisading pattern (Fig. 1J). Hepatic tumor shows similar histologic features with the pancreatic mass, and partially cystic neoplasm infiltrates the adjacent liver parenchyma with microcystic and trabecular growth pattern (Fig. 1K). There was an expression of CK7, synaptophysin, chromogranin, and CK20 (Fig. 1L). These findings resulted in the diagnosis of a well differentiated neuroendocrine tumor of pancreatic origin with hepatic metastasis (1).

## DISCUSSION

Pancreatic neuroendocrine tumors (PNETs) are rare neoplasms that affect only 1 in 300000 individuals in the United States annually, and comprise 1% to 2% of all pancreatic tumors (1, 2). The major categories of gastroenteropancreatic neuroendocrine tumors defined by the WHO are well-differentiated neuroendocrine tumors, which show benign behavior or uncertain malignant potential, well-differentiated neuroendocrine carcinomas, which are characterized by low-grade malignancy, and poorly differentiated (usually small cell) neuroendocrine carcinomas of high-grade malignancy (3).

Most malignant PNETs metastasize to the liver where they derive their blood supply from hepatic artery branches (75–80%) (4). Hepatic metastasis occurs in 30–85% of PNETs, and is potentially and completely resectable only in 7–15% of the patients (5, 6). Because of the varying presentation of the patients with metastatic hepatic neuroendocrine tumors and the rarity of this disease, it is often difficult to define the ideal treatment approach based upon the current literatures. In patients with isolated hepatic diseases, an increasing number of published findings report the improved survival rates in the patients undergoing complete resection (6).

Hepatic metastases are most frequently hypointense or iso-

tense compared to the surrounding normal liver parenchyma on T1-weighted image, and are hyperintense on T2-weighted images. On dynamic gadolinium enhancement phases, metastases typically showed early enhancement (7). Cystic appearance of neuroendocrine carcinoma (NEC) metastases is rare. The low prevalence of NEC and the unusual, large solitary cystic lesion in the liver made this differential diagnosis quite unlikely, which initially resulted in the interpretation as an echinococcal cyst based on the monophasic contrast enhanced CT (8). Despite newer chemotherapeutic and immunologic agents, surgical therapy remains the most efficient approach to metastatic disease and offers the longest lasting benefits. Surgical debulking of the hepatic disease improves the chance of survival (9).

In this article, we described the CT and MRI findings on cystic appearance of hepatic metastases from PNETs, which was mistakenly interpreted as complicated or hydatid cyst. Because of variability of neuroendocrine tumor appearance in imaging, percutaneous biopsy should be considered for atypical imaging to avoid delayed diagnosis and wrong treatment.

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## 양성낭성종괴로 오인된 고분화췌장신경내분비종양의 단발성 간전이 1예: 증례 보고

안수좌 · 최승준 · 김형식 · 김정호 · 최혜영

췌장신경내분비종양과 그들의 간전이는 극히 일부에서 전산화단층촬영이나 자기공명영상에서 양성종괴로 보이는데 이로 인해 양성낭성종괴나 염증성종괴로 오인되어 치료가 늦어지거나 부적절한 처치를 할 수 있다. 저자들은 수개월 동안 상복부 통증을 주소로 내원한 환자가 시행한 전산화단층촬영과 자기공명영상에서 복합낭종이나 포충낭으로 오인된 양성 변화를 동반한 췌장신경내분비종양의 간전이를 경험하였기에 영상 소견을 간단한 문헌 고찰과 함께 보고하고자 한다.

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