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Case Report

Myoepithelial hamartoma as a solitary mass in the pancreatic parenchyma: the first case report

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Solid hamartoma of the pancreas is very rare, and only 3 cases have been reported thus far. A patient underwent pancreaticoduodenectomy due to a mass in the head of the pancreas which was suspected to be a borderline malignant tumor, but the histologic diagnosis turned out to be myoepithelial hamartoma (MEH) or adenomyoma. It was characterized by benign duct and glandular structures surrounded by proliferating smooth muscle, and acinus formation was not observed. Immunohistochemical stain for smooth muscle actin (SMA) was positive in spindle cells, and CD34 was negative, differentiating it from the three previously reported cases of solid hamartoma of the pancreas. MEH is an entity that is on the same spectrum as heterotopic pancreas. MEH is rare and has usually been reported in the gastrointestinal tract. To the best of our knowledge, MEH has never been reported in the pancreas. Therefore we report the world's first documented case of MEH of the pancreas. **(Korean J Hepatobiliary Pancreat Surg 2012;16:80-83)**

Key Words: Hamartoma; Pancreas; Myoepithelial hamartoma

INTRODUCTION

Common features of myoepithelial hamartoma (MEH) are abnormal glandular formations lined by columnar epithelium and surrounded by smooth muscle cells.[1] MEH has been referred to by various names such as incompletely differentiated pancreas, adenomyomas, adenomyosis, and foregut choristomas.[2] The existence of various names indicates that the pathogenesis and clinicopathologic features of this disease entity are not well understood. The rarity of MEH is in part responsible for this.

We experienced a case in which a patient underwent pancreaticoduodenectomy due to a mass in the head of the pancreas that was thought to be a borderline malignant tumor, but which turned out to be a benign tumor histologically. The histologic diagnosis was MEH. MEH itself is an extremely rare disease entity that has been reported in sites in the gastrointestinal tract such as the stomach and small intestine. To the best of our knowledge, MEH has never been reported in the parenchyma of the pancreas. Herein, we report the world's first documented case of MEH of the pancreas with a review of the literature.

CASE

A 41 year old female was referred to the Seoul National University Hospital for further evaluation of a mass in the head of the pancreas detected by ultrasonography during a regular check-up at a health-care clinic. There were no abnormalities in her last check-up 2 years ago. She had neither a significant past medical history nor family history. Review of her symptoms and physical examination did not reveal any remarkable symptoms or signs. She was admitted to the department of surgery for further evaluation and management.

A 2.3×1.5 cm sized well demarcated mass was identified at the pancreas head and uncinate process by computed tomography. The mass showed low attenuation during the early and late arterial phase, and high enhancement at the portal phase. The possible differential diag-

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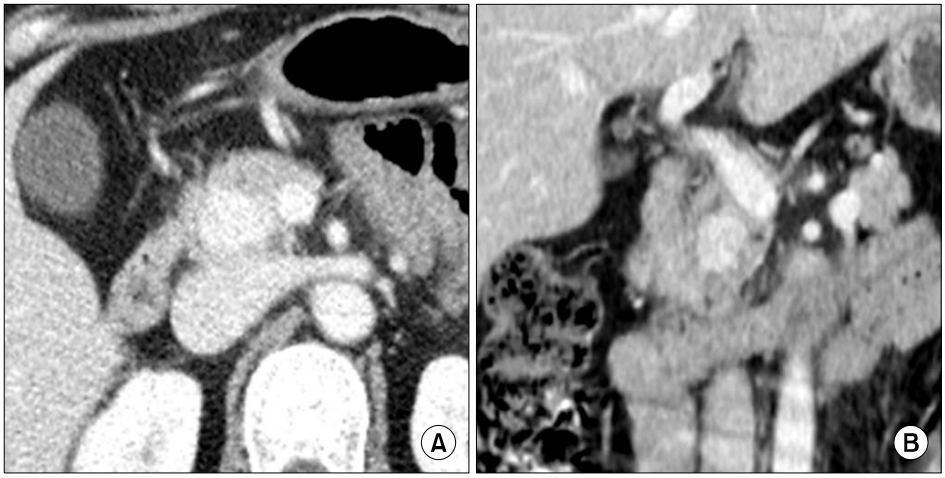

Fig. 1. CT findings of the pancreatic mass. During the computed tomography, a 2-cm sized mass with enhancement during the portal phase is demonstrated in cross sectional view (A) and coronal section (B).

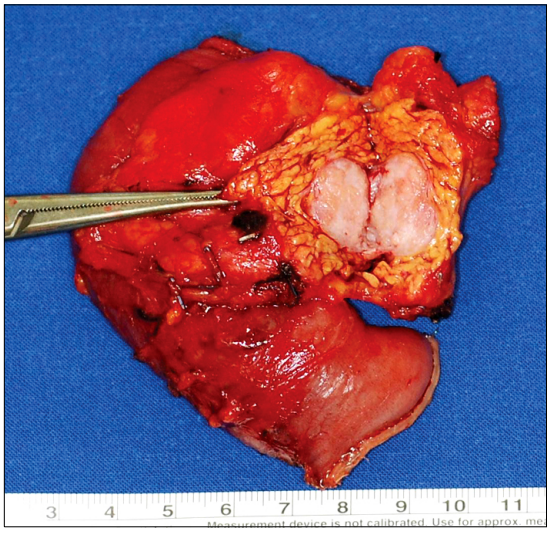

Fig. 2. Gross photograph of the specimen. On gross examination, a 2×1 cm-sized well-demarcated whitish mass with lobulation without evidence of degeneration or hemorrhage was visible in the parenchyma of the pancreas head.

noses were neuroendocrine tumor (NET) or solid pseudopapillary neoplasm (SPN), although the CT finding was not typical. There was no evidence of bile duct or vascular invasion (Fig. 1).

Tumor markers were normal with CEA and CA-19-9 levels of 1.8 ng/ml (reference range: 0-5 ng/ml) and 9.2 U/ml (reference range: 0-37 U/ml). Chromogranin A, insulin, and glucagon levels were 45.21 ng/ml (reference: 24-97 ng/ml), 5.2 μIU/ml (reference: 2-25 μIU/ml), and 28.7 pg/ml (reference: 40-130 pg/ml), respectively.

An operation was planned as the tumor was thought to be possibly malignant or borderline malignant. Ascites, shelf, or seeding were not present, and a 2×1 cm sized firm mass was palpable at the pancreas head. Pylorus preserving pancreaticoduodenectomy was performed. On gross cross section examination of the specimen, the mass was located at the head of the pancreas. It was a well encapsulated, whitish mass with lobulations. Degenerative changes or hemorrhage were not visible (Fig. 2). The patient recovered uneventfully and was discharged on the 9th postoperative day in good condition.

Microscopic examination revealed a well-defined pancreatic parenchymal mass with a pushing border. Hematoxylin-eosin staining showed benign duct and glandular structures surrounded by proliferating smooth muscle cells. Acinus formation was not observed (Fig. 3). Immunohistochemical stain for smooth muscle actin (SMA) was positive in the spindle cells and CD34 was negative (Fig. 4). The histopathology diagnosis was myoepithelial hamartoma (adenomyoma).

DISCUSSION

Non-neoplastic mass-forming lesions of the pancreas which include hamartoma are extremely rare. Hamartomas are defined as a localized excessive overgrowth of mature normal cells and tissues in an organ composed of identical cellular elememts but without the normal architecture of the surrounding tissue. Hamartoma of the pancreas can be divided into two subgroups: “solid and cystic lesion” and “solid mass”.[3] There have been 15 reported cases of pancreatic hamartoma, and only 3 of them were documented solid hamartoma. The 3 documented solid hamartomas by Pauser et al.[4] and Nagata et al.[3] were positive for CD34

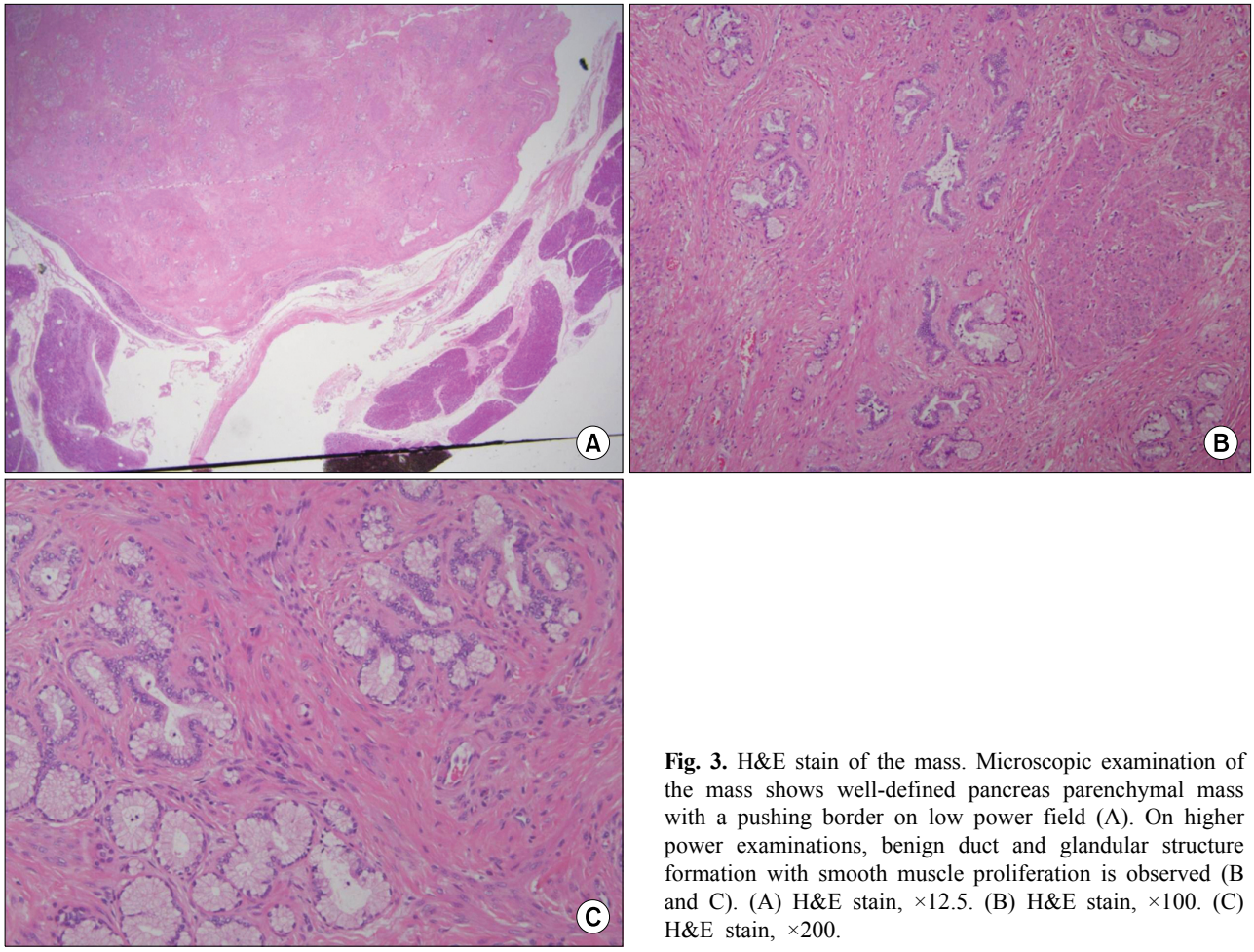

Fig. 3. H&E stain of the mass. Microscopic examination of the mass shows well-defined pancreas parenchymal mass with a pushing border on low power field (A). On higher power examinations, benign duct and glandular structure formation with smooth muscle proliferation is observed (B and C). (A) H&E stain, ×12.5. (B) H&E stain, ×100. (C) H&E stain, ×200.

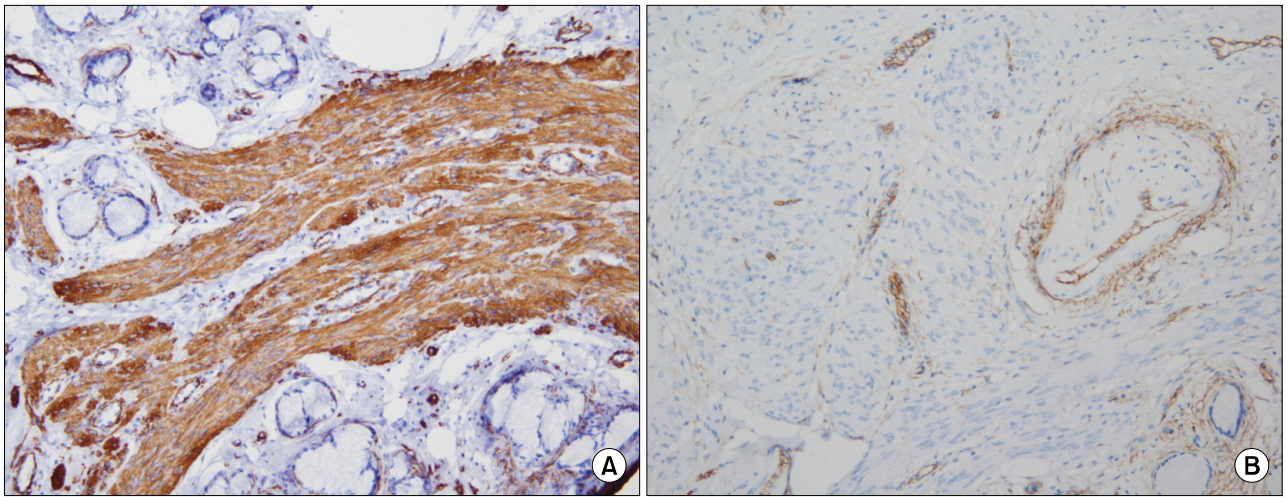

Fig. 4. Immunohistochemical stain of the mass. The spindle cells show positive staining for smooth muscle actin (SMA) (A) and negative staining for CD34 (B). (A) SMA stain, ×200. (B) CD34, ×200.

and c-kit, resembling GIST. The current case of solid pancreatic hamartoma is distinguishable from these 3 cases by the finding of negative CD34 and the presence of smooth muscles (SMA positive) in the present case. Also, there are no reports of MEH in the pancreas to date, adding uniqueness to the current case.

MEH and heterotopic pancreas or ectopic pancreas should be understood as a spectrum of lesions of the same process. Heterotopic pancreas is characterized by the presence of pancreatic acinar, islet, or ductular elements, usually in association with smooth muscle proliferation, outside of the topographic boundaries of the pancreas.[5] Abnormally arranged pancreatic elements that lack acini formation and islet-like tissue and consist of duct and smooth muscle proliferation are termed MEH or adenomyoma. The pathogenesis of MEH was presented by Lewin and Appleman,[6] who stated that MEH in the gastrointestinal tract arises during embryogenesis from pancreatic metaplasia or, alternatively, from a displaced pancreatic anlage. The degree of differentiation of the tissue determines the final diagnosis. The reported sites of occurrence of MEH include the gastrointestinal tract and the biliary tree. Whether adenomyomas of the gallbladder and extrahepatic bile duct are related to MEHs in the intestinal tract in terms of the pathogenesis is controversial. Some scholars believe that adenomyomas of the biliary tree are related to diverticulosis,[7] and are unrelated to pancreatic anlage. Thus, the discussion will limit MEHs to gastrointestinal MEH.

The incidence of MEH is unclear and is only known to be rare. The reported sites of occurrence include stomach, duodenum, jejunum, ileum, and rectum. The stomach seems to be the most common site of occurrence, with 38 case reports until 1998 since the first report in 1903.[2] From 1940 to 2011, twenty-four cases of adenomyoma have been reported in the small intestine, mostly in the ileum.[8] Four cases of duodenal MEHs were reported by Ryan and colleagues[2] in 1997, and a single case of rectal MEH was reported in 1961 by Pouget and Bacon.[9]

Most of the MEHs are thought to be symptomless and go undetected. However, most of the cases that are reported presented with symptoms and signs such as bleeding or obstruction.[8,10-14] Other than such extrinsic problems, Ryan et al.[2] state that anything that can occur in the pancreas can happen in heterotopic pancreatic tissue, including pancreatitis and carcinoma. Cases of adenocarcinomas associated with adenomyomas in the stomach, duodenum or jejunum have been reported as well.[15] It is unclear and doubtful whether adenomyoma undergoes malignant change or increases the risk of malignancy. Even so, the statement that MEH is completely benign should not be taken for granted.

Our case report is the first to report an MEH as a solitary mass in the pancreatic parenchyma. Currently, no such disease entity exists in the literature and textbooks of pathology. The microscopic finding is consistent with the definition and common features of MEH. Therefore, it could not be but diagnosed as MEH histologically. In order to acquire better understanding of MEH in the pancreas, we urge more case reports of MEH in pancreas be made when encountered.

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