Oncocytic Type Intraductal Papillary Mucinous Neoplasm Mimicking Mucinous Cystic Neoplasm of the Pancreas
- A Case Report -

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Intraductal papillary mucinous neoplasm (IPMN) of the pancreas is now a well-recognized clinical and pathologic entity. IPMN usually results in a dilated pancreatic duct, which frequently communicates with the tumor. IPMN is commonly seen in the head of the pancreas. Mucinous cystic neoplasm (MCN) has minimal clinicopathological differences compared to IPMN and is more frequently seen in the body and the tail of the pancreas, involving the peripheral pancreatic ductal system. Oncocytic cells are characterized by an abundance of mitochondria. In the pancreas, tumors composed predominantly of oncocytic cells are rare. In 1996, Adsay et al. proposed a new disease entity known as an intraductal oncocytic papillary neoplasm (IOPN) of the pancreas. Recently, IOPN was classified as an oncocytic type of IPMN. However, the clinical and pathological characteristics of IOPN remain to be elucidated. We report one case of IOPN that is clinically and pathologically different from those cases described in past reports.

CASE REPORT

A 35-year-old woman visited our hospital in April 2007 with a chief complaint of epigastric pain for one month. Her family history and past medical history were unremarkable. Laboratory data at admission were within the normal range. Enhanced computed tomography (CT) revealed a multilocular cystic mass with a maximum diameter of approximately 15 cm located in the distal body and tail of the pancreas. A nodular enhancing lesion was present inside the multilocular cystic tumor (Fig. 1A). Endoscopic retrograde cholangiopancreatography (ERCP) showed no communication between the mass and the main pancreatic duct, and no ductal dilatation was seen (Fig. 1B). Under the preoperative diagnosis of MCN with hemorrhagic contents, a distal pancreatectomy and splenectomy were performed. The surgically resected specimen showed a well-demarcated cystic mass measuring 15 × 12 × 10 cm, located in the distal body and tail of the pancreas. Serial sectioning showed a multilocular cyst with brownish mucous and hemorrhagic jelly-like contents. Ductal communication was not noted. There was a nodular papillary projection measuring 6 × 4 × 4 cm in diameter in the cystic wall (Fig. 2A). The wall of the cyst was fibrotic, well-defined, and thin. Microscopical examination also demonstrated a fibrous cyst wall, with papillary nodules with thin stalks. The papillary nodule was composed of plump cells with abundant eosinophilic cytoplasm. Red granules were detected in the cytoplasm of tumor cells on modified Gomori trichrome stain. Ultrastructurally, the tumor cells contained abundant cytoplasm packed with numerous mitochondria and intercellular lumina. We describe an oncocytic type intraductal papillary mucinous neoplasm having the clinical characteristics of a mucinous cystic neoplasm.

Key Words : Intraductal papillary mucinous neoplasm; Oncocytic type; Pancreas
and granular cytoplasm (Fig. 2B). A spectrum of architectural complexity was seen, ranging from simple, stratified lining, to extensive papillary arborizations; these findings were highly suggestive of carcinoma. The cellular ovarian-type stroma as seen in classic MCN was not present in this case. A mucinous component in the form of interspersed goblet-type vacuolations and glandular lumina was present throughout the neoplasm. Adjacent pancreas revealed fibrosing pancreatitis with acinar atrophy and scattered lymphocytes. Red granules were detected in the cytoplasm of tumor cells on modified Gomori trichrome stain. Immunohistochemically, the oncocytic tumor cells showed

Fig. 1. Radiologic images of the pancreatic cystic tumor. (A) The enhanced abdominal CT shows multi-loculated macrocystic mass with partial enhancing solid lesion on body and tail of the pancreas. (B) There is no definite evidence of communication between the pancreatic duct and cystic mass on ERCP.

Fig. 2. Pathologic findings of the pancreatic mass. (A) The cut section reveals a multilocular cystic lesion with nodular papillary projection in the inner wall surface (arrow). (B) Polypoid tumor exhibits extensive arborizing papillary growths and focally cribiform pattern, lined by plump cells with abundant eosinophilic and granular cytoplasm (H&E, ×400).

Fig. 3. Ultrastructurally, tumor cells show numerous mitochondria and intracellular lumen in the cytoplasm (bar in A, 470 nm; bar in B, 1 μm).
a positive p53 signal (DakoCytomation, Carpinteria, CA, USA) and a high Ki-67 (Zymed Laboratories, Inc., South San Francisco, CA, USA) labeling index. Ultrastructurally, the tumor cells contained abundant cytoplasm packed with numerous mitochondria (Fig. 3A). Intracellular and intercellular lumina were present (Fig. 3B). These clinicopathologic findings led to the diagnosis of oncocytic type IPMN having clinical characteristics of MCN.

DISCUSSION

Oncocytic differentiation has been described in acinar and islet cell neoplasms, as well as in solid-pseudopapillary neoplasms of the pancreas, but it is uncommon in intraductal and cystic tumors of the pancreas. Adsay et al. reported 11 cases of intraductal mucin producing pancreatic tumors with oncocytic features and proposed the term IOPN for this entity. Recently, IOPN was classified as an oncocytic type of IPMN. This tumor is defined by the following pathologic features: a pancreatic duct lesion signifying complex, arborizing, and proliferating papillary structures; tumor cells with a finely granular, eosinophilic cytoplasm; and diffusely scattered goblet cells. In addition, the cytoplasm of tumor cells stains diffusely with anti-mitochondrial antibodies. Electron microscopic images show abundant tumor cell cytoplasm rich with mitochondria. Our case also manifested these characteristics. IPMN and IOPN share a number of features, but our case did not demonstrate the pancreatic duct dilatation or communication with the main pancreatic duct frequently seen in IPMN. However, reports of dilatation of the main pancreatic duct or mucin excretion from the duodenal papilla are limited in IOPN. In addition, a recent report described a case of IOPN having the clinical characteristics of MCN. Microscopically, MCN can be easily differentiated from IOPN. Papillary structures, if present, are often less complex and arborizing in MCN, and MCN lacks oncocytic cells and intraepithelial lumina. On microscopic examination, IOPN typically lacks cellular ovarian-type stroma, which is characteristically present in MCN.

IPMNs are currently divided into benign, borderline, and malignant lesions. Intraductal papillary mucinous carcinoma shows severe dysplastic epithelial change with cribriform pattern and micropapillary growth. We believe that our case reveals enough cytoarchitectural atypia to confer on it the diagnosis of intraductal papillary mucinous carcinoma. Furthermore, this case exhibited the positive p53 signal and high Ki-67 labeling index frequently observed in malignant tumors. Because the oncocytic type of IPMN is a recently recognized entity, at present there is inadequate literature information to make a definitive statement concerning its clinicopathologic behavior.

In conclusion, the present case manifested characteristics that were different from those seen in previously described oncocytic type IPMNs which had dilated ducts communicating with the main tumor located in the pancreatic head. The clinical significance of the oncocytic type of IPMN is unclear at this time, due to the scarcity of clinical reports, and this tumor warrants further investigation.

REFERENCES