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CASE REPORT

췌장후부에서 발생한 원발성 후복막 점액낭샘종

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A Case of Primary Retroperitoneal Mucinous Cystadenoma Arising from the Retropancreatic Area

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Primary retroperitoneal mucinous cystadenoma is an extremely uncommon tumor, even though mucinous cystadenoma often develops in the ovary and less frequently in the pancreas. A 21-year-old female was admitted to our hospital due to severe abdominal pain. A well-demarcated, oval shaped cystic tumor at the retropancreatic area with displacement of the pancreas and surrounding major vessels was observed on CT and MRI. Exploratory laparotomy was performed, and complete excision of the entire cyst was performed without complication. The pathologic finding was consistent with primary retropancreatic mucinous cystadenoma. To the best of our knowledge, this report is the first to describe a case of retropancreatic mucinous cystadenoma arising from the retropancreatic area in Korea. **(Korean J Gastroenterol 2014;63:187-190)**

Key Words: Mucinous cystadenoma; Retroperitoneal; Primary

INTRODUCTION

Primary retroperitoneal mucinous cystadenoma (PRMC) is an extremely uncommon tumor, even though mucinous cystadenoma often develops in the ovary and less frequently in the pancreas.[1] Gross and microscopic appearance of PRMC is similar to that of mucinous cystadenoma of the ovary.[2] It can arise at various locations in the retroperitoneal area. PRMC usually remains asymptomatic in the earlier stage. However, as the size of the tumor increases, symptoms generally develop due to pressure and obstruction on adjacent organs.[3] Imaging and laboratory studies are not diagnostic; therefore, surgical resection is needed in order to establish an accurate diagnosis and definite treatment. We report on a case of PRMC arising from the retropancreatic area. To the best of our knowledge, this was the first case reported in the literature to describe a PRMC arising from the retropancreatic area.

CASE REPORT

A 21-year-old female was admitted to our hospital due to severe abdominal pain lasting two days. The patient had dull-natured epigastric pain, radiating to the back area. On physical examination of the abdomen, she complained of epigastric tenderness without a palpable mass. She denied any systemic disease and previous surgery or drug history. The initial vital signs were stable.

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Laboratory tests on admission were as follows: white blood cells 8,470/mm^3, hemoglobin 14.1 mg/dL, platelet 303,000/mm^3, AST 23 IU/L, ALT 14 IU/L, amylase 110 IU/L, and lipase 25 IU/L. Serum CEA and CA 19-9 levels were 20.7 ng/mL (reference 0-5) and 41.5 U/mL (reference 0-37), respectively. A well-demarcated, oval shaped cystic tumor measuring 3.0×6.0 cm in size at the retropancreatic area with displacement of the pancreas and surrounding major vessels was observed on contrast enhanced CT and MRI (Fig. 1). The cystic mass consisted of protein-rich liquid contents with irregular thickening of the cyst wall (Fig. 2).

An exploratory laparotomy was performed. The cystic mass was located at the posterior aspect of the pancreatic neck portion with compression of the splenic vein. The cyst was not adhered to the pancreas and could be easily dissected without opening of the pancreatic capsule. Thick mucus contents were aspirated from the cyst. Complete excision of the cyst was performed without complication. Gross examination showed a cystic mass measuring 5.5×3.5 cm in size and weighing 11.8 g, which consisted of thick mucus material on the cut surface (Fig. 3). Histopathologic examination showed that the cyst wall was lined with a single layer of mucinous epithelial cells (Fig. 4). The lining of the cyst consisted of tall columnar cells and cuboidal cells. No pancreatic compounds were found in the resected specimen. Results of immunohistochemical staining were positive for MUC2 and MUC5AC antibodies. The pathologic findings were consistent with primary retropancreatic mucinous cystadenoma. The patient made a full recovery, with no postoperative complications and remained asymptomatic on regular follow-up.

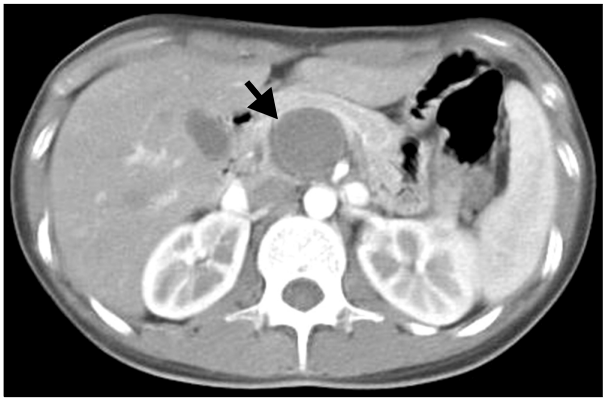

Fig. 1. CT scan of the abdomen. A well-demarcated oval shaped cystic tumor measuring 3.0×6.0 cm in size at the retropancreatic area with displacement of the pancreas and surrounding major vessels was observed (black arrow).

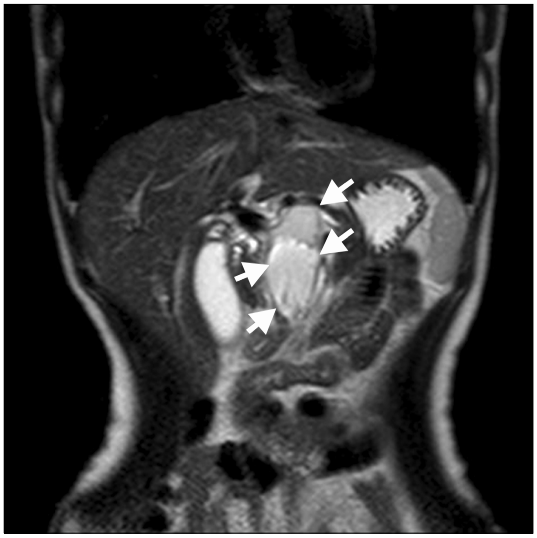

Fig. 2. MRI of the abdomen. The cyst consisted of protein rich liquid contents with irregular thickening of the cyst wall (white arrows).

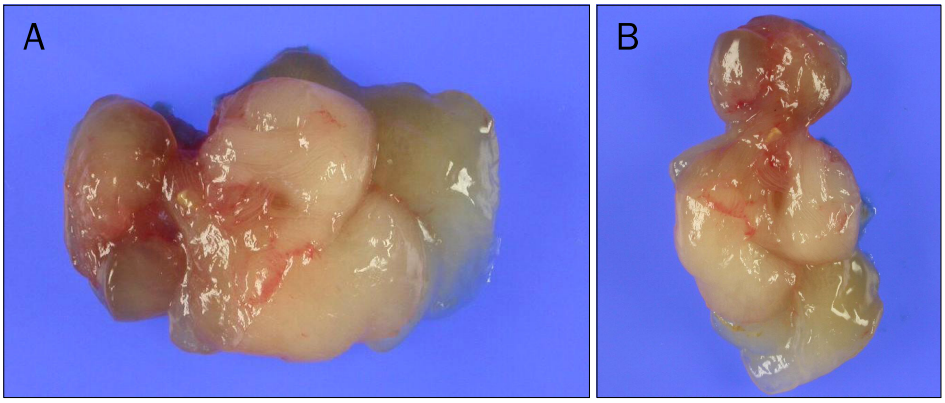

Fig. 3. Gross finding of the resected specimen. The cystic mass measured 5.5 x 3.5 cm in size and weighed 11.8 g, and consisted of thick mucus material on the cut surface.

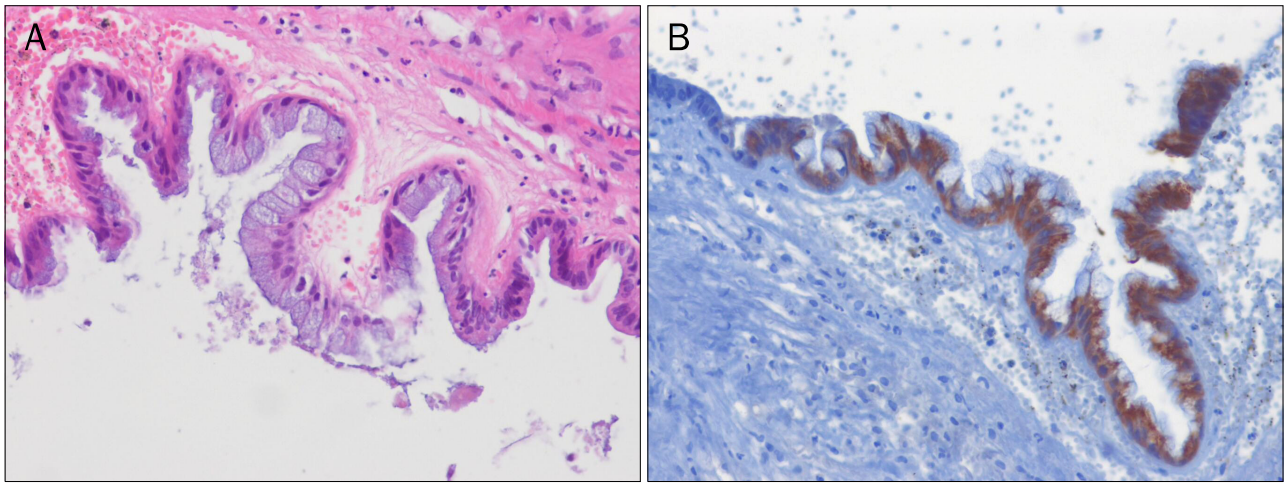

Fig. 4. Microscopic finding. (A) The cyst wall was lined with a single layer of mucinous epithelial cells (H&E, ×200). (B) On immunohistochemical staining, MUC2 and MUC5AC antibodies were positive (×200).

DISCUSSION

PRMC is a very rare neoplasm. Therefore, its epidemiology is not yet well established.[2] PRMC is usually found incidentally in asymptomatic patients, and is more common in young women. Gross and microscopic features of PRMC are similar to those of benign mucinous cystadenoma of the ovary.[4] Both neoplasms are multiloculated cystic tumors, lined with a single layer of tall columnar cells.[5] In addition, cells of both tumors contain clear cytoplasm and a basal nucleus. The histologic difference is the presence of cuboidal cells in PRMC, compared to ovarian mucinous cystadenoma.[5] In our case, the lining of the cyst was composed of tall columnar cells and cuboidal cells.

The histogenesis of retroperitoneal mucinous cystadenoma is unclear; however, several theories explain its formation.[6] According to the hypotheses, the tumor can arise from ectopic ovarian tissue, teratoma, and remnants of the embryonic urogenital apparatus.[7] According to one recent hypothesis, a tumor can arise from invagination of the peritoneal mesothelial layer that undergoes mucinous metaplasia.[5] In our case, PRMC was located at the retropancreatic area; therefore, this case can more likely be explained by a recent hypothesis.

Cystic mesothelioma, cystic lymphangioma, and pancreatic pseudocyst can be confused with PRMC.[5] An exploratory laparotomy is usually indicated for a definite diagnosis. In our case, the cystic mass was difficult to distinguish from a pancreatic cyst on pre-operative imaging studies. However, diagnosis of PRMC was possible because the cystic mass was positioned at the retropancreatic area and could easily be separated from the pancreas without opening of the pancreatic capsule during the operation.

Although pre-operative diagnosis of PRMC is not easy, imaging modalities, including CT and MRI, can facilitate achievement of an accurate diagnosis.[3] PRMC usually manifests as a homogeneous, unilocular cyst on the CT scan, and can appear as hyperintense on T1-weighted images on MRI.[8] In our case, CT scan showed a round, unilocular cystic mass measuring 3 cm in size and MRI showed a well defined tubular cyst with high signal intensity fluid content.

Malignant potential is high in patients with PRMC because ovarian mucinous cystadenoma can progress to malignancy, and cases of primary retroperitoneal mucinous cystadenocarcinoma have been reported.[9] Therefore, complete surgical excision is recommended for treatment of PRMC, as well as accurate diagnosis in order to eliminate any risk of malignant change, even in asymptomatic patients.[10]

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