Mucinous cystadenoma of the spleen

Report of a rare case

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ABSTRACT

A rare case of mucinous cystadenoma of the spleen is reported. The patient was a 63-year-old female with a cystic mass in the spleen demonstrated by echo and CT scan. Surgical exploration revealed the tumor was confined in the spleen with extravasation of mucinous material from a rupture into left subphrenic space. Microscopically, the cystic space was lined by a single layer of mucin-producing columnar epithelial cells without malignant change. We reviewed the literature and found this may be the second case of splenic mucinous cystadenoma.

Key words: mucinous cystadenoma, spleen

INTRODUCTION

Mucinous cystadenomas are relatively uncommon benign cystic tumors. Most

of them are found in the ovary, pancreas, and appendix. However, they have also been identified in other unusual sites such as retroperitoneum **(1)**, fallopian tube **(2)**, lung **(3)**, urinary bladder **(4)**, liver **(5)** and spleen **(6).** In the English literature, there were three cases of splenic mucinous cystadenocarcinoma reported previously **(7,8,9)**, and Miracco reported a case of splenic cyst lined with mucin-secreting epithelium **(6)**. Herein, we describe a mucinous cystadenoma of the spleen in a 63 year-old woman.

CASE REPORT

A 63 year-old Taiwanese woman had had a mass in her left abdomen for one year. She had also suffered from cerebrovascular accident one year earlier. Otherwise, her past history was unremarkable.

Physical examination revealed a palpable mass about two finger-breadths below her left costal margin. Laboratory data was unremarkable. Abdominal plain film showed a faint mass shadow in left upper abdomen, about 7 cm in diameter. Abdominal ultrasonography revealed a cystic lesion in the spleen with multiple internal septa, some of which had strong echogenicity **(Figure 1).** Computerized tomography showed an 8 cm low-density cyst with internal septa and peripheral calcification in the spleen **(Figure 2)**.

On surgical exploration, splenectomy was done. A multiloculated cystic mass was confined in the spleen and had a rupture on its upper pole, from which mucinous

material came out and into the left subphrenic space. There was no any communication between the spleen and the pancreas. Grossly, the tumor was measured about 9×6×4.5 cm in diameter. The cut surface showed multiloculated cystic mass with smooth inner surface and profuse insipissated mucus. Microscopically, papillary structure was found on the inner surface of cystic space , which was lined by a single layer of mucin-producing columnar epithelial cells without malignant change **(Figure 3).** The splenic tissue was outside the cystic wall. No pancreatic tissue was found in the specimen. So, mucinous cystadenoma of the spleen was diagnosed although calcification was even found.

The postoperative course was uneventful. She was discharged one week later, and abdominal ultrasonography demonstrated no recurrence one year later.

DISCUSSION

Splenic tumors are uncommon neoplasms. In 1985, Morgenstern et al. **(10)** classified splenic tumors roughly into four categories: lymphoid, non-lymphoid tumors, metastatic tumors, and tumor-like lesions, such as cysts and hamartomas. The most common non-lymphoid tumors are vascular tumors. However, primary splenic tumors from epithelial origin appear to be extremely rare.

From a few of case reports, splenic mucinous cystic tumors were defined as

cystic spaces lined by mucin-producing columnar cells and ranged from benign cystadenoma to frankly malignant cystadenocarcinoma. To our knowledge, there were three cases of mucinous cystadenocarcinoma in the spleen reported previously. Two of them revealed a mucinous tumor arising in an aberrant intrasplenic pancreas **(7,8).** The third case of mucinous cystadenocarcinoma in the spleen arose from undefined origin **(9)**. The author postulated that the splenic lesion was a delayed metastatic recurrence of appendiceal carcinoma, either from a developmental misplacement of endodermal epithelial tissue such as a heterotopic pancreas, or from mesothelium of the splenic capsule. On the other hand, there was a case of splenic mucinous cystadenoma according to the description of Miracco **(6)**. So, our case may be the second case of splenic mucinous cystadenoma.

Clinical presentation of patients with mucinous cystic tumors depends on the site of the tumor and usually reveals nonspecific findings. In the image diagnosis, sonography and CT scan may demonstrate a cystic lesion containing internal septum or not. Peripheral calcification in the tumor may be seen more frequently in cystadenocarcinoma than in cystadenoma **(11)**, but the rule is not applied to our case. Anyway, surgical intervention is needed for their definite diagnosis.

The exact histogenetic mechanism of splenic mucinous cystic tumors is unknown except those arise from heterotopic pancreatic tissues **(7,8)**. Elit et al.

reported a squamous cell carcinoma in an epidermoid cyst of the spleen **(12)** and Ough et al. reported a case of mesothelial cyst of the spleen with squamous metaplasia **(13)**. Epidermoid cysts of the spleen are postulated that epithelial lining cells of the cysts may originate in an invagination of the splenic capsular mesothelium that undergoes metaplastic changes **(13)**. Invagination of the capsular mesothelium may be congenital or traumatic. Our case is a mucinous cystadenoma of the spleen with a rupture on its upper pole which caused focal “pseudomyxoma peritonei” in the left subphrenic space. There are no definite heterotopic pancreatic tissues in and around the splenic lesion. So, we don’t know what is the exact histogenetic mechanism. By way of Morinaga’s presentation **(9)**, we postulate that the splenic tumor may originate in the invagination of the splenic capsular mesothelium.

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FIGURE LEGENDS

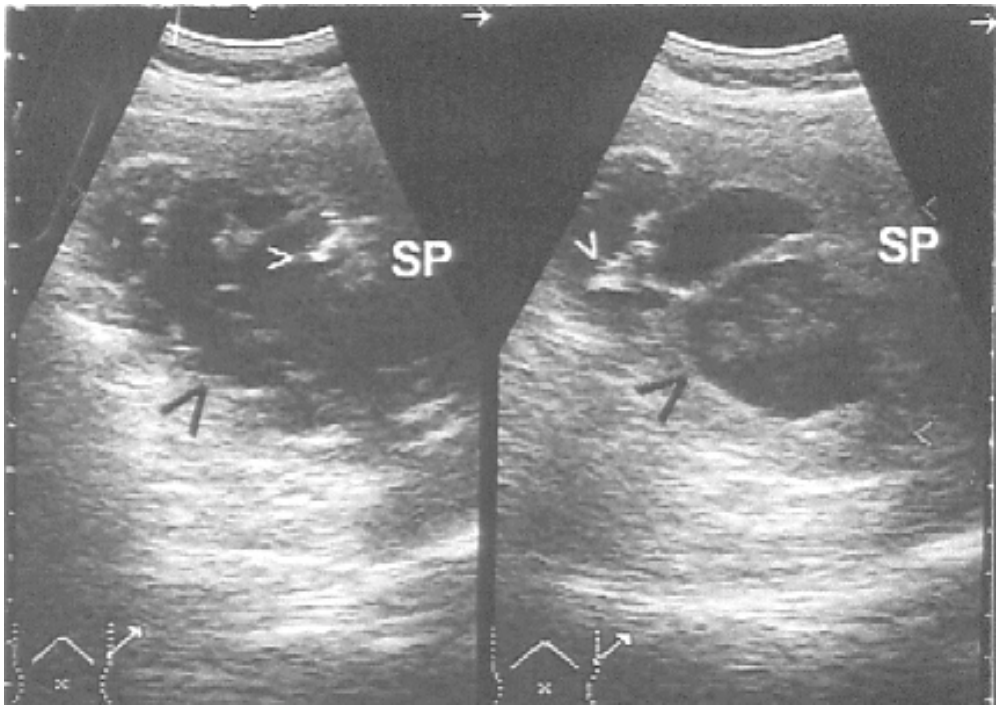

Fig.1. Abdominal echo shows a cystic lesion in spleen (large arrowhead) with multiple internal septa. Some of the septa have strong echogenicity (arrowheads). SP: spleen

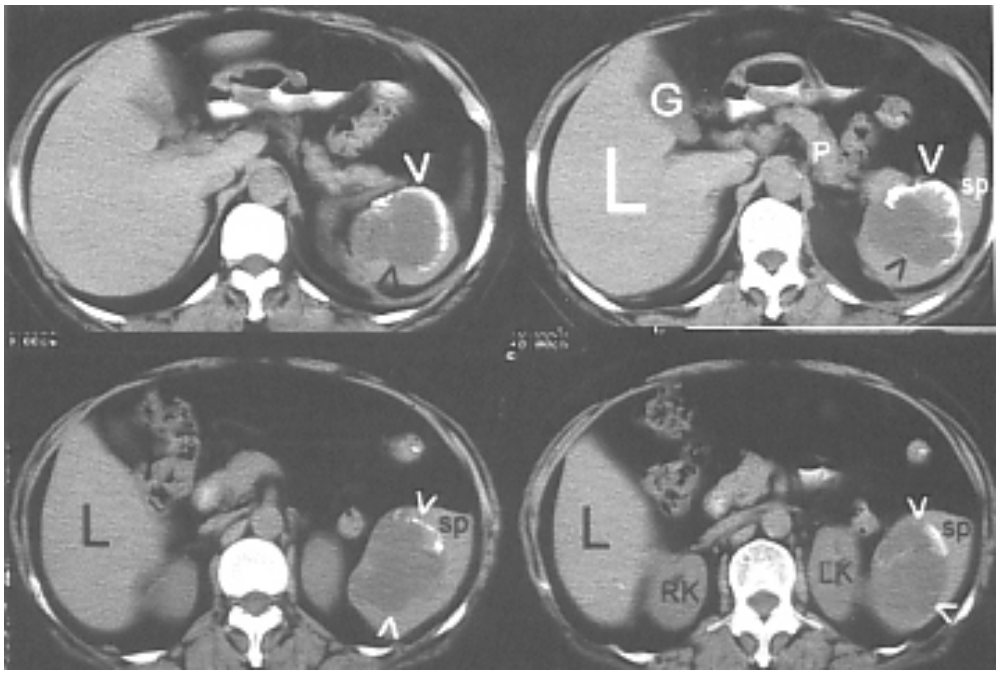

Fig.2. A series of abdominal CT of the patient shows an 8-cm low density mass (small arrowhead) with peripheral calcifications (large arrowhead) and internal septa in the spleen. SP: spleen; L: liver; G: gallbladder; RK: right kidney; LK: left kidney; P: pancreas

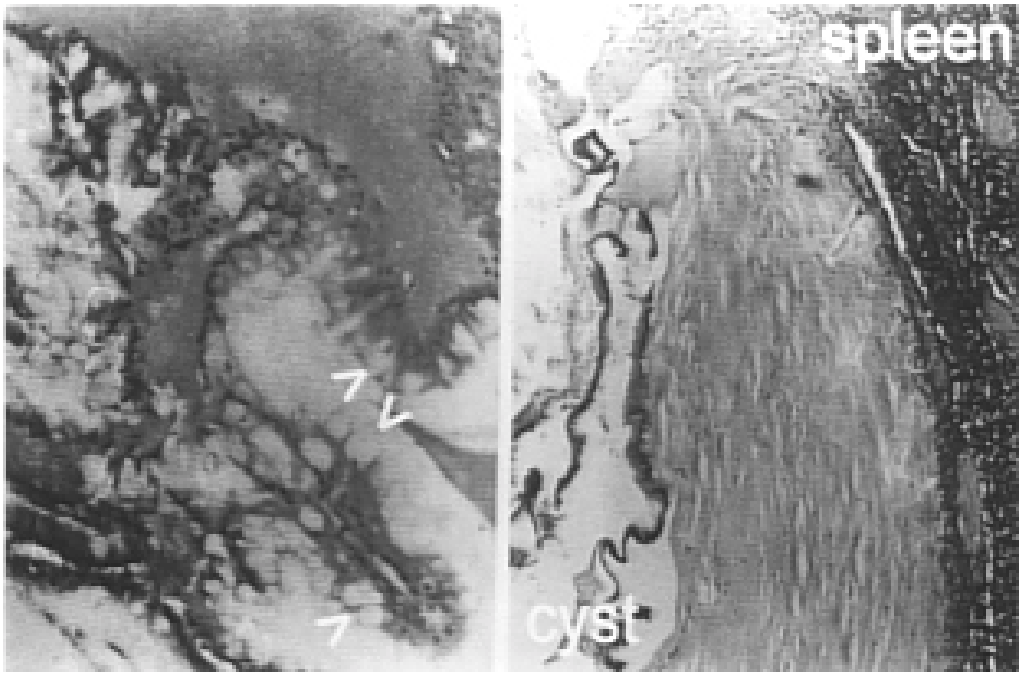

Fig.3. Right: Cystic structure is found in the splenic tissue. (H & E stain 40x)

Left: The lining epithelium of the cyst is columnar epithelial cells with prominent goblet cells (arrowheads) in papillary projection configuration (H & E stain 100x)