A case of splenic low-grade mucinous cystadenocarcinoma resulting in pseudomyxoma peritonei

Abstract
Primary splenic mucinous cystadenocarcinoma (MCCa) is extremely rare, and only six cases appear to have been reported previously. We present herein a case of primary splenic MCCa resulting in pseudomyxoma peritonei (PMP). A 66-year-old Japanese woman presented to a hospital with a chief complaint of upper abdominal pain and a 7-year history of splenic cyst. Cyst rupture was noted on computed tomography, and splenectomy was performed. The abdominal cavity was filled with a large amount of gelatinous ascites, with the appearance of PMP. On the cut surface, multiple cysts containing mucinous material were found within and outside the spleen. Microscopically, splenic parenchyma was occupied by large mucinous pools focally lined with mucinous epithelial cells and mesothelial cell-like cells, which were considered benign. Outside the spleen, a low-grade MCCa component was found. No ectopic pancreatic or intestinal tissue was identified. Although most PMP cases are known to be caused by low-grade mucinous appendiceal tumor, the present case represents the first report of a splenic MCCa resulting in PMP.

Key words
Mucinous cystadenocarcinoma · Spleen · Pseudomyxoma peritonei

Introduction
Primary nonhematopoietic malignant tumor of the spleen is extremely rare, and primary splenic mucinous cystadenocarcinoma (MCCa) is even more rare. To the best of our knowledge, only six cases have been reported previously. However, there have been no reports of primary splenic MCCa associated with PMP. We present herein the first case of primary splenic MCCa resulting in PMP.

Clinical summary
A 66-year-old Japanese woman was admitted to hospital with a chief complaint of upper abdominal pain. A splenic cyst had been identified on a medical check-up 7 years earlier. She had undergone right upper lobectomy for pulmonary tuberculosis 45 years earlier and uterine myomectomy 22 years earlier.

Laboratory values were unremarkable, other than slightly elevated levels of carcinoembryonic antigen (CEA) (9.1 ng/ml; upper normal limit, 5 ng/ml) and carbohydrate antigen (CA)19-9 (59.9 U/ml; upper normal limit, 37 ng/ml). Peripheral blood counts were normal. Fiberoptic studies of the gastrointestinal tract showed no mucosal abnormalities. Abdominal computed tomography (CT) revealed a huge multicystic mass within the spleen. The pancreas was normal and separate from the spleen. Ascites was noted in the perisplenic and perihepatic areas, and was suspected to have been caused by rupture of the cysts. No history of trauma that might have caused the rupture was elicited.

Magnetic resonance imaging also showed a multicystic splenic tumor with low signal intensity on T1-weighted imaging and high signal intensity on T2-weighted imaging (Fig. 1a). The pouch of Douglas was filled with a large amount of ascites, including floating, irregular nodal architecture (Fig. 1b). Magnetic resonance cholangiopancreatography showed no dilatation of the common bile duct or main pancreatic duct.

Splenectomy and biopsy of the peritoneum and diaphragm were performed. The abdominal cavity was filled with a large amount of gelatinous ascites and mucinous nodules, with an appearance consistent with that of PMP. A large multicystic mass containing mucinous material was found within the spleen. This mucinous material had leaked around the stomach and beneath the diaphragm. No connections were found between the spleen and pancreas. The pancreas, liver, and gastrointestinal tract appeared normal.
and intact. The appendix was previously resected, probably at the time of uterine myomectomy, and the appendiceal resection stump was clear. Both ovaries appeared normal. Postoperatively, serum CEA and CA19-9 levels normalized. The patient received chemotherapy for PMP after providing informed consent. Repeated CT examinations have demonstrated no ascites or intraabdominal masses. As of 11 months postoperatively, the patient was well without evidence of recurrence or metastasis.

Materials and methods

Sampling tissues

Written informed consent was obtained to use the surgical samples for pathological examination, research, and publication. Excised specimens were fixed in formalin and embedded in paraffin. Sections 3 μm thick were prepared and stained using hematoxylin and eosin. Periodic acid–Schiff and Alcian blue double-stainings were also performed. Immunohistochemical staining was performed using an autostainer (XT System Benchmark; Ventana Medical Systems, Tucson, AZ, USA) according to the manufacturer’s instructions with an ultra View TM Universal DAB Detection kit (Ventana, Yokohama, Japan) for the following primary antibodies: cytokeratin (CK)7 (OV-TL 12/30) (Dako, Glostrup, Denmark); CK20 (Ks20.8) (Dako); CEA (rabbit polyclonal) (Dako); CA19-9 (116-NS-19-9) (Japan Tanner, Osaka, Japan); MUC2 (Cep58) (Novocastra, Newcastle-upon-Tyne, UK); MUC6 (CLH5) (Novocastra); M-GGMC-1 (HIK1083) (Kanto Chemical, Tokyo, Japan); calretinin (rabbit polyclonal) (Zymed, San Francisco; CA, USA); WT-1 (6F-H2) (Dako); D2-40 (D2-40) (Dako); p53 (DO-7) (Novocastra); and Ki-67 (MIB-1) (Dako).

Results

Macroscopic findings

The spleen measured 13 × 10 × 7 cm and weighed 580 g. On the cut surface, the parenchyma was widely occupied by multiple cysts containing large amounts of yellowish, translucent, mucinous material. The diameter of the largest cyst was 5.5 cm. Variously sized cysts were also found outside the spleen (Fig. 2). The intra- and extrasplenic cysts were connected via a common cyst wall.

Microscopic findings

Multiple cysts in the splenic parenchyma were surrounded by thick, fibrous tissue. The inner lining epithelium was widely desquamated, and tall, columnar, mucinous cells remained in some parts. Mesothelial cell-like cells were also observed. Transitions between mucinous and mesothelial cell-like cells were identified (Fig. 3a). Both cell types showed no atypia and a papillary configuration. Leakage of mucinous fluid into the splenic parenchyma was observed. On the other hand, the variously sized cysts outside the spleen were lined by taller and partly stratified epithelium with hyperchromatic and somewhat irregular nuclei (Fig. 3b). Furthermore, a papillary configuration and back-to-back structures were also observed (Fig. 3c,d). These carcinomatous components were only found outside the spleen.