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Case report

Pseudomyxoma retroperitonei

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Abstract. We report a rare case of pseudomyxoma retroperitonei in a 58-year-old woman with a past history of severe appendicitis. The imaging showed a multicystic mass similar to pseudomyxoma peritonei, but the tumor was located in the retroperitoneal space.

Key words: Retroperitoneal neoplasms – Retroperitoneal space – Computed tomography – Magnetic resonance imaging – Ultrasonography

Introduction

Pseudomyxoma peritonei (PP) is a rare but well-known condition in which masses of gelatinous and mucinous material spread and accumulate throughout the peritoneal cavity, resulting usually from the rupture of a mucinous lesion of the appendix or ovary [1–7].

Most pseudomyxomas are in the peritoneal cavity. To our knowledge, only 12 cases of pseudomyxoma retroperitonei (PRP) have been reported [8–17]. We report one such case of PRP that developed as a multicystic mass in the retroperitonal space.

Case report

A 58-year-old woman suffering abdominal pain was referred to our hospital. An appendectomy had been performed 28 years previously. She said that the appendicitis had been severe.

On ultrasonography (US) (Fig. 1), computed tomography (CT) (Fig. 2) and magnetic resonance (MR) imaging (Fig. 3), a huge multicystic mass was seen in the right retroperitoneal space. There was some internal echo in the tumor on US. The density of the mass on CT and the intensity on T1-weighted MR imaging was low, but slightly higher than that of the urine. T2-weighted MR imaging showed a high-intensity mass. The mass had a thick capsule and septa, with tiny calcifications and nodes. Post-contrast studies on CT and MR imaging showed enhancement of the walls and nodes. The right kidney and the ascending colon were shifted anteriorly. The lesion extended to the pelvic extraperitoneal space below the right iliac muscle. The right ureter in the pelvis was shifted medially, and the urinary bladder was compressed by the lesion. The images were similar to PP. However, the lesion was not located in the peritoneal cavity, but in the retroperitoneal space. Hence we made a preoperative diagnosis of PRP.

Transabdominal exploration located the retroperitoneal cystic masses. There was no lesion in the peritoneal cavity. Neither the appendix, the stump of the appendix, nor an ovarian tumor was found. Histological examination revealed mucinous cystadenoma. It could not be determined histologically whether the lesion was secondary or primary.

Discussion

PP [1–7] is characterized by multiple, massive implants containing abundant gelatinous or myxomatous material. PP occurs when these mucin-producing lesions rupture into the peritoneal cavity. Potential primary sites include the mucocele of the appendix and the mucinous neoplasm of the ovary; also the uterus, bowel, urachus, common bile duct, and pancreas, among others. However, its origin may not be clear due to extensive organ involvement. Both the malignant and benign forms have the same macroscopic appearance and also share many common microscopic features. The US, CT, or MR appearance of PP is of loculated fluid collections, hepatic scalloping, and masses with thick cystic walls or septa. Curvilinear or punctate calcifications are often seen on CT [6].

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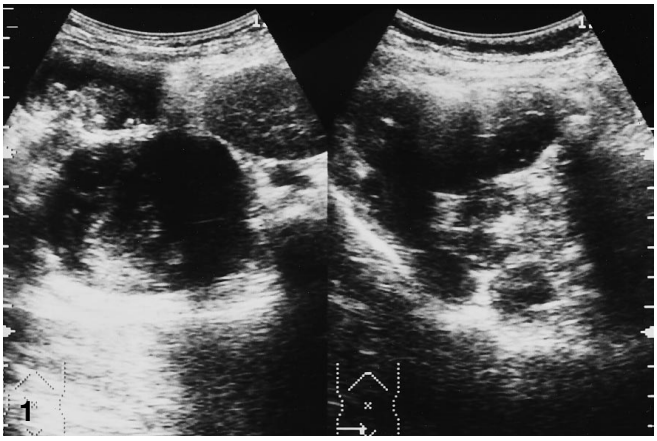
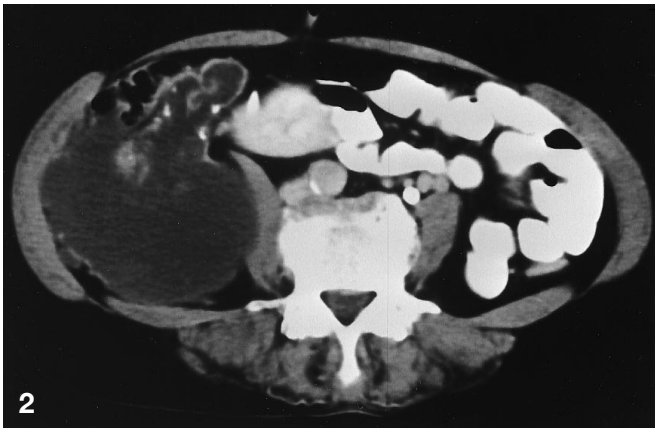


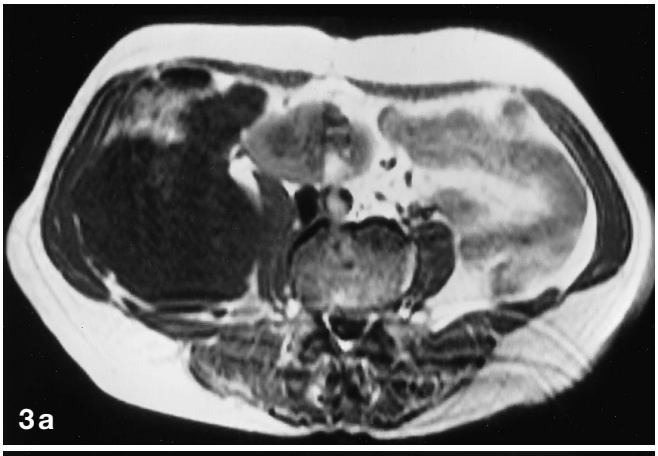


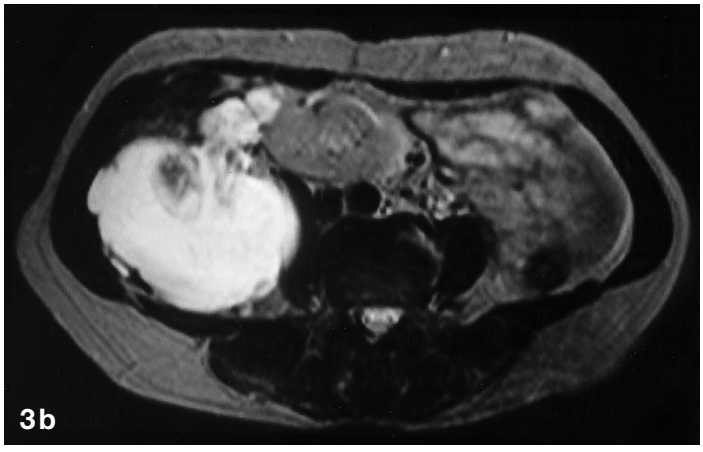
PRP is a very rare condition in which the retroperitoneal space is full of a thick, gelatinous, myxoid material. To our knowledge, 12 cases of PRP have been reported [8–17], comprising seven men and five women aged 39–82 years (mean age 66 years). The sites of origin were diagnosed as or suspected to be the appendix in all cases. PRPs were described as multicystic or multiseptate. The images were similar to that of PP. Calcifications were seen in two cases [12, 15], and air in two cases [12, 17]. The cause of pseudomyxoma spreading from the appendix to the retroperitoneal space may be the position of the retrocecal appendix and its fixation to the posterior abdominal wall [14]. A chemical relationship may exist between the cartilage in growing bone and the chemical composition of mucin, both of which are very similar glycoproteins [18].

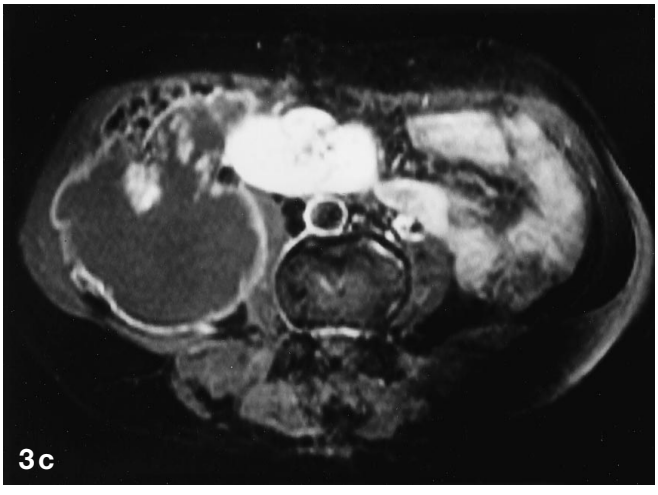
Fig. 1. US scan shows multiple cysts in the right abdomen. The internal echoes in the cysts are due to solid portions or gelatinous material

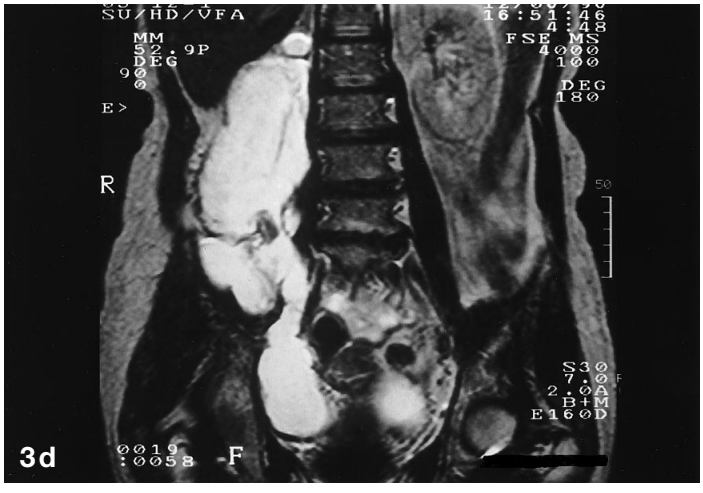
Fig. 2. Contrast-enhanced CT scan shows a multicystic mass with nodes and tiny calcifications in the right retroperitonal space

Fig. 3 a–d. MR images show the retroperitoneal mass. The intensity of the mass is low on T1-weighted imaging (**a**), and high on T2-weighted axial (**b**) and coronal (**d**) images. The capsule, septum, and nodes are enhanced on postcontrast T1-weighted imaging with fat suppression (**c**). The tumor extends to the pelvis (**d**)









Primary retroperitoneal mucinous cystadenoma (RMC) and mucinous cystadenocarcinoma (RMCCa) are also very rare [15, 19–23]. Rothacker et al. [23] reviewed 13 primary RMC and 15 primary RMCCa. These patients were all women, their ages ranging from 17 to 69 years (median 36 years). As far as could be determined, the primary RMC and RMCCa consisted of a unilocular cyst or a central large cyst with some peripheral small cysts [20–23]. The images of these tumors were not similar to that of PP, but rather to ovarian or pancreatic cystic lesions. The histogenesis of primary RMC and RMCCa is not clear, but it has been suggested that these tumors may arise from teratomas, ectopic supernumerary or accessory ovaries, or celomic metaplasia [20]. The derivation from celomic epithelium during gonadal descent is now the most widely accepted theory [23].

Our patient told us that her appendicitis had been so severe that the period of her hospitalization was longer than usual. It was possible that the appendix, a part of the tissue of the appendix, or adenoma of the appendix was adherent to the retroperitoneal space. In this case, it was histologically impossible to determine whether the lesion was primary or secondary. However, the lesion was multicystic, and its images were similar to that of PP, so we diagnosed PRP radiologically.

Among other retroperitoneal cystic lesions are cystic lymphangioma, cystic teratoma, cystic neurinoma, cystic mesothelioma, alimentary tract duplication, hematoma, abscess, and hydatid cyst [15, 24, 25]. Davidson and Hartman [25] reported 19 cases of retroperitoneal lymphangioma. CT showed a unicameral mass in 57% and a septate mass in 43% of these cases. The walls were thin and smooth in 79%, and thick and irregular in 21%. In two cases the mass contained mural calcification. Nobusawa et al. [15] reported that the presence of a 'neck' (between the cystic mass and paraaortic region) strongly suggested lymphangioma. Liposarcoma, leiomyosarcoma, fibrosarcoma, and malignant teratoma – which are common primary malignant tumors of the retroperitoneal space – generally have a solid appearance on CT, but they may be cystic on rare occasions when the mass has become necrotic [24]. The differential diagnoses of a retroperitoneal multicystic mass should include pseudomyxoma retroperitonei.

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