Abdom Imaging 24:78-81 (1999)

Abdominal Imaging

Peritoneal serous papillary carcinoma: radiological appearance

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Received: 12 November 1997/Accepted: 31 December 1997

Abstract

Background: The radiological appearance of peritoneal serous papillary carcinoma (PSPC) is described.

Methods: Three cases of PSPC were analyzed retrospectively with regard to the radiological appearance and histopathological features.

Results: All three patients were women, aged 44–71 years. Massive ascites and a greater omentum tumor were observed on computed tomography in all patients. Double-contrast enema performed in one patient showed irregularity on the upper aspect of the transverse colon. Radiological examinations excluded primary tumors in both gastrointestinal and genital organs in all patients. Histological diagnosis was made from the surgical specimen in two patients and from an autopsy specimen in one patient. All patients had a large omental tumor involving the transverse colon, but the ovaries were not involved or only minimally involved on the surface. Serum CA125 was markedly elevated, and immunohistochemical staining for CA125 was positive within the tumor cell cytoplasm in all three patients.

Conclusion: PSPC cannot be diagnosed from radiological findings alone because of its similarity to metastatic peritoneal carcinomatosis and peritoneal mesothelioma. Marked elevation of serum CA125 may help with PSPC diagnosis. Response to treatment is promising, and exploratory laparotomy is thus justified when a patient shows characteristic radiological findings and high CA125 level.

Key words: Peritoneum—Neoplasm—Computed tomography—Barium enema—CA125.

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Since 1959, when Swerdlow reported the first case of peritoneal serous papillary carcinoma (PSPC) as a ''mesothelioma resembling papillary ovarian adenocarcinoma'' [1], PSPC has become recognized as an independent pathological entity. The etiology of PSPC is uncertain but it is believed to be a primary peritoneal tumor arising from the secondary Müllerian system in the mesothelium. The histopathological features of this tumor are identical to those of ovarian papillary serous carcinoma (OPSC), but in PSPC the ovaries are intact or only their surface is affected [2]. The clinical features of PSPC are also similar to those of peritoneal carcinomatosis due to OPSC and the response to using cisplatin-based chemotherapy regimen is favorable. Thus, a correct diagnosis of PSPC, distinguishing it from the other peritoneal malignancies with a poor prognosis, is required before treatment. There are few descriptions of the radiological appearance of PSPC in the literature; therefore, we analyzed the radiological, clinical, and histopathological features of this carcinoma in our institution.

Case reports

Case 1

A 59-year-old woman presented with rapidly progressing abdominal distention and loss of appetite. She had a history of chronic thyroiditis for 25 years, but the thyroid function had normalized during that period. Physical examination showed diffuse goiter, massive ascites, and palpable induration in the lower abdomen. Cytological examination of ascitic fluid showed adenocarcinoma cells, which did not stain with periodic acid–Schiff with diastase digestion. Serum carbohydrate antigen 12-5 (CA125) was markedly elevated to 11,100 IU/mL (normal < 35 IU/mL). Computed tomography (CT) demonstrated a poorly demarcated smudgy tumor in front of the transverse colon (Fig. 1A), but no involvement of the genital organs was observed. Double-contrast enema showed irregularity on the upper aspect of the transverse

colon (Fig. 1B). This finding suggests colon involvement through the gastrocolic ligament, which is typically seen in an advanced gastric cancer [3], but no abnormality was detected in the stomach on endoscopic and double-contrast examination. Because there was no evidence of a primary site in the gastrointestinal system, an ovarian tumor was suspected. After one course of chemotherapy using cisplatin, bilateral adnexectomy was performed and a peritoneal infusion catheter was implanted. At surgery, a large omental tumor involving the transverse colon was found, and multiple nodules were seen to be scattered on the peritoneum and on the surface of the ovaries. Histopathological examination of surgical specimens of the tumor demonstrated a tubulopapillary structure of the tumor cells with considerable nuclear atypism and psammoma bodies (Fig. 1C). On immunohistochemical staining, the tumor cells were found to be positive for CA125. As a result, PSPC was diagnosed. The patient received chemotherapy postoperatively, by both intravenous and intraperitoneal infusion, with a cisplatin-based regimen. The intraperitoneal tumors then disappeared, and she has been free of disease for 4 years.

Case 2

A 44-year-old woman visited our clinic having suffered from abdominal distention for 1 month. Physical examination showed a large amount of ascites, and cytological examination of ascitic fluid showed adenocarcinoma cells. On ultrasonography, there was a vague tumor echo in the peritoneal space surrounded by massive ascites. CT also showed ascites and an omental tumor but no other abnormality (Fig. 2). Because serum CA125 was elevated to 3,598 IU/mL, an ovarian tumor could not be excluded. Exploratory laparotomy was then performed. At surgery, there was a huge omental tumor involving the colon, but the ovaries could not be observed, being obscured by diffuse adhesive carcinomatosis. Despite chemotherapy, the patient died 110 days after surgery and autopsy was performed. Macroscopically, there was minimal invasion of the ovarian cortex and of the fallopian tubes bilaterally. Light microscopic examination of the tumor cells demonstrated a tubulopapillary structure, and immunohistochemical staining showed CA125-positive cells. These macroscopic and microscopic findings were thus consistent with a diagnosis of PSPC.

Case 3

A 71-year-old woman was admitted to our hospital with rapidly progressive abdominal distention. Massive ascites was found on physical examination, and adenocarcinoma cells were detected in the ascitic fluid. Serum CA125 was extremely elevated to 10,329 IU/mL. A gynecological survey, including ultrasonography, did not show any neoplastic lesion in the genital organs. One week after admission, induration was felt on palpation in the left lower abdomen. Abdominal CT showed a poorly defined hazy mass in the anterior portion of the peritoneal cavity, together with retroperitoneal lymphadenopathy and ascites (Fig. 3). Althogh no definitive diagnosis was obtained, intraperitoneal infusion of cisplatin was started. The ascites was then diminished, and the serum CA125 level fell to 7124 IU/mL.

Sixty-six days after admission, the patient complained of sudden abdominal pain. On abdominal radiography, free air was observed intraperitoneally. At surgery, the transverse colon was obstructed by an omental tumor, and perforation at the cecum was observed. Multiple nodules were found on the peritoneum, but a primary tumor of the genital organs was excluded macroscopically. The affected transverse colon was resected, but its mucosa was free of disease. Microscopically, no neoplastic lesion was found in the biopsied ovaries and a histological survey of the tumor demonstrated papillary serous carcinoma with CA125-positive cells. The diagnosis, based on surgical and pathological findings, was PSPC.

Discussion

Neoplasms of the female peritoneum are classified into three groups: mesothelial, Müllerian, and metastatic [4]. Embryologically, the germinal epithelium of the ovary and the mesothelium of the peritoneal cavity derive from the same coelomic epithelium [5]. PSPC belongs to Müllerian tumors, arising from the mesothelium and with a capacity to differentiate the second Müllerian system, located external to the cavities of the original Müllerian ducts. The histopathological features of PSPC are identical to those of OSPC, but in PSPC the ovaries are intact or only the surface of the ovaries is affected. The biological behavior of PSPC is also similar to that of OSPC. Three cases of PSPC with long-term survival have been reported by Chen and Flam [6], and our patient 1 has survived for 4 years. Strnad et al. reported five patients with complete response to cisplatin-based chemotherapy and recommended therapeutic guidelines for advanced ovarian carcinoma, including initial surgical cytoreduction followed by cisplatin-based combination chemotherapy [7]. In view of good response, it is crucial to recognize PSPC and differentiate it from other peritoneal tumors with poor a prognosis, such as peritoneal carcinomatosis originating from sources other than ovarian cancer or malignant mesothelioma.

The clinical and pathological features of PSPC have been clarified, but there are few descriptions of its imaging features in the literature. In our cases, three characteristic CT findings were noted: a large amount of ascitic fluid, a greater omentum tumor, and exclusion of primary tumor in both the gastrointestinal system and the genital organs. However, these findings do not seem to be specific to PSPC. Walkey et al. described CT findings in patients with peritoneal carcinomatosis; the most common feature was ascites, and the most common origin of the carcinomatosis was ovarian [8]. They found that the detectability of the primary site was only about 40–50%. In addition, the greater omentum was the favored site for peritoneal seeding. The primary sites in their patients with omental caking differed considerably and included the ovary, fallopian tube, colon, stomach, and peritoneum.

Peritoneal mesothelioma is another type of primary peritoneal tumor. It has some histopathological variants, and Fox classified it into four groups: cystic mesothelioma, well-differentiated papillary mesothelioma, fibrous mesothelioma, and diffuse mesothelioma [4]. The CT findings in mesothelioma differ greatly and include peritoneal thickening, nodular apperance of the peritoneum, omental caking, different amount of ascites, and so on [9–11]. Occasionally, the CT appearance of peritoneal mesothelioma is similar to that of our PSPC, making it difficult to differentiate these two tumors.

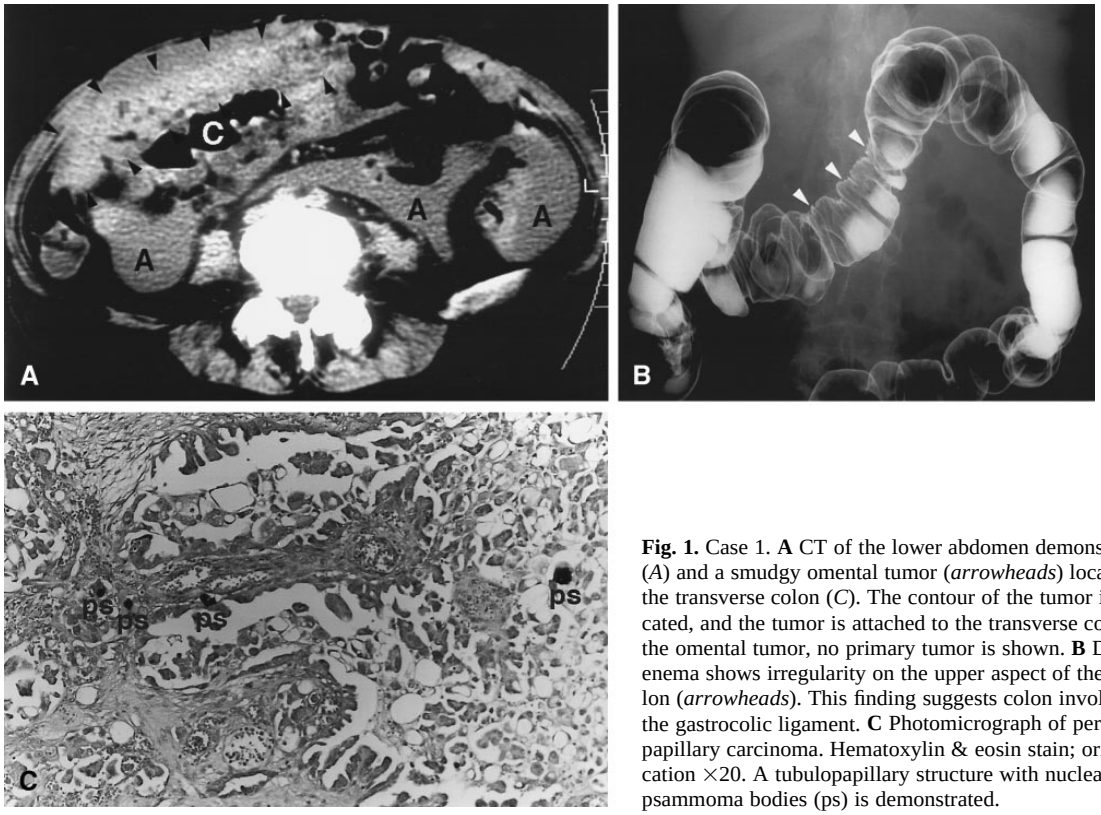

Fig. 1. Case 1. **A** CT of the lower abdomen demonstrates ascites (*A*) and a smudgy omental tumor (*arrowheads*) located in front of the transverse colon (*C*). The contour of the tumor is poorly demarcated, and the tumor is attached to the transverse colon. Except for the omental tumor, no primary tumor is shown. **B** Double-contrast enema shows irregularity on the upper aspect of the transverse colon (*arrowheads*). This finding suggests colon involvement through the gastrocolic ligament. **C** Photomicrograph of peritoneal serous papillary carcinoma. Hematoxylin & eosin stain; original magnification ×20. A tubulopapillary structure with nuclear atypism and psammoma bodies (ps) is demonstrated.

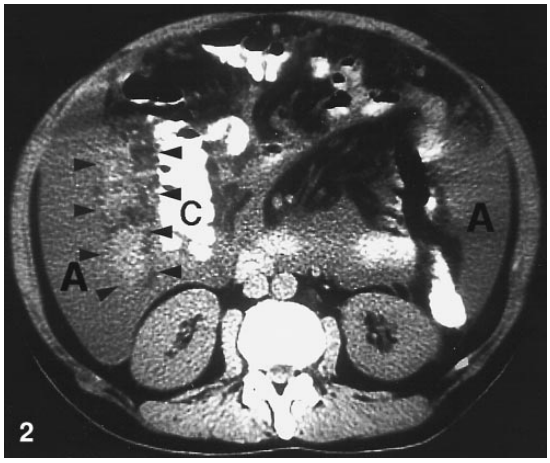

Fig. 2. Case 2. CT at the level of kidney shows a poorly demarcated tumor (*arrowheads*) surrounded by fat density adjacent to the colon (*C*). No primary site in another location is detected. The peritoneal cavity is occupied by ascites (*A*).

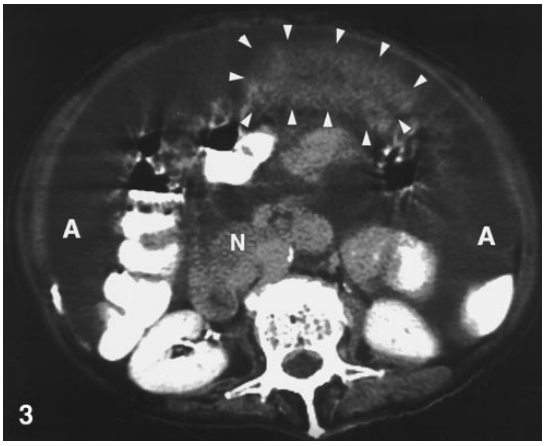

Fig. 3. Case 3. CT at the level of the kidney with contrast administration. A poorly defined hazy mass (*arrowheads*) is located in the anterior portion of the peritoneal cavity, in which massive ascites (*A*) is collected. Retroperitoneal lymphadenopathy is also demonstrated (*N*), but the gastrointestinal system and genital organs appear free from disease.

Cooper et al. analyzed the radiographic appearances of benign and metasatic malignant omental disease [12]. They identified four distinct patterns: omental caking, finely infiltrated fat with a ''smudged appearance,'' cystic masses, and discrete nodules. The CT appearances of the omental lesion in our cases differed somewhat; case 1 showed a ''smudged appearance'' and cases 2 and 3 showed omental caking. Because both patterns are common in metastatic disease, it is difficult to differentiate PSPC from metastatic peritoneal carcinomatosis.

In case 1, double-contrast enema demonstrated involvement of the superior part of the transverse colon.

This finding implies invasion in the area between the taenia omentalis and the taenia mesocolica [3]. This area easily becomes involved by extension of malignancy through the gastrocolic ligament, as is commonly seen in cases of gastric cancer. However, if the greater omentum is attached to the transverse colon at the taenia omentalis, primary omental disease could also extend to the same portion of the colon. The radiological findings on CT and double-contrast enema in our PSPC patients were indistinguishable from those in metastatic peritoneal carcinomatosis or peritoneal mesothelioma; therefore, we conclude that PSPC cannot be diagnosed on radiological grounds alone.

CA125 is a useful marker for epithelial ovarian cancer. In our cases, the serum CA125 levels were extremely elevated, to values ranging between 3598 and more than 10,000 IU/mL. A case of PSPC with a high CA125 level has been reported by Rosen et al. [13]. Bast et al. measured CA125 levels in patients with epithelial ovarian carcinoma and in different control groups [14] and found a value of more than 2000 IU/mL only in epithelial ovarian carcinoma. If a patient with peritoneal carcinomatosis had such a high level of CA125, epithelial ovarian malignancy was usually suspected. Therefore, in combination with exclusion of ovarian malignancy on radiological examination, findings of an extremely high level of CA125 and characteristic CT features such as ascites and omental tumor give strong reasons to suspect PSPC, and exploratory laparotomy should be undertaken for a definitive diagnosis.

Because the biological behavior of PSPC is supposed to be similar to that of OSPC, multidisciplinary management with cell reduction surgery and chemotherapy, which are applied for stage III and IV ovarian carcinoma, would appear to be the optimal treatment of PSPC. In case 1, combination chemotherapy using intravenous and intraperitoneal infusion of anticancer drugs was used in addition to surgery. This treatment has been effective for 4 years. In cases 2 and 3, the prognoses were poor; intensive chemotherapy was not possible because of poor general condition. On the assumption that the therapeutic strategy in PSPC should be same as that in advanced ovarian cancer, an aggressive approach to PSPC is desirable.

With the recent advances in cancer treatment, the prognosis of PSPC has been improved. Radiological examinations, including CT, and serum CA125 measurement are recommended in patients with suspected PSPC to arrive at a precise diagnosis.

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