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Abdominal Imaging

Synchronous mucinous tumors of the ovary and the appendix associated with pseudomyxoma peritonei: CT findings

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Abstract
Background: To present the computed tomographic (CT) findings of synchronous mucinous tumors of the ovary and the appendix associated with pseudomyxoma peritonei (PMP).
Methods: Imaging studies, mainly abdominal CT scans, of three women aged 49–75 years were reviewed. Attention was directed to the ovarian masses, peritoneal seeding, and the presence of an appendiceal mucocele.
Results: The ovarian tumors and the appendiceal mucocele were clearly demonstrated in two cases, and they were part of the extensive PMP in the third patient. Ascites was found in all cases, with internal septation in one. Associated scalloping of the liver margins and hypodense peritoneal implants, with extensive bowel involvement, were seen in another one. Pathologically, there was one case of right ovarian mucinous cystadenoma and villous adenoma of the appendix, one case of right ovarian and appendiceal mucinous cystadenocarcinoma, and one case of bilateral metastatic ovarian implants of appendiceal mucinous cystadenocarcinoma. PMP was found in all. In the case with benign tumors of the ovary and the appendix, the PMP was classified as a benign mucinous spillage. This patient returned 33 months after surgery with PMP, in which epithelial cells were found.
Conclusions: Radiologists should be familiar with the clinical occurrence of synchronous mucinous tumors of the ovary and the appendix associated with PMP and with the typical CT findings of the latter two entities. Alternatively, when the imaging findings suggest ovarian cystic tumor with PMP, the radiologist should be alerted to the probability of a clinically unsuspected appendiceal mucocele and should search for it.

Key words: Appendix, neoplasm—Ovary, neoplasm—Pseudomyxoma peritonei—Computed tomography.

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Ovarian cystadenocarcinomas are responsible for more than 50% of pseudomyxoma peritonei (PMP) [1, 2]. The concurrent occurrence of a cystic tumor in the appendix in these cases has been well established in large clinicopathological series [3–7]. The ovarian and appendiceal tumors are usually synchronous, but one may appear years before the other. The causal link between these tumors is uncertain as to whether they have independent origins or the ovarian tumor represents a secondary deposit from the appendiceal neoplasm [8, 9]. Seidman et al. claimed that this situation represents a "multifocal neoplasia of the ovary, appendix and the peritoneum" [5].

We present the computed tomographic (CT) findings of three women with ovarian cystic tumors and coexisting appendiceal mucocele associated with PMP, in whom the diagnosis was suspected preoperatively.

Material and methods

The clinical data and imaging studies from three women, aged 49–75 years, were retrospectively reviewed. They were diagnosed during a

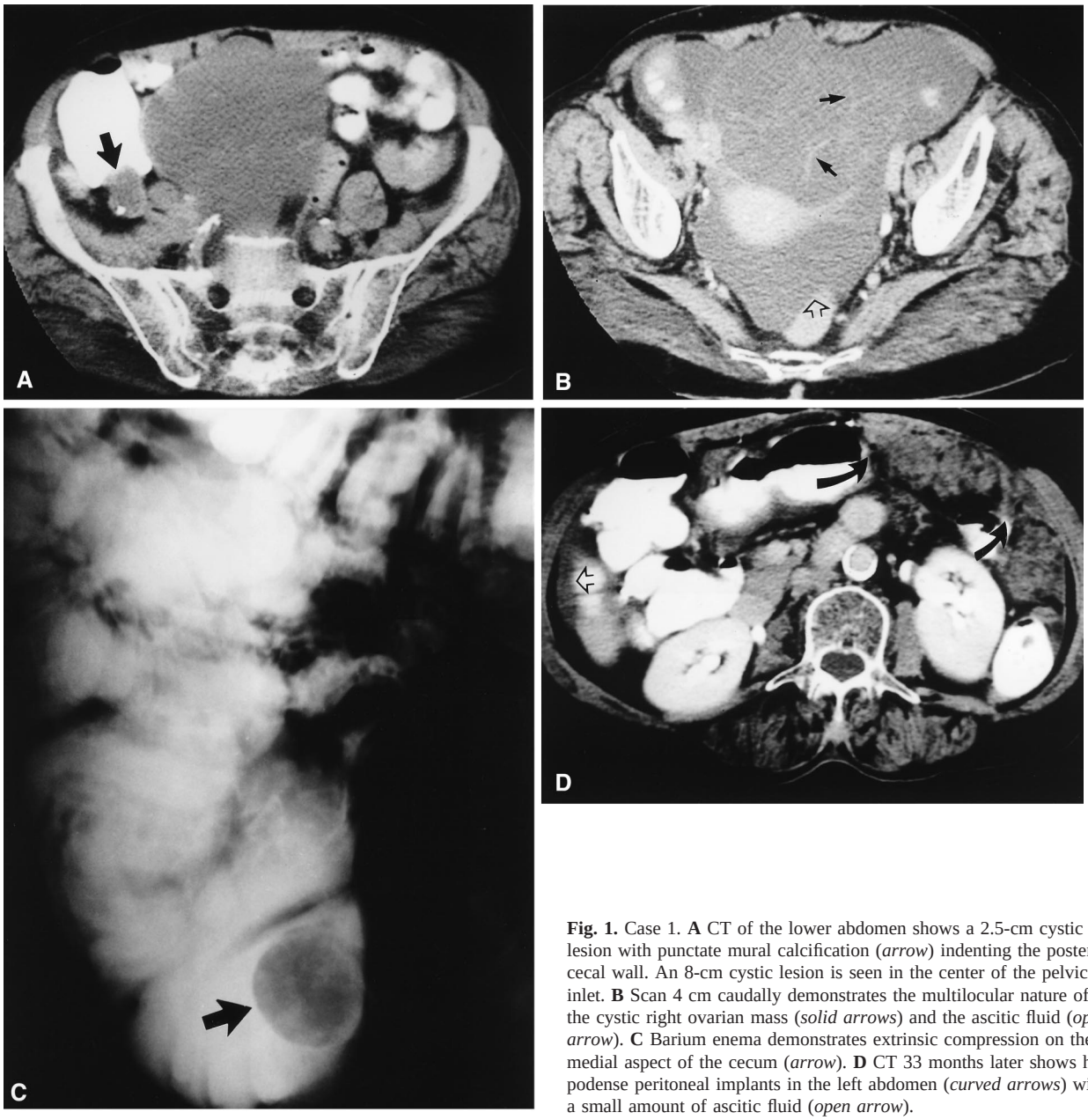

Fig. 1. Case 1. **A** CT of the lower abdomen shows a 2.5-cm cystic lesion with punctate mural calcification (*arrow*) indenting the posterior cecal wall. An 8-cm cystic lesion is seen in the center of the pelvic inlet. **B** Scan 4 cm caudally demonstrates the multilocular nature of the cystic right ovarian mass (*solid arrows*) and the ascitic fluid (*open arrow*). **C** Barium enema demonstrates extrinsic compression on the medial aspect of the cecum (*arrow*). **D** CT 33 months later shows hypodense peritoneal implants in the left abdomen (*curved arrows*) with a small amount of ascitic fluid (*open arrow*).

5-year period as having cystic ovarian tumors associated with an appendiceal mucoceles and PMP. Two of these patients were described previously, with an emphasis on the CT findings of mucocele of the appendix [10].

CT was the initial examination to suggest the diagnosis of appendiceal cystic tumors or PMP in all patients. There were four abdominal CT scans because one patient had a second CT 33 months after surgery. In addition, a barium enema was performed in two patients. All patients underwent surgery.

CT was performed on either an Elscint 2400 Elite or an Elscint CT Twin with 10-mm collimation and 1.0-cm interval from the diaphragm to the symphysis pubis. Oral contrast material, 1000 mL of a flavored solution with 4% Telebrix (meglumine-ioxitalamate), was administered 2 h before the examination and an additional 250 mL was administered just before the study. Intravenous contrast medium (80–100 cc Telebrix) was manually injected by bolus in all patients.

Case reports

Case 1

A 71-year-old woman was admitted with a pelvic mass detected during a routine physical examination. Contrast-enhanced CT showed a large right ovarian multilocular cyst measuring 8 cm, a 2.5-cm cystic lesion with punctate calcification indenting the cecal wall, and ascites in the pelvis and left gutter (Fig. 1A,B). Barium enema showed extrinsic compression on the medial aspect of the cecum with an intact mucosa (Fig. 1C). At surgery, gelatinous ascites, right ovarian mass, and an appendiceal mucocele were found. Hysterectomy, bilateral salpingooophorectomy, and right hemicolectomy were performed. The pathologic examination of the surgical specimen demonstrated PMP with scattered cellularity, right ovarian cystadenoma, and villous adenoma of

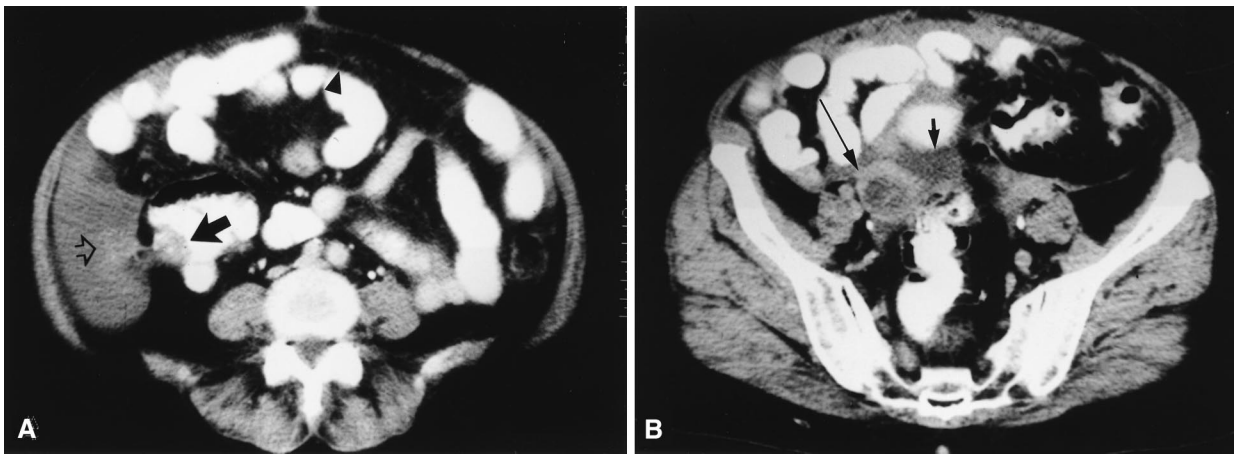

Fig. 2. Case 2. **A** Contrast-enhanced CT shows a 2-cm cystic lesion with punctate mural calcifications indenting the lateral cecal wall (*solid arrow*). Ascitic fluid with fine internal septations (*open arrow*) is seen in the right gutter, as is a delicate omental infiltration (*arrowhead*). **B** In the pelvis, a 3-cm right ovarian cystic lesion with minimal mural thickening and internal septation is seen (*long arrow*). A small amount of free peritoneal fluid is seen (*short arrow*).

the appendix. Thirty-three months later, she returned with progressive abdominal distention and vague abdominal pain. A repeat CT scan disclosed low-density peritoneal implants and a moderate amount of ascites (Fig. 1D). Malignant PMP was diagnosed by needle tap of the ascitic fluid.

Case 2

An asymptomatic 75-year-old woman with a history of breast cancer was found to have a cystic ovarian lesion detected by a transvaginal sonogram. Contrast-enhanced CT showed a 2-cm right ovarian septated cyst, a 3-cm cystic lesion with punctate calcification indenting the cecal wall, and ascites in the pelvis and right gutter (Fig. 2A,B). There were internal septations within the ascitic fluid and minimal infiltration of the omentum (Fig. 2A). At surgery, gelatinous ascites, a right ovarian cyst, and mucocele of the appendix were found. Hysterectomy, bilateral salpingooophorectomy, and right hemicolectomy were performed. The pathologic diagnosis was ovarian and appendiceal mucinous cystadenocarcinoma and PMP.

Case 3

A 49-year-old woman presented with rapidly progressing abdominal distention, abdominal pain, and nausea. On physical examination, an abdominopelvic mass was found. On ultrasonography, a 12- × 5- × 7-cm complex, mainly solid, pelvic mass was found with a small amount of peritoneal fluid. Double-contrast barium enema showed an arcuate compression on the medial aspect of the cecum, separating adjacent bowel loops, indicating a mass (Fig. 3A). It was suspected to represent an appendiceal mucocele. Contrast-enhanced CT showed a large pelvic mass embedding the internal genitalia and the sigmoid colon (Fig. 3B,C). In addition, multiple hypodense peritoneal implants were seen between contrast-filled bowel loops (Fig. 3D) and in the ligamentum teres (Fig. 3E). A small amount of pelvic fluid was also seen. The presumed preoperative diagnosis was PMP associated with appendiceal and ovarian tumors.

At surgery, massive gelatinous peritoneal implants were found surrounding the entire internal genitalia, sigmoid colon, terminal ileum, and cecum. Debulking of the peritoneal masses, hysterectomy with bilateral salpingooophorectomy, right hemicolectomy, and sigmoid resection with ileotransverse anastomosis and Hartman's procedure were performed. The pathologic diagnosis was metastatic mucinous cystadenocarcinoma involving both ovaries and the sigmoid colon, Duke's C2 mucinous adenocarcinoma of the cecal region, and PMP. The adjacent wall of the right colon was also involved by the tumor, and the right hemicolectomy specimen showed an almost completely destroyed cecum without an identifiable appendix. The primary tumor was diagnosed as consistent with an appendiceal tumor. A 4.5-cm uterine leiomyoma was found.

Discussion

PMP is an unusual disease found in two of 10,000 laparotomies. Three of four patients affected by PMP are women [2, 11]. In the case of PMP associated with mucinous ovarian tumor, the neoplasm is frequently benign or of low malignant potential (borderline) [4, 6, 9, 11]. In most reported cases, there is a coexisting adenoma or carcinoma of the appendix, which typically cause its macroscopic dilatation, termed *mucocele* [3]. There are several publications concerning the coexistence of mucinous tumors of the ovary (mainly right sided or bilaterally) and the appendix associated with PMP [3–7].

The simultaneous occurrence of neoplastic diseases in female patients might be explained on the basis of two separate neoplasms or by spread from one organ to another [8, 9]. Ronnett et al. reported the largest series, of 30 women with multifocal peritoneal mucinous implants with ovarian mucinous tumors and coincident appendiceal or colonic tumors. No case was of unequivocal ovarian origin, and in 28 cases the ovarian tumors were considered as secondary involvement. They concluded

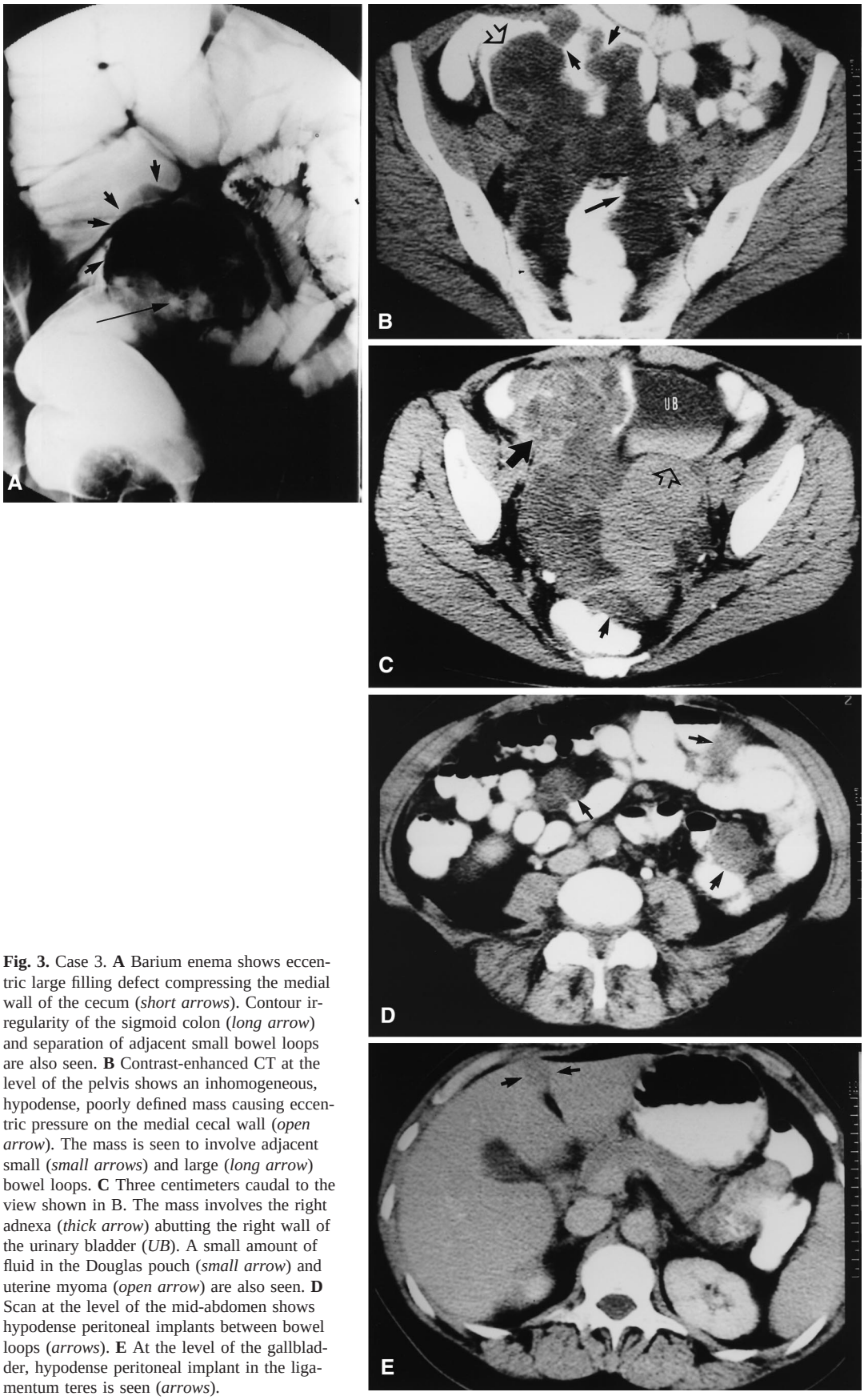

Fig. 3. Case 3. **A** Barium enema shows eccentric large filling defect compressing the medial wall of the cecum (*short arrows*). Contour irregularity of the sigmoid colon (*long arrow*) and separation of adjacent small bowel loops are also seen. **B** Contrast-enhanced CT at the level of the pelvis shows an inhomogeneous, hypodense, poorly defined mass causing eccentric pressure on the medial cecal wall (*open arrow*). The mass is seen to involve adjacent small (*small arrows*) and large (*long arrow*) bowel loops. **C** Three centimeters caudal to the view shown in B. The mass involves the right adnexa (*thick arrow*) abutting the right wall of the urinary bladder (*UB*). A small amount of fluid in the Douglas pouch (*small arrow*) and uterine myoma (*open arrow*) are also seen. **D** Scan at the level of the mid-abdomen shows hypodense peritoneal implants between bowel loops (*arrows*). **E** At the level of the gallbladder, hypodense peritoneal implant in the ligamentum teres is seen (*arrows*).

that the ovarian mucinous tumor is rarely, if ever, the primary site of the condition of PMP [6].

Ovarian cancer is the second most common gynecological cancer and the leading cause of death due to malignant tumor in the reproductive tract in women. The diagnosis and staging by CT is not universally accepted, and its use in the preoperative evaluation of ovarian masses is controversial and uncertain [12]. CT of the abdomen is, however, an important diagnostic tool in the diagnosis and evaluation of coexistent pathologic processes in patients with ovarian mucinous tumors bcause appendiceal cystic tumors and PMP may have typical imaging findings [10, 13, 14]. Preoperative imaging and diagnosis are desirable to plan the preferable treatment [11]. Imaging is also a great aid in the assessment of recurrent disease.

The typical CT finding of an appendiceal mucocele is a cystic, well-encapsulated mass, sometimes with mural calcification, in the expected location of the appendix, causing extrinsic pressure on the cecal wall without any surrounding inflammatory reaction. Most cases of mucocele should be regarded as a mucinous neoplasm, either benign or malignant [10, 15, 16]. In two of our patients, the mucocele was spherical with mural calcification. In the third patient, who had no history of prior appendectomy, the appendix could not be identified by imaging or in the operative specimen. It was embedded within the tumoral mass that had grown from it.

Pathologically, PMP is defined as large pools of peritoneal mucin that contain epithlial glandular cells. This is in contrast to a self-limited, benign process associated with spillage of acellular mucin, around the appendix associated with a ruptured appendiceal adenoma [6, 9].

The diagnostic CT criteria for PMP are scalloping of the liver and splenic margins, septated ascites or ascites with attenuation slightly higher than water, and hypodense peritoneal implants that may cause extrinsic pressure on bowel loops [10, 11, 13, 14]. These implants occasionally show curvilinear or punctate calcifications. We observed at least one of these findings in two of our cases and established the correct diagnosis preoperatively. These patients were found to have PMP at the time of surgery. Absence of such imaging findings, however, does not exclude PMP, as happened in our first case. That patient had CT findings of a right ovarian cystic tumor, an appendiceal mucocele, and a mild amount of ascitic fluid in the pelvis and left gutter. On pathology, benign tumors were found, and the fluid was classified as a "benign" form of PMP, resulting from spillage of mucin into the peritoneal cavity. About 3 years later, however, PMP had been proven by needle biopsy, associated with CT findings compatible with that diagnosis.

Ronnett et al. also classified two categories with different pathologic features and prognosis of PMP, which emphasizes its complexity [17]. They defined peritoneal mucinous carcinomatosis as peritoneal mucinous implants, which contain epithelial tumor cells, arising from a gastrointestinal cystadenocarcinoma. Thirty percent of those cases had ovarian involvement, 70% bilaterally, with histology similar to that of the primary and peritoneal lesions. The other category, disseminated peritoneal adenomucinosis, was defined as benign peritoneal implants. Associated ovarian stromal involvement, similar to that of the peritoneal tumor, was found in 23% of the cases, usually bilaterally.

In clinical series, however, the diagnosis of PMP is based on the gross intraoperative finding of an abdominal cavity filled with gelatinous or mucinous material regardless of its cellularity [3–5, 7]. These collected series of PMP represent a heterogeneous group of the two described pathologic entities. We assume that acellular PMP may not show the "classic" CT findings reported in the "true" PMP. These findings probably result from the islands of tumor cells found within the intraperitoneal mucus deposits. These two pathologic processes did show different imaging findings in our first patient, but this thesis has to be evaluated further in radiologic–pathologic analysis.

In conclusion, radiologists should be familiar with the rare occurrence of synchronous mucinous tumors of the ovary and the appendix associated with PMP. Findings of an appendiceal mucocele, a cystic ovarian mass, mainly right sided or bilaterally, and ascitic fluid should arouse the suspicion of the diagnosis, especially if additional suggestive imaging findings of PMP are present.

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