SUNTEXT REVIEWS

SunText Review of Case Reports & Images
ISSN: 2766-4589

Open Access
Case Report
Volume 4:7

An Aggressive Pseudomyxoma Peritonei Secondary to an Appendiceal Mucocele: A Case Report and Review of Literature

Choayb S*, El Harras Y, Messaoud O, Jerguigue H, Latib R and Omor Y

Department of radiology, National Institute of Oncology, Mohammed V University, Rabat, Morocco

***Corresponding author:** Choayb S, Department of radiology, National Institute of Oncology, Mohammed V University, Rabat, Morocco; E-mail: choayb.safaa@gmail.com

Received date: 04 September 2023; **Accepted date:** 07 September 2023; **Published date:** 12 September 2023

Citation: Choayb S, El Harras Y, Messaoud O, Jerguigue H, Latib R, Omor Y (2023) An Aggressive Pseudomyxoma Peritonei Secondary to an Appendiceal Mucocele: A Case Report and Review of Literature. SunText Rev Case Rep Image 4(7): 206.

DOI: https://doi.org/10.51737/2766-4589.2023.106

Abstract

Pseudomyxoma peritonei (PMP) is an unusual condition responsible for mucinous ascites in the peritoneal cavity. The most common cause is a mucinous neoplasm of the appendix. We present the case of a 67-year-old postmenopausal patient who presented to the department of gynecology with a 6-month history of abdominal pain and distension. Her abdominal ultrasound revealed a large amount of fluid within the peritoneal cavity and bilateral cystic pelvic masses. An MRI showed an appendiceal mucocele associated with bilateral cystic ovarian masses and a large amount of ascites.

Keywords: Pseudomyxoma peritonei; Mucinous neoplasms; Appendix; Ovary

Introduction

PMP, also referred to as “jelly belly”, is characterized by the presence of mucinous effusion in the peritoneal cavity. It is a rare condition, frequently secondary to mucinous neoplasms of the appendix [1]. Occasionally, PMP may emerge from other abdominal neoplasms, including those of the stomach, pancreas, gallbladder, urachus, urinary bladder, or pelvic organs, in which the ovary is a rare cause [2,3]. PMP should be considered a borderline malignant process, as this disease has a wide spectrum from slow growth to rapidly infiltrative progressive disease [3]. Imaging plays an important role in preoperative diagnosis and guides the therapeutic approach.

Case Report

A 67-year-old postmenopausal patient presented to the department of gynecology with a 6-month history of abdominal pain and distension. She previously had an abdominal ultrasound, which revealed a large amount of fluid within the peritoneal cavity and bilateral cystic pelvic masses. The patient didn’t have a family history of cancer or toxic habits. Physical examination revealed a distended abdomen; no palpable mass was detected. Laboratory tests revealed elevated tumour markers: carcinoembryonic antigen (CEA) (236 ng/mL), carbohydrate antigen 19.9 (CA 19.9) (<2 IU/mL), and CA 125 (36 ng/mL). The upper endoscopy and colonoscopy were negative for pathology. A CT scan showed a large amount of ascites with scalloping on the liver surface associated with nodular peritoneal thickening. The ovarian masses were cystic and had bilateral curvilinear calcifications. No lymph node enlargement was noted. The appendix was difficult to assess due to the large amount of ascites (Figures 1,2). An abdominal MRI showed an appendiceal mucocele, bilateral cystic ovarian masses, a large amount of ascites, and nodular peritoneal thickening (Figures 3,4). The patient underwent an exploratory laparoscopy that was converted to a laparotomy due to an important amount of yellowish, gelatinous ascites with adhesions. At exploration, carcinosis completely engulfed the right diaphragmatic cupule, porta hepatis, lesser omentum, stomach, and spleen. Several implants were also noted in the lumbar and iliac regions, in addition to bilateral ovarian masses. The small bowel was massively involved. The PCI (peritoneal carcinosis index) was 36. A biopsy of the large omentum, mesenterium, and abdominal wall and

aspiration of 6 l of ascites were performed. They revealed a low-grade mucinous peritoneal carcinoma. Both histopathology and immunoassays (CK20+ and CK-) confirmed the diagnosis of disseminated peritoneal mucinous carcinomatosis of appendiceal origin. As the surgery was impossible, the patient was referred to the palliative care department. In this case, we couldn't confirm the origin of the ovarian masses based on histopatholgy, and they were considered secondary spread from the appendiceal mucinous neoplasm.

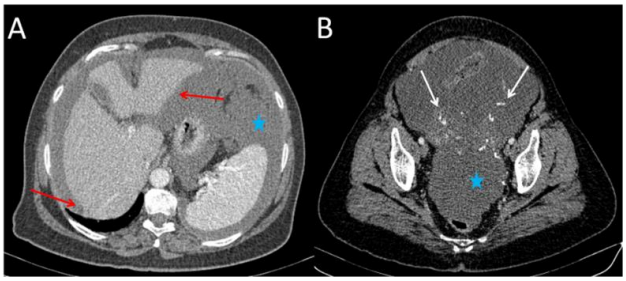

Figure 1: *A and B: axial CT scans of the abdomen revealing diffuse ascites (stars) with scalloping on the liver surface (red arrows). Note the calcifications within the pelvic masses.*

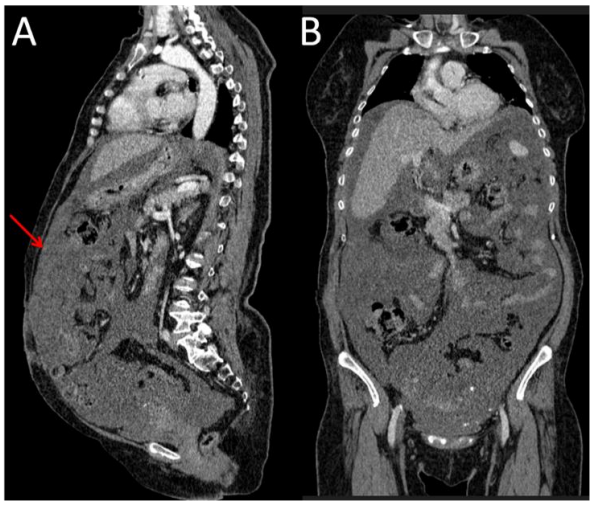

Figure 2: *A and B, thoraco-abdominal sagittal and coronal CT scans show an important amount of ascites within the intraperioneal spaces, mesentery, and omentum. Note the omental caking on the sagittal view (red arrow).*

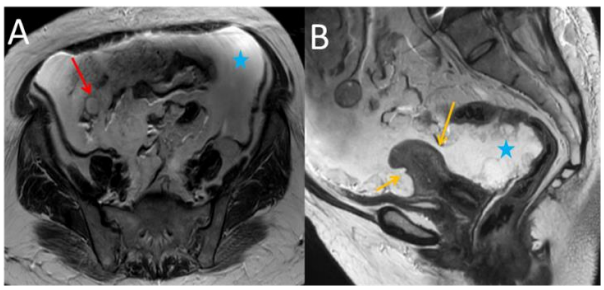

Figure 3: *Axial (A) and sagittal (B) T2WI of the pelvis showing abundant ascites (stars) and a high-signal spherical mass contiguous to the cecum, corresponding to an appendiceal mucocele (red arrow). Note the scalloping of the uterine surface (yellow arrows) by the ascites.*

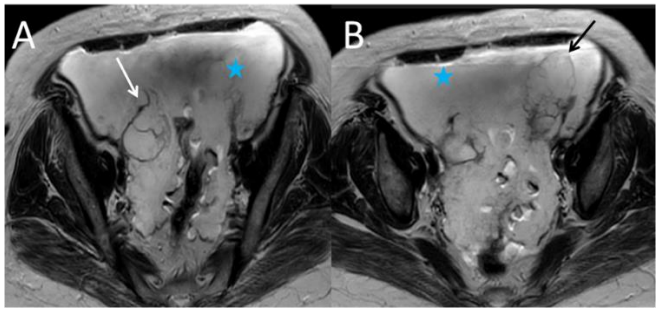

Figure 4: *Axial (A and B) T2WI of the pelvis showing abundant ascites (stars) and bilateral cystic masses of the ovaries (Right ovary: white arrow; Left ovary: black arrow).*

Discussion

In 1842, Carl Rokitansky was the first to describe an appendiceal mucocele, and in 1937, a German gynecologist called Robert Michaelis Von Olshausen suggested that after appendiceal cyst rupture, epithelial cells took roots in the peritoneal cavity and continued the secretion of gelatinous material, leading to PMP [3]. The term PMP was first described by Werth in 1884 in a patient who was supposed to have a ruptured pseudomucinous cystadenoma of the ovary. The term pseudomyxoma derives from a type of mucin called pseudomucin that was used to portray the content of the locules present in ovarian pseudomucinous cystadenoma [4]. It also refers to mucus-free production in the peritoneal cavity or cystic gelatinous masses [4]. According to Carr et al., PMP could include mucinous ascites, omental cake, peritoneal implants, and ovarian involvement. It describes the macroscopic appearance of the gelatinous ascites and thus is not a histological diagnosis [1]. The incidence of PMP is about 1 to 4 individuals per million per year, with a net female predominance [1]. A recent study by Smeenk et al. found that the incidence of mucinous epithelial neoplasm of the appendix is estimated to be around 0,3% and 20% of these patients progress to PMP [3]. In the initial stages, PMP is frequently asymptomatic; the diagnosis is often made during surgery for suspected appendicitis, gynecological cancer, or peritonitis [1]. The origin of this disease is usually an appendiceal neoplasm that eventually takes on the appearance of mucoceles [4]. Most of the time, low-grade mucinous carcinoma peritonei is related to low-grade appendix mucinous neoplasms, and high-grade mucinous carcinoma peritonei is related to appendiceal mucinous adenocarcinoma [2]. The appendix and the ovary could be involved concurrently or successively in females [2].

It was long believed that PMP might be secondary to ovarian malignancies, but immunohistochemistry and molecular genetic examinations proved that most ovarian mucinous tumours come from perforated appendiceal mucinous tumours [1]. Only a few

rare cases of PMP emerging from a mature ovarian cystic teratoma are left as possible causes of PMP [4]. Ovarian tumours require a complete evaluation of the upper and lower digestive tracts to exclude metastasis, as primary malignant ovarian mucinous tumours are very rare [2]. Ovarian metastasis of low appendiceal mucinous neoplasms appears as a cystic ovarian mass mimicking a mucinous carcinoma or a borderline tumour of the ovary [5]. Several criteria help distinguish primary ovarian mucinous tumours from secondary ovarian mucinous tumours. Unilaterality, a large tumour exceeding 10cm, a smooth surface without extra-ovarian disease, the presence of benign or borderline areas, an expansile pattern of invasion, a low grade and low stage at presentation, and less aggressive behaviour — all these signs indicate a primary ovarian neoplasm. On the other hand, bilateral ovarian involvement, smaller size, ovarian surface involvement, multiple nodules, and an infiltrative pattern of stromal invasion favour an extra-ovarian origin [6]. Immunohistochemical markers such as CK20 and CK7 may help in determining the origin of an ovarian tumour [5]. CK20+ and CK- favour an appendiceal origin; however, overlap in the expression of these markers exists [5]. SATB2 (Special AT-rich sequence-binding protein 2) is a specific immunohistochemical marker that helps distinguish primary ovarian mucinous tumors from metastatic ovarian mucinous tumours. It is expressed by the cells that line the lower part of the digestive tract, mainly in the colon and the appendix. STAB2 is negative in primary ovarian mucinous neoplasms, excluding teratoma [2-5]. The risk of an appendiceal mucinous tumour includes familial adenomatous polyposis, KRAS, GNAS, and TP53 mutations [1]. The latter appears to be associated with a high-grade disease, a poor outcome, and female sex [1-7].

In PMP, the mucus tends to collect in the pelvis, the paracolic gutters, the liver capsule, the omentum, the retrovesical pouch, and the retrohepatic space. This is because the mucus follows the normal flow of the peritoneal fluid and the pull of gravity. When the ovaries are involved in a trans-celomic spread, they appear as a large, multiloculated mucin mass [3-7]. PMP is often found earlier in women; this may be explained by the rapid enlargement of the ovaries, which becomes symptomatic or clinically obvious. On the other hand, males are often diagnosed in the advanced stages, as the disease was asymptomatic initially. The tumour rupture is often not associated with any pain, and the mucus seals the luminal communication to the cecum and therefore prevents gross bacterial contamination. Incidental funding after appendectomies can lead to early detection in both males and females [3]. Clinical symptoms are not specific and do not correlate with the disease's progression. Patients may present with appendicitis-like syndrome, irritation bowel syndrome, bowel obstruction secondary to progressive fibrous adhesions, ascites, abdominal discomfort, distension, and palpable abdominal masses (ovarian masses, omental cake). Occasionally, tumours with mucin ascites accumulate in the inguinal hernia. Rectal examination may reveal deposits in the pouch of Douglas [1-7]. The mobile small bowel is typically spared in the early stages of the disease, but the fixed parts may be heavily involved. In cases of aggressive tumours, small bowel involvement can occur early [3]. PMP typically does not spread beyond the peritoneal cavity and does not metastasize to lymph nodes, especially in its low-grade form [7]. But there have been reports of extra abdominal spread, particularly to the pleural cavity [3]. Imaging plays an important role in the diagnosis. Ultrasound may detect peritoneal ascites and an appendiceal mucocele that appears as a cystic mass in the right iliac fossa with internal echogenic layering called the onion skin sign and possible mural calcifications exhibiting shadowing. On CT and MRI, the gelatinous ascites are distinguished from the fluid ascites by the scalloping on the surfaces of the spleen and liver and exhibit a low or proteinaceous attenuation [1-6]. CT and MRI also detect peritoneal nodules that manifest as omental cake in advanced stages and may cause extrinsic pressure on bowel loops [1-6]. Amorphous or curvilinear calcifications within the peritoneal implant may be seen [6].

Detecting the involvement of the small bowel mesentery, serosa, and porta hepatis using oral and intravenous contrast determines the possibility of complete resection [3]. Nevertheless, small bowel involvement is often best assessed during the surgery [3]. MRI also helps in the assessment of small bowel and hepatoduodenal ligament involvement [3]. DWI with a low b value helps visualize the septa within the intraperitoneal fluid and thus helps diagnose PMP [6]. It also has better sensitivity and specificity to detect peritoneal metastasis [3]. In all cases of PMP, the appendix should be well observed. On cross-sectional images, the mucocele appears as a well-encapsulated cystic mass with occasional mural calcification. Nevertheless, a residual perforated appendix can be difficult to detect [6]. PET CT is more interesting for detecting extraperitoneal metastasis. Percutaneous biopsies are unhelpful, but when performed, the acellular mucin finding is suggestive of PMP [3]. Serum tumour markers help with diagnosis, prognosis, follow-up, and early detection of recurrence [1]. High tumour markers preoperatively could guide the interval and frequency of follow-up. The three tumour markers used are the carcinoembryonic antigen (CEA), expressed by tumours of the gastrointestinal tract, especially colorectal cancers; carbohydrate antigen 125 (CA 125), a marker for ovarian tumours; and carbohydrate antigen 19.9 (CA 19.9), typically associated with pancreatic and upper gastrointestinal tract tumours but also expressed in peritoneal malignancy. In patients with any source of peritoneal irritation, CA125 and CA19.9 can be elevated [3]. The treatments include complete cytoreduction surgery (CCSR) and hyperthermic intraperitoneal chemotherapy

(HIPEC). CCSR includes bilateral parietal and diaphragmatic peritonectomy, right hemicolectomy, radical greater omentectomy with splenectomy, cholecystectomy, and liver capsulectomy; a pelvic peritonectomy with or without recto-sigmoid resection; and bilateral salpingo-oophorectomy with hysterectomy in females [1-3]. Even when the appendix is macroscopically normal, appendicectomy is always recommended in PMP [7]. When complete excision is not feasible, maximal tumour debulking is performed, permitting a significant survival advantage [3]. Systemic chemotherapy should be considered only in patients with no surgical options [4]. CRS and HIPEC permit disease-free survival at 1, 5, and 10 years up to 75%, 56–70%, and 67%, with an overall 5-year survival rate of 69–75% and overall 10-year survival rates of 57%, but are associated with major morbidity (7% - 49%) and mortality (0.6% - 4.4%) [3]. A CT scan with serum tumour markers is recommended in the follow-up, 1 year after the surgery, and annually for 10 years. If recurrence is suspected or the patient is symptomatic, imaging is performed earlier [3].

Conclusion

PMP is a complex clinical-pathological condition originating from a perforated appendiceal mucinous neoplasm in the majority of cases. It may spread secondarily to the ovaries and mimic ovarian tumours. Imaging plays an important role in diagnosis and in guiding the therapeutic approach. The optimal treatment is CCSR associated with HIPEC.

References

1- Kostov S, Kornovski Y, Slavchev S, Ivanova Y, Dzhenkov D, Yordanov A, et al. Pseudomyxoma peritonei of appendiceal origin mimicking ovarian cancer - a case report with literature review. Prz Menopauzalny. 2021; 20: 148-153.

2- Yan F, Shi F, Li X, Yu C, Lin Y, Li Y, et al. Clinicopathological characteristics of pseudomyxoma peritonei originated from ovaries. Cancer Manag Res. 2020; 12: 7569-7578.

3- Mittal R, Chandramohan A, Moran B. Pseudomyxoma peritonei: natural history and treatment. Inter J Hyperthermia. 2017; 33: 511-519,

4- Morera-Ocon FJ, Navarro-Campoy C. History of pseudomyxoma peritonei from its origin to the first decades of the twenty-first century. World J Gastrointest Surg. 2019; 11: 358-364.

5- Schmoeckel E, Kirchner T, Mayr D. SATB2 is a supportive marker for the differentiation of a primary mucinous tumour of the ovary and an ovarian metastasis of a low-grade appendiceal mucinous neoplasm (LAMN): A series of seven cases. Pathol Res Pract. 2018; 214: 426-430.

6- Suh DS, Song YJ, Kwon BS, Lee S, Lee NK, Choi KU, et al. An unusual case of pseudomyxoma peritonei associated with synchronous primary mucinous tumours of the ovary and appendix: A case report. Oncol Lett. 2017; 13: 4813-4817.

7- Carr NJ, Bibeau F, Bradley RF, Dartigues P, Feakins RM, Geisinger KR, et al. The histopathological classification, diagnosis and differential diagnosis of mucinous appendiceal neoplasms, appendiceal adenocarcinomas and pseudomyxoma peritonei. Histopathol. 2017; 71: 847-858.