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Pseudomyxoma Peritonei: A Challenging Clinical Diagnosis. Case Report and Review of the Literature

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Abstract. *Background: Pseudomyxoma peritonei (PMP) is a clinical entity of subtle onset abdominal pain, ascites, and distention associated with characteristic imaging. In most cases, laparoscopic exploration will give the definitive diagnosis and histopathologic verification. However, usually there are difficulties in the diagnosis of this disease. Case Report: Herein, we present a case of a 51-year-old female who developed ascites over 5 months. An investigational laparotomy established the diagnosis of PMP, after the discovery of a mucinous, grey-brown tumor that was CK20 positive and CK7 negative. Subsequently, chemotherapy with oxaliplatin combined with 5-FU (FOLFOX4 regimen), was initiated and the patient survived for 30 months. We also present a comprehensive review of the English literature concerning the different symptoms and radiological findings of this rare entity. According to the literature review, 35 cases of PMP with different clinical and radiological findings have been described. In the majority of the cases, ultrasound, computed tomography or magnetic resonance imaging was orientating towards a proper diagnosis before a diagnostic laparotomy. Conclusion: The combination of a clinical picture with the characteristic imaging findings enables a prompt diagnosis of PMP, making prognosis more favorable.*

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Pseudomyxoma peritonei (PMP) is a rare disorder within the abdomen clinically presented with abdominal pain and mass, fatigue, and weight loss. It is characterized by progressive accumulation of mucin in the peritoneal cavity and the presence of gelatinous ascites and peritoneal implants. Due to the fact that the symptoms are not specific, it is usually incidentally discovered during surgery for other reasons (1).

PMP is commonly associated with malignant tumors in the appendix, but there is a controversy regarding the epithelium from which it originates (2, 3). Theories of ovaries being an alternative primary site of PMP origin are considered to be convincing, although not fully explained when it comes to immunochemistry. The term is still being applied for a wide range of diseases and some oncologists and pathologists apply PMP to any condition with gelatinous material in the abdomen and pelvis (4).

The disease is three to four times more common in females than males and its incidence estimated at 1-2 per million per year (5, 6). This case report presents the challenging clinical diagnosis of a PMP and the intriguing effect that it had on the choice of treatment.

Case Report

A 51-year-old female presented to the emergency department with a five month history of gradually deteriorating abdominal pain, abdominal distension and ascites. The clinical examination did not reveal any palpable masses of the

Table I. *Patients with pseudomyxoma peritonei (PMP).*

Patient no.	Age/Sex	Presenting symptoms	Duration	Diagnostic imaging findings	IHC tissue	Histology	Refs.
1	66/M	Abdominal pain, nausea, and constipation	2 years	CT: peritoneal and liver lesions, perigastric ascites EUS: hypo-echoic peri-gastric and omental masses and perigastric ascites	CK7: + CK20: +	Low-grade mucinous adenocarcinoma	Darr *et al*. (8)
2	38/F	Progressive abdominal distension	N/A	CT: ascites and multilocular cystic mass MRI: cystic mass	CK7: – CK20: + CDX: +	Moderately differentiated adenocarcinoma in the cystic teratoma	Gohda *et al*. (9)
3	45/F	Pelvic effusion	N/A	u/s: hypoechoic fluid	N/A	Low-grade mucinous adenocarcinoma	Anania *et al*. (10)
4	42/F	None	N/A	u/s/MRI: adnexal cyst	N/A	Low-grade mucinous adenocarcinoma	Anania *et al*. (10)
5	35/M	Diffuse abdominal pain, abdominal girth	4 months	CT: mucinous soft tissue	N/A	Low-grade mucinous adenocarcinoma	Ghosh *et al*. (11)
6	30/F	Lower abdominal distension	N/A	U/S: multicystic mass	1. Ovary: CK7: + CK20: – CDX: – 2. Appendix: CK7: – CK20: + CDX: +	1. cells containing abundant cytoplasmic mucin with minimal atypia 2. mucinous cystadenoma	Zhou *et al*. (12)
7	80F	Swollen right thigh and fluctuation in the right lower quadrant of the abdomen	3 months	MRI thigh: fluid-containing lesion MRI calf: fluid-containing lesion CT: multiloculated cystic mass	N/A	Low-grade mucinous carcinoma peritonei	Joo *et al*. (13)
8	50/M	Lower abdominal pain	4 months	U/S: unilocular, hypoechogenic mass CT: polycystic mass with a calcified wall	N/A	Low-grade mucinous neoplasm	Agrawal *et al*. (14)
9	32/M	Mucinuria	N/A	U/S/CT: cystic mass	N/A	Mucinous urachal adenocarcinoma	Martínez *et al*. (15)
10	59/M	Abdominal turgidity and discomfort	N/A	U/S: cystic mass and massive ascites CT/MRI: mass with a calcified wall and ascites	N/A	Mucinous adenocarcinoma	Kebapçı *et al*. (16)
11	37/M	Abdominal pain	N/A	MRI: cystic mass and ascites	N/A	Mucinous borderline tumor of low malignant potential	Nozaki *et al*. (17)
12	63/M	Chronic urinary retention	N/A	MRI: cystic mass and nodularity within the abdomen and the pelvis	N/A	Mucinous urachal adenocarcinoma	Lamb *et al*. (18)
13	58/M	Abdominal fullness	N/A	CT/MRI: cystic mass with a calcified wall and ascites	N/A	Well differentiated adenocarcinoma	Sugiyama *et al*. (19)
14	14/F	Weight loss, bowel obstruction	7 months	U/S: ascites	N/A	Invasive moderately differentiated mucinous adenocarcinoma	Khalid *et al*. (20)
15	32/F	Infertility	N/A	CT: cystic masses	N/A	Mucinous urachal adenocarcinoma	Sugarbaker *et al*. (21)
16	47/M	Abdominal girth and ramping	1 year	CT: cystic mass and mucinous carcinomatosis	N/A	Mucinous urachal adenocarcinoma	Sugarbaker *et al*. (21)

Table I. *Continued*

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Patient no.	Age/Sex	Presenting symptoms	Duration	Diagnostic imaging findings	IHC tissue	Histology	Refs.
17	47/M	Mucusuria, abdominal extension and pain	N/A	CT: cystic mass	CK20: + CDX-2: + CK7: –	Well-differentiated mucinous adenocarcinoma	Yan *et al*. (22)
18	54/M	Left inguinal hernia	N/A	U/S/CT: cystic mass and ascites	N/A	Mucinous urachal cystic tumor of low malignant potential	Shinohara *et al*. (23)
19	82/M	Abdominal fullness	N/A	CT/MRI: cystic mass with a calcified wall	N/A	Mucinous urachal adenocarcinoma	Takeuchi *et al*. (24)
20	50/M	Abdominal fullness	N/A	CT: cystic masses with calcification MRI: cystic masses	N/A	Mucinous cystadenocarcinoma	Yanagisawa *et al*. (25)
21	54/M	Abdominal pain, rectal bleeding	6 months	u/s: cystic mass CT: unilocular mass with a calcified wall	N/A	Mucinous urachal adenocarcinoma *in situ*	Stenhouse *et al*. (26)
22	39/M	Abdominal pain, urinary problems, and fistula producing mucous	N/A	CT/MRI: multilobular cystic mass	N/A	Mucus with a moderate number of epithelial cells with moderate atypia	de Bree *et al*. (27)
23	34/M	Recurring tumor of urinary bladder	2 years	N/A	N/A	Low-grade mucinous urachal adenocarcinoma	de Bree *et al*. (27)
24	45/M	Abdominal distension	N/A	CT/MRI: cystic mass	N/A	Intermediated grade mucinous adenocarcinoma	Sasano *et al*. (28)
25	35/M	Gross painless hematuria	N/A	CT: cystic mass	N/A	Well-differentiated signet-cell adenocarcinoma	Loggie *et al*. (29)
26	71/M	Intermittent diarrhea	2 months	CT: ascites and cystic mass with a calcified wall	CK7: – CK20: +	Low- and high-grade mucinous adenocarcinoma	Touloumis *et al*. (30)
27	60/M	Abdominal pain	N/A	CT: fluid-filled dilated appendix with mural calcifications and intraperitoneal low-attenuation mass	N/A	Appendiceal mucinous adenocarcinoma	Idjuski *et al*. (31)
28	56/F	Abdominal pain and altered bowel movement	N/A	U/S: tumor-like mass	N/A	Borderline mucinous appendicular cystadenoma	Mavrodin *et al*. (32)
29	58/F	Abdominal pain, distension, and weight loss	2 months	MRI: cystic mass and ascites.	CK7: – CK20: + CDX-2: +	Low grade mucinous adenocarcinoma	Quiñonez *et al*. (33)
30	53/F	Abdominal girth and bloating	N/A	CT: multiloculated cystic mass, peritoneal carcinomatosis and ascites	N/A	Low-grade mucinous adenocarcinoma with associated pseudomyxoma peritonei	Peixoto *et al*. (34)
31	41/M	Abdominal girth and a weight loss	N/A	CT: mucinous ascites	N/A	Low-grade disseminated peritoneal adenomucosis	Becude *et al*. (35)
32	54/M	Abdominal pain, nausea, and fever	4 days	U/S: the loops of the intestines puffed up with gases	N/A	Mucinous cystadenoma	Demetrashvili *et al*. (36)
33	35/F	Abdominal pain, nausea, and anorexia	1 day	U/S: viable gestation	N/A	Simple mucocoele of the appendix	Idris *et al*. (37)
34	25/F	Infertility	12 months	CT/MRI: ovarian cyst with abundant peritoneal ascites	CK7: heterogenous CK20: + CDX-2: +	Ovarian teratoma containing an appendiceal-like structure with mucocele and LAMN	Csanyi-Bastien *et al*. (45)
35	51/F	Ascites with gradually deterioration	5 months	CT: ovary enlargement	CK7: – CK20: +	Mucinous adenocarcinoma	Present case

Duration: Duration of symptoms; CT: computed tomography; F: female; M: male; MRI: magnetic resonance imaging; N/A: not assessed; u/s: ultrasound; IHC: immunohistochemistry.

abdomen beyond marked ascites. The computed tomography (CT) scan of the upper and lower abdomen revealed an ovary enlargement and ascites. Serum tumor markers were also measured to be as follows: CEA=312.9 ng/ml, CA125=72.9 IU/ml, CA19-9=520.6 ng/ml.

The patient underwent an investigational laparotomy, and a tumor was found in the right iliac fossa, infiltrating the right ovary, right fallopian tube, and omentum. A second tumor was found in the left ovary as well.

An extrafascial total abdominal hysterectomy along with bilateral salpingo-oophorectomy and resection of the infracolic omentum was performed. Macroscopically, the right iliac tumor was described as having a mucinous, cystic form (5×5×3cm) with an irregular inner surface and a multilobar, mucinous, grey-brown outer surface. The second tumor sallied out from the left ovary, laterally, towards the fallopian interspace, infiltrating almost the entire left fallopian tube. It was a multilobar, mucinous, grey-brown tumor (3×3×2.5cm). The histopathology and immunohistochemistry report (CK20 positive and CK7 negative) confirmed the diagnosis of a mucinous adenocarcinoma, in the context of PMP.

Although the extensive sampling of the surgical specimen did not reveal histological evidence of appendix residue (CK20 positive rather than CK7) and elevated CEA, the patient was treated with oxaliplatin combined with 5-FU (FOLFOX4 regimen), as first line treatment for stage IV colorectal cancer. The re-evaluation after six cycles showed virtually stable disease and it was decided that she should continue with second line chemotherapy, using the combination of irinotecan and 5-FU (FOLFIRI regimen).

Three months later, the patient was stable and underwent plastic surgery for omphalocele and white line hernia. The pathology report showed infiltration with mucinous cystadenocarcinoma, with extracellular and intracellular mucin production (signet-ring cells). Palliative treatment was offered, however, the patient died 30 months after the primary diagnosis.

Review of the literature. We searched MEDLINE for papers using the following key words: "pseudomyxoma peritonei", "cancer", "malignancy", "presenting symptoms" and "laparatomy". Reference lists in the retrieved papers were checked to identify any other published data. Cases were included if there was both clinical and histological evidence of PMP. Only papers written in the English language and cases with patients older than 14 years were included in the analysis. Data regarding the primary tumor, age, sex, presenting symptoms, and the clinical and diagnostic work up were analyzed.

Table I summarizes the 35 cases, including ours, that were identified in the literature review for patients experiencing symptoms of "pseudomyxoma peritonei", presented with a variety of clinical pictures, stages at time of surgery, and histologic types.

Discussion

This report describes the clinical experience of managing a rare PMP case. The correct diagnosis of the disease is considered of high importance since the discovery of the origin indicates the appropriate treatment regimen. It is emphasized that even with no histological evidence of appendix residue; we must consider the primary origin as colorectal cancer (7). In this case, the immunohistochemistry findings (positive CK20 and negative CK7) and elevated tumor markers (CA19-9 and CEA) strongly supported the colorectal origin of this PMP. What merits a particular mention is the classification that pathologists have compiled for PMP subtypes, with different prognosis for each one: disseminated peritoneal adenomucinosis (DPAM), peritoneal mucinous carcinomatosis (PMCA), and an intermediate PMCA type (PMCA-1) (7).

PMP is a slowly progressive disease that can be presented with a variety of symptoms. Usually, PMP exhibits the characteristic "jelly belly", as the amount of mucoid fluid fills the peritoneal cavity. Symptoms that have been described are abdominal pain, nausea, vomiting, fatigue, and urinary abnormalities. It can also be presented with local symptomatology, mirroring the location of the primary or metastatic tumor, like appendicitis (38).

In the majority of cases as we can see in Table I, the diagnosis was made only by CT (11 cases) with the assistance of ultrasound (u/s) or magnetic resonance imaging (MRI), followed by histopathologic verification of an extensively sampled tumor. In six cases, only u/s was performed (10, 12, 20, 32, 36, 37), but in two of them (36, 37), u/s misled the diagnosis, resulting in delayed treatment. Thus, CT or MRI would be probably the most appropriate imaging option, offering accurate and prompt diagnosis. Ultrasound usually describes cystic masses and mucinous substance as retroperitoneal fluid. CT and MRI will differentiate the fluid (watery *vs*. mucinous) and confirm the cystic nature of the masses.

Another important aspect for the diagnosis and follow-up of PMP patients is the assessment of serum tumor markers CEA and CA19-9. CEA has been reported to be increased in 56% to 75% of patients and CA19-9 in 58% to 67% respectively, whilst the baseline tumor marker values seem to be related to the extent of tumor and completeness of resection (39, 40).

Analysis of immunohistochemical markers' expression, such as cytokeratin (CK) 7, 20, CEA, and CDX-2 is very useful in characterizing the origin of tumors. Positive expression of CK20, CEA, and CDX-2 imply primary colorectal or appendiceal origin while CK7 and CA125 indicate a primary ovarian origin (41).

There is currently no consensus regarding the optimal treatment for progressive PMP. Cytoreductive surgery, with

or without hyperthermic intraperitoneal chemotherapy (HIPEC), despite possible concomitant morbidity is considered the proper treatment (42). Although several approaches have been described so far, the radical removal of all intra-abdominal and pelvic disease and the administration of intraperitoneal heated chemotherapy (mitomycin, 5-FU) have been adopted by most clinicians (43, 44).

Conclusion

To conclude, the reported symptoms for PMP vary, the clinical course is vague, and there is no certain pathognomic signs that can lead to the prompt diagnosis and management of this clinical entity. Therefore, clinicians should demonstrate high index of clinical suspicion, as to discover this medical entity without delay.

Conflicts of Interest

No potential conflicts of interest exist in relation to this study.

Authors' Contributions

All Authors equally contributed to this study regarding the conception and design of the study, literature review and analysis, drafting, critical revision, editing, and final approval of the final version.

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