International Journal of Innovative Research in Medical Science (IJIRMS)
Volume 09, Issue 04, April 2024,
https://doi.org/10.23958/ijirms/vol09-i04/1859

Case Report

Pseudomyxoma Peritonei: A Case Report of a 73-Year-Old Male Presenting with Abdominal Distension

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Received 14 March 2024; Accepted 28 March 2024; Published 01 April 2024

Abstract

Pseudomyxoma peritonei is characterized by diffuse mucinous ascites and mucinous implants on the peritoneal surfaces. The authors report a case of a 73-year-old man with complaints of abdominal distention, nausea, fatigue, and asthenia for the past two months. The patient underwent diagnostic laparoscopy, and histological examination revealed pseudomyxoma peritonei secondary to low-grade mucinous carcinoma with intestinal differentiation. The patient was referred to Oncology for hyperthermic intraperitoneal chemotherapy along with complete cytoreduction surgery.

Keywords: ***pseudomyxoma peritonei, mucinous ascites, oncology, laparoscopy, hyperthermic intraperitoneal chemotherapy.***

Introduction

Pseudomyxoma peritonei (PMP) is a rare clinical entity with an estimated incidence of 1 to 4 cases per million per year and appears to be more common in women [1]. It is characterized by diffuse mucinous ascites along with mucinous implants on the peritoneal surfaces that results from mucus-producing neoplasms [2]. Although it can originate from every gastrointestinal organ, the appendix appears to be the most common [3]. Nonappendiceal origin mucin-producing neoplasms have a lower incidence but are more likely to be adenocarcinoma. However, despite the histopathology, their prognosis is not worse for reasons that are not yet fully understood [4]. In most cases, PMP adopts an indolent behavior and was once considered a benign disease. However, if left untreated, it can rapidly progress, leading to severe symptoms such as markedly increased abdominal girth secondary to large volume ascites, intestinal perforation, or intestinal obstruction [2,5]. Here we report a case where a common symptom was caused by a rare clinical entity.

Case report

A 73-year-old male with a prior history of hypertension, diabetes, and prostate adenocarcinoma in remission after radical prostatectomy presented to the emergency department with complaints of increasing abdominal distension for the past two months, along with nausea, fatigue, and asthenia. He denied any other symptoms such as fever, weight loss, change of bowel habit, melena, or hematochezia.

On physical examination, there was marked abdominal distension with shifting dullness on percussion without tenderness, guarding, or rebound on palpation. The remainder of the physical examination was unremarkable. Laboratory work showed elevated C reactive protein (8.94 mg/dL, reference range 0–0.5 mg/dL) and discrete normocytic and normochromic anemia with hemoglobin of 11.9 g/dL (reference range 13.0–17.0 g/dL). An abdominal ultrasound was performed, revealing septated ascites that did not move when externally compressed (**Figure 1A**). A contrast-enhanced abdominal CT scan was then performed, which showed large volume ascites and scalloping of the liver (**Figure 1B**).

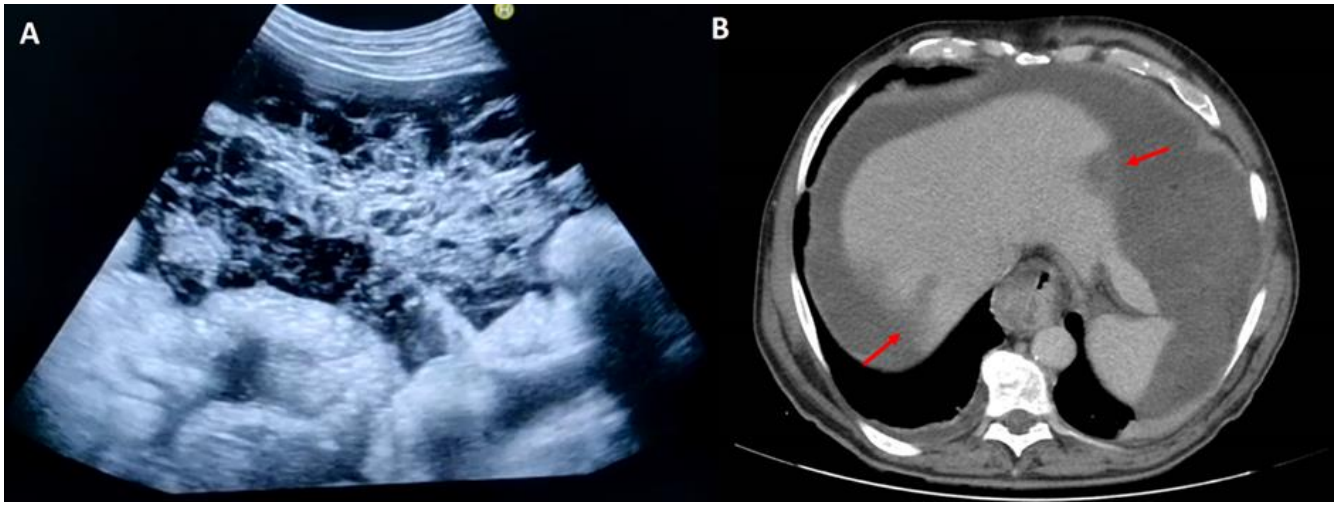

Figure 1: A - Abdominal ultrasound revealing ascites with septation; B – Abdominal CT scan showing large volume ascites and scalloping of the liver (red arrows).

The patient was admitted to the Internal Medicine ward for further investigation. An upper endoscopy and a total colonoscopy were performed, but both showed no relevant abnormalities. He was then subjected to diagnostic and therapeutic paracentesis, and approximately 5 liters of viscous dark yellow fluid were drained. Ascitic fluid cytology could not be performed because of its viscosity, but the chemical examination showed an elevated protein level of 4.9 g/dL and a serum to ascites albumin gradient of 0.5 g/dL. Adenosine deaminase and amylase levels were normal. The histopathology of the ascitic fluid revealed atypical cells but was otherwise inconclusive; therefore, a diagnostic laparoscopy was performed, which revealed a marked accumulation of gelatinous mucus in peritoneal cavity (**Figure 2**).

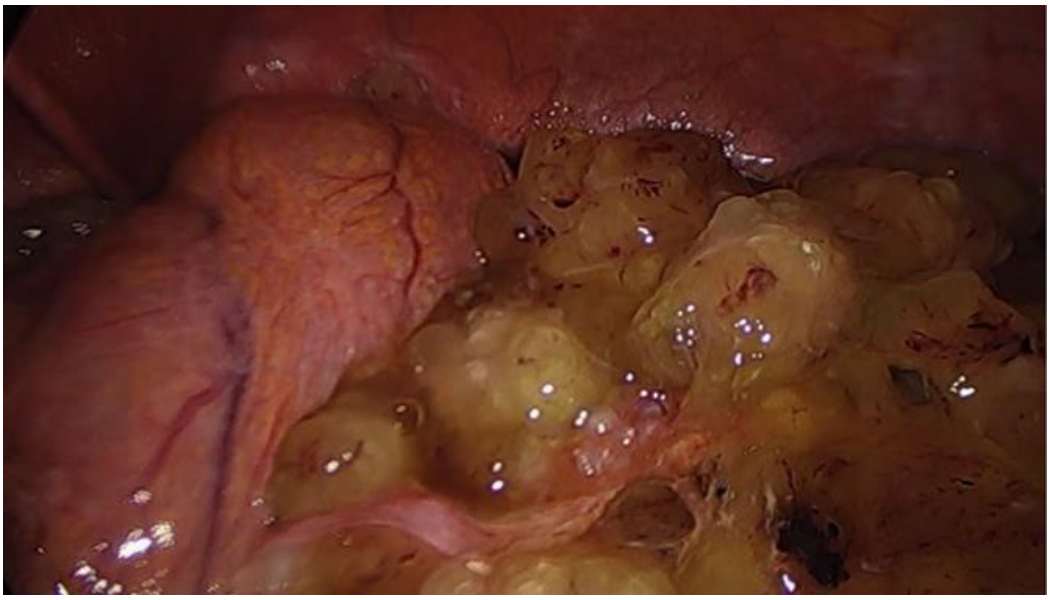

Figure 2: Diagnostic laparoscopy showing accumulation of gelatinous mucus in peritoneal cavity.

Several biopsies were performed, and the histopathology was compatible with PMP secondary to low-grade mucinous carcinoma with intestinal differentiation. Immunochemical analysis showed a RAS mutation in addition to positive expression of CK20 and CDX2 but negative expression of CK7 markers [6]. The patient was referred to Oncology for hyperthermic intraperitoneal chemotherapy (HIPEC) and complete cytoreduction surgery.

Discussion

Most cases of PMP originate from the appendix, although they can be derived from any mucin-producing neoplasm in the gastrointestinal tract. In this case report, PMP did not have an appendiceal origin but was secondary to low-grade mucinous carcinoma with intestinal differentiation. Despite the less common etiology, the clinical presentation was one of the most common, according to the literature, namely increased abdominal girth. Other frequent presentations are acute appendicitis, and suspected internal hernia [5]. PMP has a wide clinical spectrum ranging from slow-growing benign lesions to rapidly progressing malignant disease, and if left untreated, it can lead to severe symptoms such as cachexia or bowel obstruction [7].

The diagnosis of PMP requires a high level of suspicion. Contrast-enhanced CT is the preferred imaging exam for diagnosis. It can identify the hallmarks of the disease, such as the scalloping of abdominal organs, particularly the liver, thus allowing the differentiation of mucinous from fluid ascites and can identify septae within mucinous ascites [8]. However, septae within mucinous ascites can also be identified by an abdominal ultrasound which was first performed in this case report, raising clinical suspicion for PMP which warranted further investigation. Histopathological samples for definitive diagnosis are usually obtained through laparoscopy or

laparotomy, as imaging-guided percutaneous biopsy is not helpful because the collected material is frequently acellular mucin [2]. In the present case, a laparoscopy was performed since the histopathology of the ascitic fluid, obtained during paracentesis, was inconclusive, despite showing the presence of atypical cells.

Treatment consists of tumor removal surgery combined with HIPEC, although it is not feasible for every patient and some of them do not meet the performance status requirements for the procedure, the gold-standard treatment consists of tumor removal surgery combined with HIPEC. If complete cytoreduction is achieved and the patient undergoes HIPEC, the 5-year predicted survival can achieve 87%. When complete cytoreduction surgery is not possible, major tumor debulking can be performed. In these cases, the 5-year survival rate drops do 34% even when in combination with HIPEC [9].

Conclusions

The diagnosis of PMP can be challenging, and physicians must have a high index of suspicion. Clinical presentation is non-specific and in some cases the disease takes an indolent course, further delaying the diagnosis. The presence of specific findings in either contrast-enhanced abdominal CT scan or ultrasound, such as septated ascites and scalloping of the organs, should raise suspicion for PMP and warrant further investigation.

Treatment with complete cytoreduction surgery combined with HIPEC is the goal, granting a good prognosis.

Ethics approval and consent to participate

Not applicable.

List of abbreviations

PMP: Pseudomyxoma peritonei
CT: Computerized tomography
HIPEC: Hyperthermic intraperitoneal chemotherapy

Data Availability

Not applicable.

Conflicts of Interest

The author(s) declare(s) that there is no conflict of interest regarding the publication of this paper.

Funding Statement

The author(s) received no financial support for the research, authorship and publication of this article.

Authors' contributions

RAP was responsible for research design, data collection, drafting of the paper and spelling revision. MCR, JC and FM were responsible for data collection. RGP was responsible for laparoscopy and pictures. HJC was responsible for revision. All authors read and approved the final manuscript.

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