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CASE REPORT

Case Report: Malignant Pseudomyxoma Peritonei

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Abstract

Pseudomyxoma peritonei (PMP) is a poorly understood condition characterized by mucinous ascites diffusely involving the peritoneal surfaces.

There is considerable debate regarding the definition, pathology, site of origin and prognosis of PMP.

In this article, we discuss how an asymptomatic middle-aged man who was being investigated for an incidental finding of ascites was finally diagnosed to have a malignant variant of PMP.

Keywords: Abdominal pain, Appendix, Ascites, disseminated peritoneal adenomucinosis, Malignancy, peritoneal mucinous carcinomatosis, Pseudomyxoma peritonei

Introduction

PMP is a rare disease, where a gelatinous ascites accumulates within the peritoneal cavity.[1, 12] Classically, PMP refers to a ruptured benign cystadenoma of the appendix with secondary seeding of mucus-producing cells into the peritoneal cavity, which then continue to proliferate and form the 'jelly belly" which is called the PMP. [2, 12]

However, in recent years, many clinicians use the term PMP to denote any mucinous ascites that arises either from a ruptured benign cystadenoma or from the dissemination of an aggressive mucin producing adenocarcinoma.[3, 4, 12]

This has led to the development of the more recent terms, disseminated peritoneal adenomucinosis (DPAM) and peritoneal mucinous carcinomatosis. DPAM refers to the ruptured benign mucinous cystadenoma, while peritoneal mucinous carcinomatosis refers to the spread of the more aggressive mucinous adenocarcinoma.[2, 3, 5]

The difference between the two can be explained by the difference in their prognosis. In one study, the age-adjusted five-year survival for patients with DPAM was 84% compared with 7% for those with peritoneal mucinous carcinomatosis and 38% for those with intermediate features.[5, 12]

The most common clinical presentation of PMP is an increase in abdominal girth (in both men and women). The second most common presentation is an inguinal hernia in men, while for women, it is an ovarian mass palpated at the time of a routine pelvic examination.[2, 12]

The Case

History and Physical examination

A 65-year-old Sudani male who was not known to have any prior medical illness presented to the gastroenterology clinic in a tertiary hospital in Bahrain following an incidental finding of ascites, which was discovered when he had sought medical care due to lower abdominal pain at his home country.

The patient had experienced acute lower abdominal pain three months back, that had lasted for a few hours and had gradually resolved over the next few days. Since then the patient had been asymptomatic.

At the time of the event, the patient had been investigated with an abdominal ultrasonogram (USG) and was found to have ascites. He was further advised additional investigations, which he decided to get done once he returned to Bahrain.

The patient was not known to have of any liver disease and had no history of notable abdominal distention, nausea or vomiting. Bowel habits were unchanged, with no history of hematemesis, coffee ground vomiting, melena or hematochezia. There was no history of weight loss, night sweats, episodes of hot flushes, diarrhea, dyspnea or fever.

The patient had no past medical history and no history of previous surgeries or medical treatment. No abnormalities were detected during general and systemic examination.

Diagnostic assessment and Investigations

Laboratory investigations

A full blood profile including complete blood count, biochemical profile (urea, electrolytes, renal and liver function test) and tumour markers were within normal limits.

Serology was negative for the presence of any of the hepatitis viruses.

Radiological investigations:

USG Abdomen showed multiloculated ascites, suspicious of Pseudomyxoma peritonei (PMP), warranting further investigations (Figures 1 and 2).

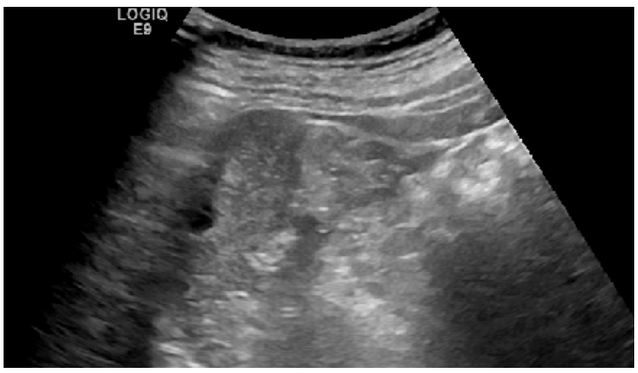

Figure 1: Left upper abdominal and lower thoracic USG scan noted subdiaphragmatic perisplenic loculated small fluid collection compressing the splenic subdiaphragmatic surface with splenic surface indentations

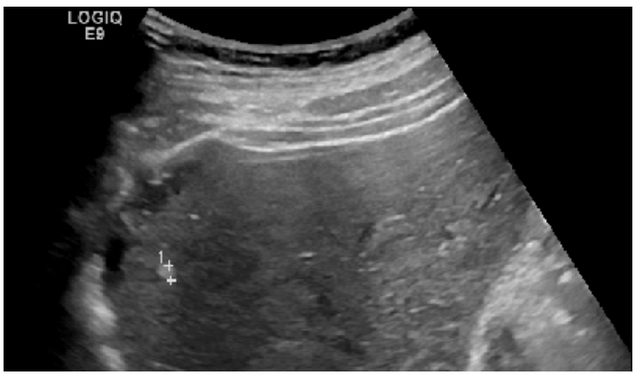

Figure 2: Right upper abdominal and lower thoracic USG scan noted subdiaphragmatic perihepatic loculated fluid collection compressing hepatic surface with surface indentations

Double contrast (oral and intravenous) enhanced CT Abdomen and Pelvis was further performed, that showed the following images as in Figures 3, 4 and 5.

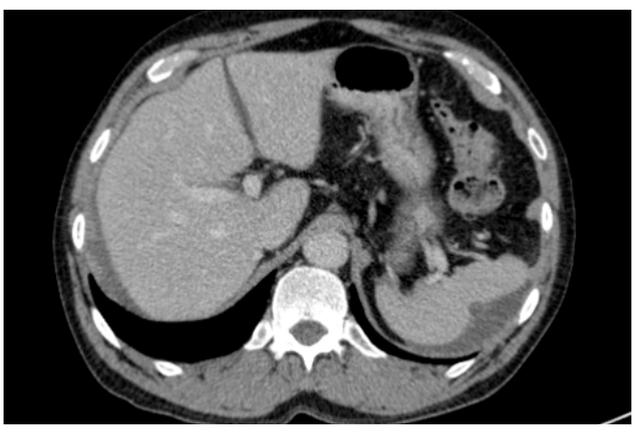

Figure 3: CECT axial scan of the upper abdomen revealed left and right subphrenic fluid collection of high density with splenic and hepatic surface indentations

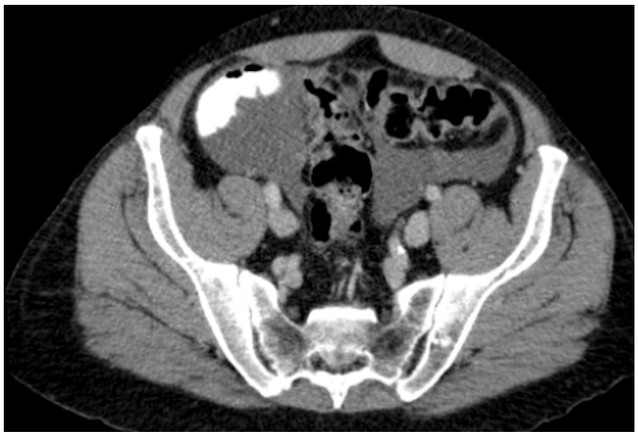

Figure 4: CECT pelvic area in axial scan revealed loculated intraperitoneal pelvic fluid collection of high density

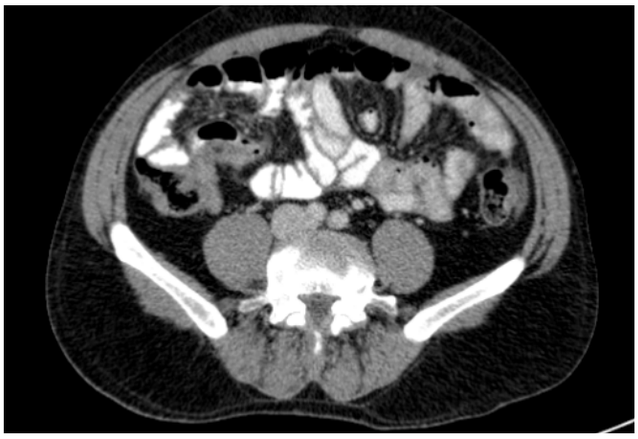

Figure 5: CECT axial scan of the pelvis revealed thickening of the mesoappendix with soft tissue density as well as appendicular noninflammatory mural thickening

The final CT report mentioned ascites with multiple loculations around liver, spleen and pelvic organs as well as appendicular enhancement and thickened wall, that were highly suggestive of pseudomyxoma peritonei with an appendicular tumour as a primary source.

Given the mucinous ascites and high clinical suspicion, fluid sampling using USG guided aspiration of the peritoneal fluid was performed. Thick gelatinous greenish fluid was collected and sent for cytology.

The report following cytology and microscopic examination of the aspirated fluid revealed thick mucin with many nests of mildly pleomorphic cells having abundant cytoplasm. The appearance was consistent with mucinous adenocarcinoma in keeping with pseudomyxoma peritonei.

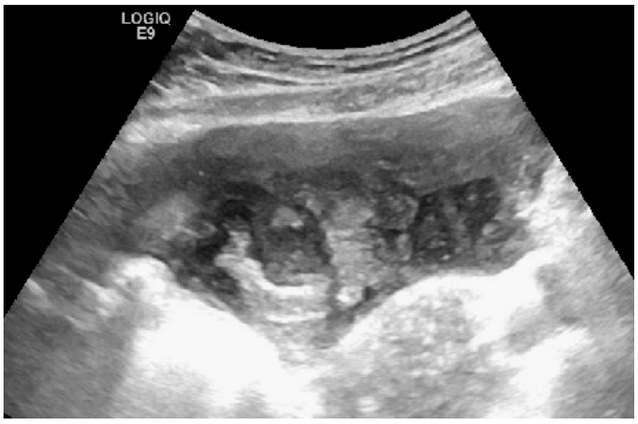

Figure 6: Pelvic U/S revealed turbid loculated nonmobile intraperitoneal fluid

A full ileo-colonoscopy was performed to further investigate the appendix and cecum. However, the study was normal and showed no pathology (Figure 7).

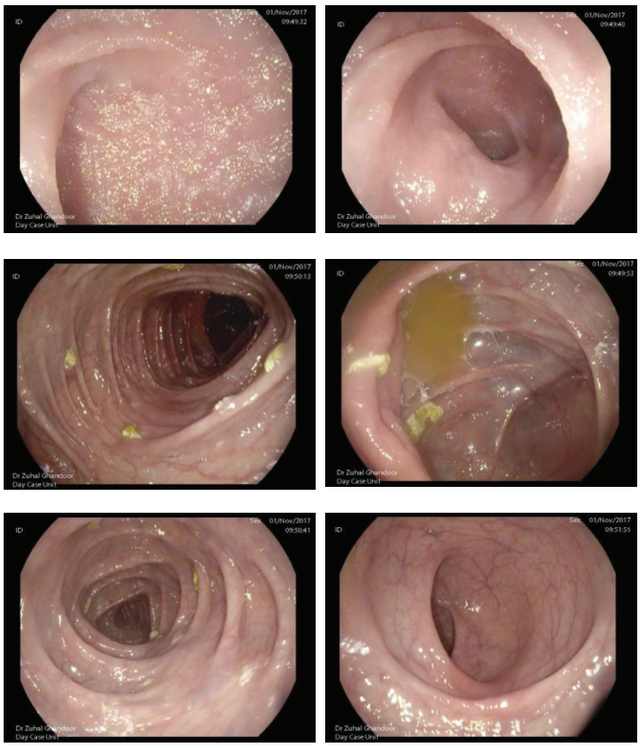

Figure 7: Normal ileocolonoscopy

Final diagnosis, therapeutic interventions and further care

The final diagnosis was Pseudomyxoma peritonei, secondary to disseminated mucinous adenocarcinoma with a high suspicion of appendiceal cancer as the primary tumor.

The patient needed specialized surgical management and hence was referred to a center specializing in peritoneal malignancy abroad where he underwent

Table 1: Timeline of investigation and managment

Date	Investigation/Treatment	Findings
26 October 2017	Ultrasound Abdomen	Multiloculated Ascites
30 October 2017	CT Abdomen and Pelvis with IV and Oral contrast	Ascites with multiple loculations, enhanced appendix with thick wall
31 October 2017	Ultrasound guided peritoneal fluid sampling	3ml of thick gelatinous greenish fluid
31 October 2017	Cytology of peritoneal fluid	mucinous adenocarcinoma
1 November 2017	Colonoscopy	Normal
Mid November 2017	Surgery + HIPEC	Low grade mucinous carcinoma
May 2018	6 months Post-op CT Abdomen and Pelvis	mural thickening of proximal small bowel with matting

right hemicolectomy, complete right peritonectomy and pelvic peritonectomy, partial left peritonectomy, omentectomy, Glisson capsulectomy, splenectomy and Hyperthermic Intraperitoneal Chemotherapy (HIPEC).

We followed up the patient at our clinic till March 2018. The patient was doing well and was asymptomatic. Repeat analysis of tumour markers showed CEA 1.58 ng/ml (previously 12.5 ng/ml) and CA19-9 16.38 U/ml (previously 22.32 U/ml). The patient also underwent a repeat CT abdomen and pelvis on 20/5/18 which showed moderate symmetrical mural thickening of the proximal small bowel with matting and no intraperitoneal or retroperitoneal fluid collection.

Discussion

PMP is an uncommon condition and the findings on physical examination might not always be helpful. The use of radiological investigations is beneficial for the diagnosis of these conditions

The routine investigations for ascites, which include serum biochemical profile and tumor markers, in addition to an initial radiological investigation with USG Abdomen will always help to point the diagnosis.[7, 8]

In this case, the only positive finding that led to the diagnosis was the initial suspicion on ultrasonography. Moreover, CT abdomen increased the suspicion and gave the impression of presence of the primary tumor in the appendix and cecum.

However, a full ileo-colonoscopy showed no evidence of any pathology in that area, revealing that a perfectly normal mucosa cannot rule out a cancerous tumour that is either intramural or subserosal.

This could indicate that use of endoscopy is limited when dealing with these types of tumors and a more invasive surgical or radiological approach would be superior in diagnosing these conditions.

Furthermore, as with most tumors, the definitive diagnosis is only made after macroscopic and microscopic findings through histopathology. In this case, ultrasound-guided fluid sampling was used for histopathological evaluation of ascites. However, as an appendiceal sample was not obtained, it was not evaluated histopathologically. Hence, the specific site of the primary lesion and its histopathology could not be identified.

The basis of management for PMP is surgical debulking, aiming to manage symptoms. However, with the inevitable recurrence, further surgery can be more complicated.[3] Therefore, more aggressive approaches are recently being considered to be of choice, with cytoreductive surgery aiming to remove all intraabdominal and pelvic disease and administration of local chemotherapy along with the use of heat.[6] The aim of the chemotherapy is to eliminate any minimal residual disease and the use of heat is to enhance local tissue drug concentration.[9] This technique is defined as Heated Intraperitoneal Chemotherapy (HIPEC) and can achieve a more satisfactory clinical outcome with PMP.[10, 11]

The limitations we faced when dealing with this case was the lack of readily available expertise on

treating this type of tumour in our region, due to its rarity. The patient, for the lack of readily available expertise, traveled abroad for high quality and experienced care.

Conclusion

This study highlights the importance of investigating the presence of ascites when detected with routine radiological and biochemical/histopathological techniques, as the etiology of ascites and its associated manifestations will define patient management and prognosis.

Moreover, this study denotes how useful the classical operator dependent ultrasonography can be. In this case, the first and only initial suspicion of an underlying PMP was through an ultrasound scan. The next radiological step was a Contrast enhanced CT Abdomen to confirm the presence of the loculation and to have a different perspective of the image in order to confirm or rule out the initial diagnosis and try to find the primary site of the tumor. [3]

Fluid sampling and histopathological evidence of the tumor is always a must for a final definitive diagnosis.

Furthermore, all these patients should be followed up at specialized surgical centers with expertise in managing such conditions as proper treatment could very well improve survival and quality of life.

Conflict of Interest

The authors have no financial or proprietary interests in any material or method mentioned.

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