Pseudomyxoma Peritonei—An Unusual Cause of Ascites: A Case Report

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ABSTRACT

Introduction: Ascites in patients with cardiac disease can be multifactorial. Serum ascitic albumin gradient (SAAG) helps in identifying the etiology of ascites. High SAAG ascites is related to hepatic or posthepatic causes. The causes of low SAAG ascites results include tuberculosis, peritoneal malignancy, or pancreatitis.

Case Presentation: We report an unusual cause of low SAAG ascites in a 48-year-old woman with valvular heart disease. The patient presented with ascites, and cross-sectional imaging revealed a right iliac fossa mass with omental deposits. The patient was finally diagnosed as having pseudomyxoma peritonei on the basis of clinicoradiopathological features.

Discussion: Pseudomyxoma peritonei is a rare cause of low SAAG ascites. It is characterized by the deposition of mucinous material on the peritoneal surfaces. The most common site of origin is the appendix, although it can arise from other intraabdominal organs as well. Excision of the tumor combined with intraperitoneal chemotherapy is the preferred modality of treatment.

INTRODUCTION

Pseudomyxoma peritonei is a rare condition[1] characterized by accumulation of gelatinous material in the abdominal and pelvic cavity along with mucinous implants over the peritoneum and omentum.[2] We present a case of pseudomyxoma peritonei as an unusual cause of ascites.

CASE PRESENTATION

Presenting Concerns

A 48-year-old woman was referred to the gastroenterology clinic for evaluation of ascites. She had been receiving medical management of rheumatic heart disease with severe mitral stenosis and regurgitation. Initially she was seen in the cardiology clinic for evaluation of worsening shortness of breath and increasing abdominal distention for the prior 3 weeks. Results of ultrasonography of the abdomen revealed moderate ascites and a solid mass in the right lower aspect of the abdomen.

The patient was referred to us for further evaluation. Her ascites revealed a jelly-like fluid (Figure 1) with a low serum ascites albumin gradient (0.9). The results of the contrast-enhanced computed tomography scan of the abdomen revealed moderate ascites with scalloping of the liver and spleen (Figure 2), omental deposits (Figure 3), and a mass (9 × 2 cm) in the right iliac fossa (70 Hounsfield units) with central calcification (140 Hounsfield units) and medial displacement of the ileal loops (Figure 4). The appendix was not separately visualized from the lesion.

The results of the colonoscopy were normal. Results of an ultrasonography-guided biopsy of the right iliac fossa mass revealed a single layer of mucinous cells with occasional goblet cells lining the fibromuscular tissue (Figure 5). On the basis of the clinicoradiopathologic findings, a diagnosis of low-grade appendiceal mucinous neoplasm with pseudomyxoma peritonei was made.

FOLLOW-UP AND OUTCOMES

The patient was referred to a surgical oncologist for treatment, but she died of her cardiac illness, with pseudomyxoma peritonei being the precipitating factor. The family refused an autopsy.

DISCUSSION

Pseudomyxoma peritonei is a rare condition, with an incidence of 1 to 2 per 1 million per year.[1] The disease is characterized by accumulation of gelatinous material in the abdominal and pelvic cavity along with mucinous implants over the peritoneum and omentum.[2] First described in a patient with a mucinous neoplasm of the ovary, the most common site of origin is the appendix, with other reported sites being the colon, rectum, gallbladder, pancreas, fallopian tube, urinary bladder, and lung. Pseudomyxoma peritonei is thought to originate in the appendix.[3,4] Pseudomyxoma peritonei is more common

Figure 1. Jelly-like ascitic fluid.

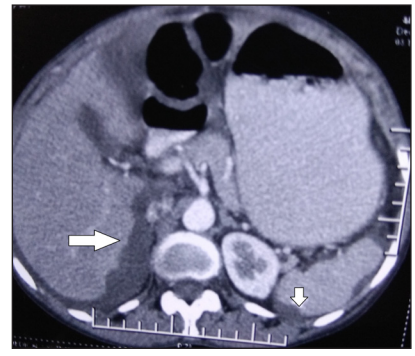

Figure 2. Contrast-enhanced computed tomography of the abdomen showing moderate ascites with scalloping of the liver and spleen (arrows).

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in women, and synchronous appendiceal and ovarian pathologies are quite prevalent. Studies suggest that ovarian disease is likely a result of metastases from a perforated appendix rather than the site of origin.[5] The most common presenting symptom is abdominal distention followed by an inguinal hernia.

The results of the cross-sectional imaging revealed characteristic findings, showing ascites with omental deposits, scalloping of the liver and spleen, calcifications, and peripheral location of the tumor with central displacement of the bowel (redistribution phenomenon).[6] The pattern of distribution of pseudomyxoma peritonei in the abdominal cavity is striking and characteristic of this condition. Gravity and physical factors such as movement of peritoneal fluid in the abdomen lead to accumulation of mucinous deposits at the omentum, retrohepatic region, and rectovesical pouch,[6] giving the characteristic appearance of visceral scalloping and omental caking. Visceral scalloping denotes the presence of mucinous fluid (exudative) and differentiates it from transudative ascitic fluid.[7] Although characteristic, visceral scalloping is not specific for pseudomyxoma peritonei and has been reported in abdominal tuberculosis as well.[8]

Conventional treatment has been limited to repeated debulking surgery and intraperitoneal chemotherapy. The current optimal treatment is macroscopic tumor excision (cytoreductive surgery) combined with hyperthermic intraperitoneal chemotherapy.[9] Five-year survival after cytoreductive surgery and hyperthermic intraperitoneal chemotherapy is 60% to 100% for low-grade disease and 0% to 60% for high-grade disease.[10-13] Five-year median survival is 79.5%, and 10-year median survival is 55.9% as reported in a systemic review conducted in 2013.[14]

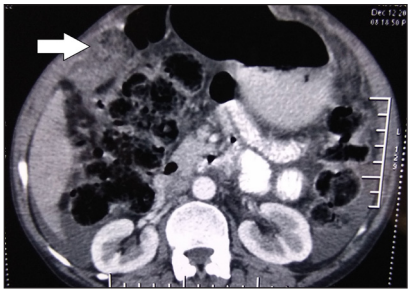

Figure 3. Contrast-enhanced computed tomography of the abdomen showing omental deposits (arrow).

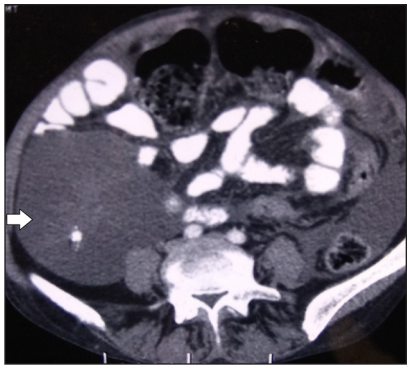

Figure 4. Contrast-enhanced computed tomography of the abdomen showing a mass in the right iliac fossa (arrow) with central calcification and displacement of the ileal loops medially.

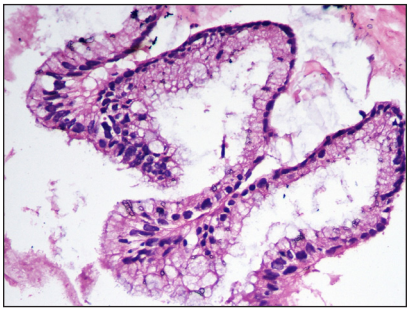

Figure 5. Biopsy specimen from the right iliac fossa mass showing a single layer of mucinous cells with occasional goblet cells lining the fibromuscular tissue.

The cause of ascites in patients with cardiac disease is not always secondary to cardiac failure. Pseudomyxoma peritonei ranks very low as an etiology for low SAAG ascites in terms of prevalence as compared to tuberculosis, pancreatitis, and peritoneal carcinomatosis. Fluid characteristics may assist in the diagnosis of pseudomyxoma peritonei, and cross-sectional imaging and histopathology may aid in confirmation. Although pseudomyxoma peritonei carries a poor prognosis, if diagnosed early and if the disease is low grade, patients can have a good 5-year survival.

Our patient had pseudomyxoma peritonei, which is a rare cause of ascites. She had underlying multivalvular heart disease, where ascites secondary to cardiac failure is more common than pseudomyxoma peritonei. The appendix was presumed to be the site of origin for pseudomyxoma peritonei in our patient without any ovarian pathology. This kind of presentation continues to be an issue of debate among oncologists interested in research on pseudomyxoma peritonei.

Table 1. Timeline of the case

Date	Presentation, treatment, and outcomes
1/17/18	The patient was evaluated for exertional dyspnea and shortness of breath. She was found to have ascites and a mass in the abdomen on USG.
1/20/18	She was referred for further evaluation. An ascitic tap was performed, which showed jelly-like fluid; SAAG was 0.9.
1/21/18	CECT of the abdomen was performed, which showed moderate ascites with scalloping of the liver and spleen, omental deposits, and a RIF mass with central calcification.
1/24/18	The patient underwent a colonoscopy, with normal results. She underwent an USG-guided biopsy of the RIF lesion.
1/29/18	HPE of the lesion showed a low-grade appendiceal mucinous neoplasm with PMP. The patient was referred to a surgical oncologist.
2/2/18	The patient died of cardiac disease. The patient's family refused an autopsy.

CECT = contrast-enhanced computed tomography; HPE = histopathological examination; PMP = pseudomyxoma peritonei; RIF = right iliac fossa; SAAG = serum ascites albumin gradient; USG = ultrasonography.

CONCLUSION

We present a case of a patient with valvular disease who presented with low SAAG ascites. Cross-sectional imaging revealed a right iliac fossa mass with omental deposits. Although pseudomyxoma peritonei is not a common cause of low SAAG ascites among tuberculosis, pancreatitis, and peritoneal carcinomatosis, a diagnosis can be suggested by the fluid characteristics and confirmed by cross-sectional imaging and histopathology. Early diagnosis has a good 5-year survival. ❖

Disclosure Statement

The author(s) have no conflicts of interest to disclose.

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Bathing the Heart

Nature [has] placed various glands round the base of the heart. From these fluid trickles out [and] bathes the entire surface of the heart, and thereby renders its movement more ready and more easy of accomplishment. … This fluid is not entirely excretory … but rather part of the nutrient Serum oozing from the blood. … It sets into white jelly when heated only a very little.

— Richard Lower, 1631-1691, English physician