Abstract

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# A rare case of mucinous ascites in an otherwise healthy patient

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## Background

Pseudomyxoma peritonei (PMP) is a rare clinical syndrome characterized by diffuse mucinous peritoneal involvement usually associated with appendiceal mucinous neoplasms. Appendiceal mucinous lesions are a rare group of lesions characterized by a distended, mucus-filled or ruptured appendix. They may be classified as non-neoplastic or neoplastic lesions (serrated polyps, low-grade or high-grade appendiceal mucinous neoplasms [LAMNs\HAMNs], or mucinous adenocarcinomas of the appendix [MACAs]).

## Case History

A 68-year old Caucasian woman was admitted to the Emergency Department complaining of malaise, dyspnea on exertion, and progressive enlargement of abdominal circumference during the previous 3 months. The patient has no preexisting disease. Because of generalized edema state and mild respiratory failure, diuretic therapy and oxygen therapy was started. At admission to the Internal Medicine ward physical examination reveal normal vital signs, distended abdomen, with peripheral edema (3+). Initial blood tests revealed increased levels of CRP (6.85 mg/dL) and slightly low serum albumin. Abdominal ultrasound showed diffuse presence of abundant corpusculated fluid, so paracentesis was performed, with limited drainage of dense gelatinous ascitic fluid (about 20ml), which cytological analysis revealed amorphous acellular material. Unfortunatelly, MRI of abdomen and pelvis was not performed due to the poor compliance of the patient. Contrast-enhanced CT scan of abdomen was performed, which documented conspicuous ascitic fluid in all recesses; splenomegaly (140 mm); regular liver without lesions. Due to the persistence of the respiratory distress, chest CT scan was performed showed thromboembolism of segmental branches of inferior pulmonary lobes in absence of pleural effusion. The patient thus started anticoagulant therapy with LMWH. Gradual improvement of the patient's respiratory functions was observed, oxygen administration was reduced and contextually peripheral edema disappeared. However, echocardiography was performed, showing marked compression of right atrium due to the abdominal ascitic fluid, with high risk for cardiac tamponade, therefore diuretic therapy was discontinued. Surgical consultation excluded surgical procedure. Despite the procedure extremely laborious, the paracentesis was repeated, with about 600 ml of fluid drained, and a new cytological examination showed amorphous material with mesothelial cells, like as PMP. After 3 weeks from admission, the patient performed CE-MRI of pelvis and abdomen; it showed voluminous right ovarian mass measuring about 25 x 20 cm that compressed liver, bladder and caused dislocation of small intestine and colon. Thus, the patient was transferred to onco-gynecologic department where she underwent surgical intervention. Histology revealed a HAMA staging IVC (pT4a M1c) sec. AJCC UICC 8th edition.

## Discussion

The term PMP is used to describe the clinical syndrome of mucinous appendiceal neoplasm with diffuse peritoneal spread and great mucin production. Appendiceal mucinous neoplasms are rare, with slight female-predominant, prevalence on V-VII decade of life, differentiated between LAMA, HAMA or adenocarcinoma. Consensus on therapeutic strategies is controversial, without scientific evidence about the necessity of cytoreductive surgery (CRS) and heated intra-peritoneal chemotherapy (HIPEC) to treat peritoneal disease after removal of neoplasm. The importance of MRI is paramount to detect appendiceal adenomas, and to differentiate large mucinous neoplasms from other intraperitoneal and retroperitoneal pathologies.