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Appendiceal Adenocarcinoma in Pseudomyxoma Peritonei (PMP) - A Case Report

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Abstract **Case Report**

Pseudomyxoma peritonei (PMP), formerly known as gelatinous disease of the peritoneum, is a very rare pathology encompassing a spectrum of lesions characterized by the accumulation of a large amount of mucus accompanied by a variable number of cells in the peritoneal cavity and pelvis, sometimes associated with the presence of peritoneal cell implants. It is considered a tumor process, linked in the vast majority of cases to a mucinous appendiceal tumor. Pseudomyxomas can benefit from a therapeutic approach combining complete cytoreductive surgery and intraperitoneal chemotherapy, which significantly improves the prognosis. The rarity of this entity and the associated therapeutic strategy require specialized care structures and the support of clinical and pathological networks. We report the case of a 71-year-old woman with pseudomyxoma peritonei diagnosed by CT scan.

Keywords: Pseudomyxoma Peritonei (PMP), Appendiceal Tumor, Cytoreductive Surgery, Intraperitoneal Chemotherapy, Mucinous Appendiceal Tumor.

INTRODUCTION

Pseudomyxoma peritonei (PMP), formerly known as gelatinous peritoneal disease, is a very rare pathology with an estimated frequency of 1-2 per million per year in the Western world [1]. PMP encompasses a spectrum of lesions characterized by the accumulation of a large amount of mucus accompanied by a variable quantity of cells in the peritoneal cavity and pelvis, sometimes associated with the presence of peritoneal cell implants [2]. The origin of PMP remains a source of controversy, however, most authors agree to recognize the appendix, the ovary, and even the peritoneum as the starting point of this disease [3, 4].

We report a case of pseudomyxoma peritoneum associated with appendiceal adenocarcinoma.

OBSERVATION

A 71-year-old woman, married, mother of 10 children, and a housewife, had a medical history of hypertension for 10 years, taking amlodipine 5 mg and undergoing subcostal cholecystectomy 10 years ago. The patient was admitted as an emergency patient with right iliac fossa pain with abdominal distension that had been developing for two days prior to the consultation. This pain, with its progressive onset, radiated throughout the abdomen and was associated with nausea. On physical examination, the patient had a general deterioration in condition, blood pressure of 140/90 mm Hg, temperature of 37.3°C, pulse of 82 beats/minute. The abdomen was distended and tender with guarding at the Mac Burney point. The rectal examination revealed tenderness of the pouch of Douglas, especially on the right side. The abdominal CT scan suggested pseudomyxoma peritonei following a ruptured appendiceal mucocele. The blood count showed a leukocyte count of 14,320/mm³ with a neutrophil predominance. The C-reactive protein (CRP) was 143 mg. The preoperative assessment had been completed by a prothrombin level (96%), an activated partial thromboplastin time (32 seconds) and a fasting blood glucose of 1.73 g/l. After a satisfactory pre-anesthetic consultation, an emergency laparotomy was performed in the operating room under general anesthesia. The operative findings were a large peritoneal effusion with a gelatinous appearance, hyperemia of the small bowel loops and a perforated appendicular mucocele with a friable base. Abscessed collections in the right iliac fossa associated with gelatinous debris, gelatinous collections in the right and left subphrenic veins and several modulo-cystic formations at the parietal and visceral levels. We performed a retrograde appendectomy; the appendix was extremely dilated and tense, the site of the cystic formations with perforation at its base. It measured 10

cm long and 6 cm wide **(Figure 1).** A peritoneal fluid sample containing nodules of peritoneal carcinomatosis was taken, followed by aspiration of the effusion and extensive lavage of the peritoneal cavity.

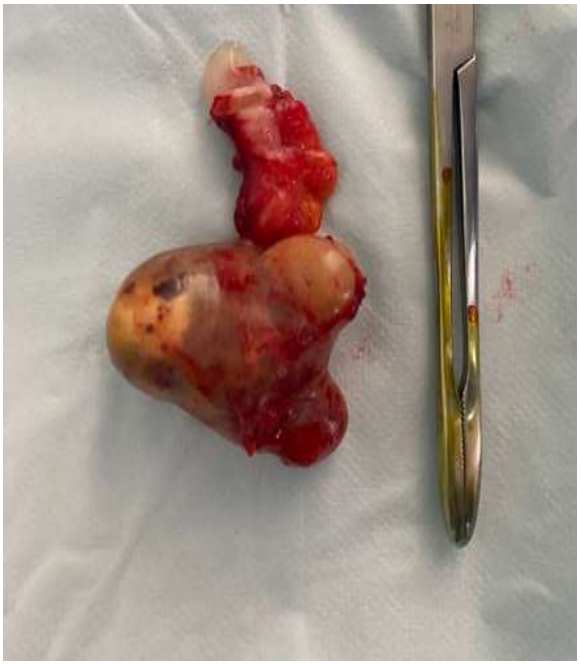

Figure 1: Surgical specimen of appendicular mucocele

The immediate postoperative course was unremarkable. The patient was discharged from the hospital on the third postoperative day. The surgical specimen sent for pathology revealed ulcerated mucinous adenocarcinoma infiltrating the lamina propria and the entire appendix wall, extending to the excisional section, and peritoneal carcinomatosis without neoplastic vascular emboli; cytology contained rare tumor cells. The patient underwent 10 sessions of Xyloda-type chemotherapy.

A follow-up chest, abdominal, and pelvic CT scan was performed, which showed no secondary lesions. A decision was made for a right hemicolectomy, including exploration of a 2*2 carcinomatosis nodule at the level of the culus of the Douglas sac. The procedure consists of a right hemicolectomy with anastomosis.

Conclusion

Pseudomyxoma peritonei is a very rare condition, often discovered late and difficult to treat. The stage of the disease is not always clear; significant gelatinous ascites is often found at laparotomy. The management of this condition also poses a challenge from a diagnostic and therapeutic perspective. Pathological examination of the appendectomy specimen allows for diagnosis. Treatment remains surgical and consists of an appendectomy or even a right hemicolectomy.

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