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Case Report

Peritoneal pseudomyxoma, a Complication of Appendicular Mucocele: A Case Report

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Abstract

Peritoneal pseudomyxoma is a rare disease, classically detected by the discovery of "gelatin" during a laparotomy. Thanks to immunohistochemistry, it is now accepted that its origin is mainly appendicular and not ovarian. It is a "borderline" malignant pathology due to its inevitable persistence and progression in the absence of appropriate therapeutic management: combination of cytoreduction surgery and intraperitoneal perioperative chemotherapy (intraperitoneal chemohyperthermia and/or immediate postoperative intraperitoneal chemotherapy), within specialized centers. Its main prognostic factors are dominated by the importance of the surgical history, the radicality of the cytoreduction and especially by the histopathological grade. We report a case of peritoneal pseudomyxoma detected at the advanced stage whose origin is a ruptured appendicular mucocele disseminated in the peritoneal cavity.

INTRODUCTION

Peritoneal pseudomyxoma or gelatinous disease of the peritoneum is a rare disease defined by the presence of an abundant gelatinous substance in the abdomen secondary to the rupture of an appendicular mucinous lesion whose cystic macroscopic appearance is mucocele.

Carl Rokitanski was the first to describe an appendicular mucocele in 1842 [1]. Subsequently, in 1884, Werth coined the term PMP [2] in relation to an ovarian tumor. In 1937, Robert Michaelis Von Olshausen, a German gynecologist, hypothesized that epithelial cells from the lining of a ruptured appendicular cyst took root in the peritoneal cavity and continued to secrete gelatinous material leading to PMP [1].

Our observation presents a case of peritoneal pseudomyxoma detected at the advanced stage whose origin is a ruptured appendicular mucocele disseminated in the peritoneal cavity.

OBSERVATION

This is a 69-year-old patient with a history of type II diabetes on insulin who was referred by her treating physician for diffuse abdominal pain at the point of departure of IDF associated with progressive abdominal distension over the past three months.

Clinical examination revealed a patient who was hemodynamically and respiratoryly stable, with a distended abdomen on inspection and a palpable mass at the level of the FID with multiple diffuse, hard and mildly painful parietal nodules and dullness.

Faced with this clinical picture, an abdominal CT scan with injection of the contrast medium was performed objectifying multiple intraperitoneal tissue nodules in cake above and under mesocolic associated with an effusion of medium abundance with hepatic and splenic impressions achieving an appearance suggestive of peritoneal carcinomatosis (Figures 1-3).

On the basis of this clinical and radiological presentation, the case was staffed at the Multidisciplinary consultation meeting and the diagnosis was ruptured and complicated by peritoneal pseudomyxoma. An ultrasound-guided biopsy of the peritoneal nodules was performed to confirm the diagnosis of mucinous cystadenocarcinoma.

The Multidisciplinary consultation meeting decision was to do Hyperthermic Intraperitoneal Chemotherapy (CHIP).

Our patient was referred to a national cancer center in Rabat for further treatment (CHIP) since it is not applicable in our structure.

DISCUSSION

The Appendicular Mucocele (AD) is a distended, mucus-filled appendix in the shape of a sac. It is observed in 0.2-0.7%

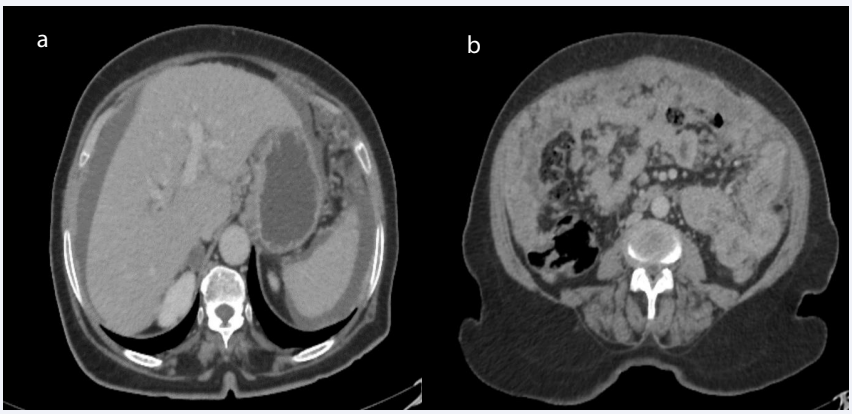

Figure 1 a). Perihepatic and perisplenic effusion of medium abundance.
b). Nodular peritoneal thickening with epiploic cake suggestive of peritoneal carcinomatosis.

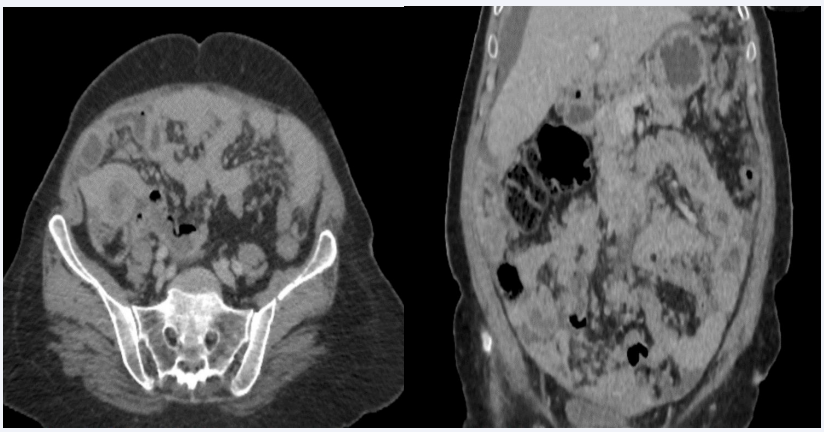

Figure 2 Multiple intraperitoneal tissue nodules above and below mesocoliaus associated with ascites of moderate abundance suggestive of diffuse peritoneal carinosis in relation to ruptured appendicular mucocele.

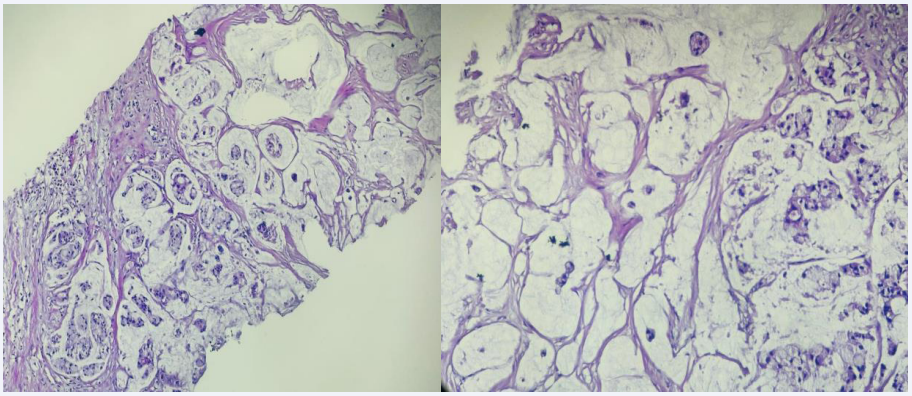

Figure 3 Carcinomatous tumor proliferation arranged in tubes surrounded by puddles of mucin. Tumor cells show cytonuclear atypia, nuclei are large, irregular, hyperchromatic and nucleolated. Figures of mitosis are noted.

of appendectomies [2]. This distension may or may not be of tumor origin. It may be a retentional cyst following obstruction by stercoris, inflammatory stenosis, or extrinsic compression.

Other causes include epithelial villous hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma. In half of the cases, it is revealed by chronic pain of the right iliac fossa. Its most formidable complication is its rupture in the peritoneum (5 to 15% of cases): pseudoperitoneal myxoma [3].

Peritoneal Pseudomyxoma (PMP) is a rare peritoneal malignancy, most often resulting from a perforated epithelial tumor of the appendix. Its incidence is estimated at one to three per million inhabitants per year [3]. PMP, also known as "jelly belly," is characterized by diffuse progressive mucinous ascites.

Its clinical presentation is variable and non-specific. The clinical presentation may manifest as acute or chronic abdominal pain localized to the FID or by a palpable mass, but most often it is asymptomatic, discovered incidentally on imaging or during a complication [4].

Also known as gelatinous peritoneal disease, it includes a spectrum of peritoneal lesions characterized by the accumulation of mucin that can be associated with a variable amount of more or less differentiated epithelial cells [5].

Complete cytoreduction surgery followed by CHIP is the reference treatment for peritoneal pseudomyxoma, recommended by a consensus of international experts in 2008 [6]. The aim of this treatment is to completely remove the peritoneal disease; only the areas affected by the disease are resected according to the recommendations of Jacquet and Sugarbaker [7]. These are usually extensive operations, requiring numerous organ resections, with an average operating time of between 8 and 10 hours.

The principle of CHIP is based on:

- complete resection of all visible peritoneal lesions;
- combined with a hyperthermic bath of intraperitoneal chemotherapy.

PATHOPHYSIOLOGY

The pathophysiology of peritoneal pseudomyxoma explains its topography [8]. In fact, at the early stage, the gelatinous substance located in the mucocele distributes locally to the rupture site and carries out a proximity inoculation. This gelatin contains mucin or even epithelial cells, more or less atypical, which will continue to proliferate and produce mucin in the peritoneum, first locally and then throughout the abdominal cavity [9].

This diffusion in the peritoneum results in a true mucinous ascites with multiple implantations of mucinous and epithelial deposits on the peritoneum. Retroperitoneum may be affected in retrocecal appendage or during surgical inoculation [10]. Peritoneal dissemination of mucinous lesions occurs by redistribution [9].

RADIOLOGICAL PRESENTATION

The peritoneal pseudomyxoma is studied according to the different imaging modalities On ultrasound, it takes on a hypoechoic appearance, associated with hyperechoic septa. Its gelatinous character makes it not very mobile unlike other ascites [11]. On CT scan [12], it appears heterogeneous, rather hypo-dense in the form of lobules whose septa are enhanced by iodinated contrast medium and associated with curvilinear calcifications. The phenomena of peritoneal distribution, proximity and redistribution guide its location. The sites of mesenteric folds as well as the areas of attachment of the intestinal surfaces to the retroperitoneum should be carefully explored. In particular, a thickening of the wall of these loops is sought. Its good spatial resolution allows for good visualization of peritoneal nodules, and images of impressions, scalloping, liver and splenic. The infiltration of the greater omentum appears rather hyperdense and nodular, Later, the epiploic cake appears as a mass of tissue density with polylobed and irregular contours.

HISTOLOGY

Histologically, B. Ronnett's classification distinguishes three main histological types of peritoneal pseudomyxoma, based on survival data from a series of patients [13]

- Disseminated peritoneal adenomucinosis: Peritoneal lesions consist of puddles of abundant extracellular mucin without or with few, minimally or moderately atypical, mucinous epithelial cells and low mitotic activity. The initial lesion is a mucinous adenoma of the appendix.
- Peritoneal mucinous carcinomatosis: peritoneal lesions include extracellular mucin associated with more abundant epithelial clusters, exhibiting cytoarchitectural characteristics of carcinoma with high mitotic activity. The initial lesion is appendicular mucinous adenocarcinoma [14]. Peritoneal mucinous carcinomatosis with intermediate appearances, CMP-I or with discordant aspects, CMP-D:

For CMP-I it is a combination of adenomucinosis lesions and rare areas of well-differentiated adenocarcinoma, the initial lesion being mucinous adenocarcinoma of the appendix with associated adenoma lesion.

For CMP-D, these are peritoneal lesions of mucinous adenocarcinoma, with the initial appendicular lesion being a mucinous adenoma or intramucosal adenocarcinoma without true invasive carcinoma.

The prognosis of the disease is directly correlated with the histological type. Disseminated peritoneal adenomucinosis, AMPD of relatively benign course, is accompanied by a survival greater than 80% at 10 years. Survival drops to less than 60% at 3 years for intermediate CMP-I forms. For malignant forms

of mucinous peritoneal carcinomatosis CMP, the evolution is serious with less than 10% survival at 3 years [13].

Treatment

There are essentially two main types of surgical management of PMP: multiple surgical "debulking" and Cytoreduction Surgery (CRS) with perioperative intraperitoneal chemotherapy: hyperthermic intraperitoneal chemotherapy (HIPC) with or without Immediate Postoperative Intraperitoneal Chemotherapy (IPPIC).

Surgical debulking

The goal of surgical debulking is to remove as much gelatin and tumor formations as possible by dissection generally limited to a right hemicolectomy, a partial resection of the greater omentum, and for women, a hysterectomy with bilateral salpingo-oophorectomy.

As this "debulking" is more often incomplete, the disease persists and leads irremediably to death. Symptomatic recurrences in the form of bowel obstruction, abdominal pain or abdominal distension are treated with further debulking.

Each reoperation becomes more difficult (due to the numerous adhesions), more ineffective (due to the tumor cell trap phenomenon) and more at risk of complications such as enterotomies, digestive fistula, postoperative peritonitis, etc [15].

Cytoreduction surgery and perioperative intraperitoneal chemotherapy

Most centers specializing in the therapeutic management of PMP recommend CRC plus CHIP with or without CIPPI [16-18].

CRC consists of resecting any visible tumor formation by performing peritonectomy procedures. The most frequently required resections are: resection of the large and minor omentum, peritonectomy of the right and left diaphragmatic cupolas, anterior peritonectomy, peritonectomy of the parietal peritoneum of the cul de sac of Douglas (with or without resection of the rectosigmoid), splenectomy, cholecystectomy, and antrectomy.

Not all of these procedures are carried out systematically. They are only performed in the event of tumor invasion of these organs. It should be noted that in addition to these surgical procedures, a right hemicolectomy and a hysterectomy with bilateral salpingo-oophorectomy in women are recommended. The goal of CRC is to resect any visible tumor leaving only tumor nodules less than 2.5 mm in diameter.

At the end of the surgical procedure, a CHIP is then started for a duration varying from 30 to 90 minutes [16-18]. The chemotherapeutic agents used in CHIP vary by centre: most commonly mitomycin C and cisplatin [19-23] and oxaliplatin. Some centers additionally administer CIPPI for five days with 5-FU [19].

CONCLUSION

Significant progress has been made in recent years in the understanding of this rare disease that is Pseudoperitoneal Myxoma. Despite its name, PMP remains a lethal disease in the short or long term and must be treated appropriately. CRC with CHIP appears to be the treatment recommended and preferred by most experienced centers. A rigorous selection of surgical candidates by a surgical team experienced in the management of such patients is the guarantee of greater success.

Conflicts of Interest

The Authors declare no conflicts of interest in relation to this study.

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