



Pseudomyxoma Peritonei with Metastatic Ovarian Tumor in a 28-Years-Old Patient: A Case Report with Review of the Literature

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Abstract

Pseudo Myxoma Peritonei (PMP) or gelatinous disease of the peritoneum is a very rare tumor that originates from a ruptured appendiceal mucocele in the abdomen. Other uncommon origins are described in the literature like ovaries and peritoneum. Although the condition occurs in both sexes, but it affects especially women between the age of 50 and 70. We hereby report the case of a 28 years old French woman diagnosed with a suspicious solid ovarian mass of 62.8 mm × 99.7 mm, associated with peritoneal effusion. The patient underwent one week later a diagnostic laparoscopy with left salpingo-oophorectomy revealing a left ovarian tumor of 10 cm associated with diffuse gelatinous ascite. The microscopic exam of the tumour showed a low-grade metastatic ovarian mucinous carcinoma associated with a pseudomyxoma peritonei with the appendix being the most probable origin.

Keywords: Appendix; Pseudomyxoma peritonei; Ovarian cancer

Introduction

Pseudo Myxoma Peritonei (PMP) is a very rare neoplasm, with poor prognosis. There is only 45 cases reported in the English Literature and it is known to affect 2/1000000 persons per year [1,2]. The course of the disease is characterized by the production of mucine into the abdomen leading to a gelatinous obstructive ascite or what it is called “jelly belly” appearances [1]. PMP has different origins: more than 80% arises from the appendix. However, Primary ovarian mucinous carcinoma presenting as PMP has been reported too [2,3], with a low stage and low grade at time of diagnosis in the majority of women. PMP has a protracted therapeutic pathway with combined repeated cytoreductive surgeries and Intra Peritoneal Chemotherapy (IPC) [4,5]. Below, we report an extremely rare case of a 28 year-old woman who developed a Pseudomyxoma peritonei originated from the appendix with an ovarian metastatic tumor.

Case Presentation

A 28 years old nulliparous woman presented to the Emergency Room for persistent diffuse pelvic pain Ten days after sexual intercourse. There is no previous personal medical or gynaecological history. She noted two diarrheal episodes that day with pollakiuria. The transvaginal ultrasound showed a left atypical solid ovarian mass of 62.8 mm × 99.7 mm, with heterogeneous peritoneal effusion. A pelvic MRI and tumor markers were ordered showing a slight increase of CEA, normal Ca 19-9, Ca 125 and Inhibin B with a primitive left septated ovarian mass suspicious of malignancy with peritoneal and pelvic effusion (Figure1).

A exploratory laparoscopy was done one week later revealing a peritoneal cavity filled with 400 ml of a viscous substance similar to gelatin with a left ovarian tumor of 10 cm (Figure 2 and 3). A left salpingo-oophorectomy was done; the appendix was not seen due to its retrocecal position and the inflammatory magma over it. The histopathological examination of the tumour reported a low-grade metastatic ovarian mucinous carcinoma associated with a pseudomyxoma peritonei most probably originating from the appendix. Later on, an immunohistochemical study revealed a strong expression of CK20 and absence of CK7. The patient is actually undergoing a cycle of hormone stimulation followed by a cryopreservation of her oocytes. Then in a second time, a cytoreductive surgery and a Hyperthermic Intraperitoneal Chemotherapy (HIPEC) will be done.

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Figure 1: Pelvic MRI: Sagittal T2-weighted image, showing an intraperitoneal tumor with tissular and cystic signal. The tumor is separated from the uterus and surrounded by pelvic effusion.



Figure 2: Immediate findings at laparoscopy showing "jelly belly" appearances.

Discussion

Pseudo Myxoma Peritonei (PMP) is a very rare entity consisting of gelatinous ascites due to the implantation of mucine on the peritoneal surfaces. In 1842, the disease was thought to originate from the ovaries when Rokitansky and then Cruveilhier were the first to describe a gelatinous degeneration in the peritoneal cavity [6]. In 1871, the disease was qualified by Pean as "gelatinous disease of the peritoneum" [6]. Then, in 1884, Werth described the rupture of an ovarian cyst with a gelatinous product [6], followed by Frankel on 1901 who mentioned a rupture of an appendicular tumor [6].

Pseudo Myxoma Peritonei (PMP) has an incidence of 2/1000000/year. Usually, the main cause is an appendicular mucocele with other rare primary sites have been also reported in the literature like the ovaries, uterus, urachus, colon, stomach, pancreas and common bile duct [1,7]. The disease is found more frequently in women than man (male: female ratio=9:11) and affects the female in general after age of 50 [7]. There are no reported cases of PMP during twenties, making our case the first in the literature to describe the occurrence of the disease at this age [1].

The path physiology of the disease is explained by the hypersecretion of mucine that leads to an overdilatation of the appendix followed by a rupture and dissemination of the mucus to the whole abdominal cavity [8].

Furthermore, studies have shown that multiple enteric bacteria play an important role in the progression of the disease (MUC2 and

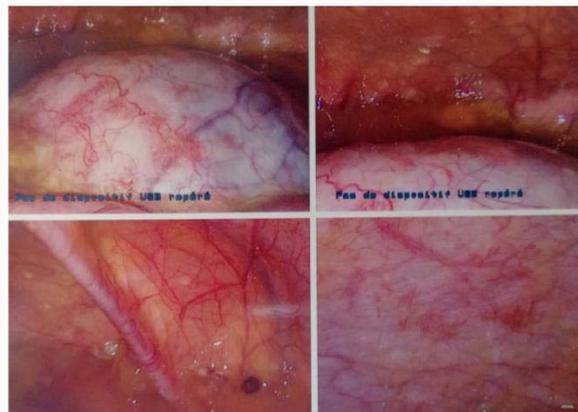


Figure 3: Intraoperative findings showing the left ovarian tumor with the diffuse gelatinous ascite around.

MUC5AC expression in disseminated peritoneal adenomucinosis and peritoneal mucinous carcinomatosis) [9,10]. The gelatin adheres to all the organs covered by the parietal peritoneum especially the omentum as shown in our case, making the surgical treatment very aggressive.

There is no pathognomonic sign for the diagnosis of PMP, but symptoms are variant and goes from a simple abdominal pain and transit disorders as seen in our patient's case, to signs of subocclusion. The imagery (endovaginal ultrasounds, RMI, and TDM) and exploratory laparoscopy stays the gold standard for the diagnosis of PMP as seen in the published literature.

Tumor markers (CEA, CA19.9, CA125) are not useful for the diagnosis of the disease. In our case, all the tumor makers were normal except of the CA125 that was slightly increased and was not specific also.

The mainstay of the treatment is cytoreductive surgery and Hyperthermic Intraperitoneal Chemotherapy (HIPEC). The surgical approach depends on the size of the lesions. But laparotomy remains the preferred method. During the operation, the appendix must obligatorily be removed; sometimes even a right hemicolectomy and hysterectomy with bilateral adnexectomy can be performed.

The prognosis is really improved by the IPC and more recently, doctors in specialized centers resort to the Pressurized Intraperitoneal Aerosol Chemotherapy (PIPAC) to defeat the pharmacokinetic limitations of intraperitoneal chemotherapy. Studies have shown that the pressured aerosol increases the drug uptake by the tumor cells with fewer complications compared to the use of IPC [11,12].

The recurrence rate of the disease is very high because the removal of the entire peritoneum and all the mesos is really impossible. The disease remains microscopic and no exam can detect a peritoneal lesion pre and post operatively. Tumor blood markers are ineffective and do not evaluate the chemotherapy efficiency. Reported cases showed a morbidity rate of 24% while the mortality is estimated at 2% [13,14].

Conclusion

Pseudo Myxoma Peritonei (PMP) is a very rare condition. The definitive diagnosis relies on laparoscopic findings combined with histopathology and immunochemical exam. As revealed in the few published cases and recently in our case, the disease can affect woman

at any age. Consequently, PMP should be considered as a differential diagnosis in any female presenting with an ovarian tumor suspicious for malignancy associated with peritoneal effusion.

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