Global Academic Journal of Medical Sciences

Available online at www.gajrc.com
DOI: 10.36348/gajms.2023.v05i03.002

OPEN ACCESS

ISSN: 2706-9036 (P)
ISSN: 2707-2533 (O)

Case Report

Peritoneal Pseudomyxoma Secondary to a Mucinous Tumor of the Ovary: About a Case

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Article History
Received: 17.04.2023
Accepted: 13.05.2023
Published: 18.05.2023

Abstract: Peritoneal pseudomyxoma, also called gelatinous disease of the peritoneum is a rare entity that corresponds to diffuse peritoneal infiltration, consisting of mucinous ascites and multifocal mucinous epithelial implants. Its prevalence is 2 cases per million populations [1]. Peritoneal pseudomyxoma may remain asymptomatic, discovered during laparotomy. CT scans are the most specific diagnostic tool. The appendicular origin of gelatinous ascites is the most common and represents 90% of causes. Mucinous tumours of the ovary are a very rare cause. We report the case of a 55- year-old woman with a peritoneal pseudomyxoma of ovarian origin [2].

Keywords: Peritoneal pseudomyxoma, ascites.

INTRODUCTION

Pseudomyxoma peritoneal corresponds to an anatomo-clinical entity without prejudging its origin, characterized by an effusion of variable abundance, of viscous or mucinous appearance, in the peritoneal cavity (synonyms: gelatinous ascites, gelatinous disease of the peritoneum) associated or not with epithelial cells, whose degree of malignancy is variable. The origin is appendicular in at least 90% of cases, secondary to a mucinous tumor of the appendix ruptured into free peritoneum. Mucinous tumours of the ovary are a very rare cause. We report the case of a 55-year-old woman with a peritoneal pseudomyxoma of ovarian origin.

PATIENT AND OBSERVATION

Patient Information:

This is a 55-year-old woman, with no pathological history. The onset of the symptom's dates to 2 months with the installation of reduced hypogastric pain, associated with homogeneous distension without cessation of matter or gas without other associated digestive or extra-digestive signs. Moreover, there were no general signs in favor of an infectious syndrome. The clinical evolution was marked by an alteration in the general condition of the patient, which motivated her consultation in our Department of Gastro-entero-hepatology at CHU Mohammed VI in Marrakech.

Clinical Result:

The general physical examination revealed a conscious patient, hemodynamically and respiratory stable, a WHO of 2, a BMI of 28.9 in the presence of ascites, white lower limb edema, soft taking the cups and arriving to legs. Abdominal examination showed homogeneous abdominal distension and diffuse dullness of the abdomen, with collateral venous circulation of the flanks, with no palpable mass. The rest of the clinical examination is unremarkable.

Diagnostic Approach:

The biological assessment was normal apart from a hyperleukocytosis at 13100 with PNN

predominance. The CEA assay was at 15N, CA125: 19.5N and for CA19.9 was at 102.8N. Abdominal ultrasound showed heterogeneous ascites of great abundance with hyperechoic images on the abdominal wall. Associated with voluminous retroperitoneal cystic formations of the right flank measuring 10*12 cms with an infiltration of the vicinity. Abdominal computed tomography (CT) revealed a cystic formation of the multi-partitioned FID with abundant dense compressive peritoneal effusion, first evoking a peritoneal pseudomyxoma.

Therapeutic Intervention and Follow-Up:

The peritoneal effusion was dense, pushing back the digestive structures inside with hepatosplenic scalloping. Due to a diagnosed doubt, the decision for an exploratory laparotomy was made. Surgical exploration revealed abundant gelatinous ascites and a large cystic tumor at the expense of the left ovary with whitish nodules disseminated on the parietal peritoneum. Furthermore, there was no appendicular lesion, nor of the digestive tract or other visceral lesions. The ascites (about 20 liters) was evacuated with left adnexectomy removing the cystic mass. An appendectomy was performed with resection of the large peritoneal implants. The postoperative course was simple. The anatomopathological study of the surgical specimen was in favor of a borderline mucinous tumor of the ovary.

DISCUSSION

Peritoneal pseudomyxoma or gelatinous disease is a rare entity, first described by R. Wyerth in 1884. Its incidence is estimated at 2 cases per million inhabitants per year and 2 cases per 10,000 laparotomies [1]. The average age of onset is 46 years with a preferential attack of women with a sex ratio of one man for two women. Our case was a 55-year-old woman. Peritoneal pseudomyxoma corresponds to diffuse peritoneal involvement, composed of mucinous ascites and multifocal mucinous epithelial implants, the fundamental histological element is the presence of extracellular mucin in the peritoneal cavity, which may be associated with mucinous epithelial cells, more or less well differentiated [2, 3].

Clinically, the symptomatology is non-specific. Peritoneal pseudomyxoma (PMP) can be revealed by late clinical signs with a general condition that is often not very altered. Thus, the diagnosis is rarely made before laparotomy [4]. The revealing signs are numerous, dominated by the progressive and isolated increase in abdominal volume and pain. The other functional signs are mainly related to the impact of the peritoneal gelatinous disease on the digestive tract and/or the urinary tract [5].

A study by Esquivel and Sugarbaker on 217 patients found that the clinical presentations were 27% suspected appendicitis, 23% progressive abdominal distension and 14% revealing inguinal hernia. Sometimes, the PMP is asymptomatic with a fortuitous discovery on imaging examination (ultrasound, CT, etc.) or during a laparotomy [6]. Surgery was long considered the gold standard for diagnosis according to Walensky *et al.,* Our patient had atypical hypogastric abdominal pain with diffuse abdominal distension.

On the radiological level, the radiograph of the abdomen without preparation (ASP) can show a predominant opacity on one side of the abdomen which is significant if it is associated with curvilinear calcifications marrying the periphery of the opacity [7]. In ultrasound as in CT, the diagnosis of pseudomyxoma is based on the identification of three lesions: mucinous ascites and its characteristics, nodular peritoneal implants if they are visible, and the primary tumor, which is only exceptionally visualized.

The mucinous ascites of pseudomyxoma is heterogeneous and hypodense. It may be partitioned and contain fine curvilinear calcifications. We can find an extrinsic compression effect on the liver or the spleen “scalloping” and a repression of the hollow organs [8]. In our patient, CT showed the same radiological signs found in the literature, without being able to locate the primary tumor responsible for the ascites. CT is particularly useful for monitoring the evolution of the disease and detecting a possible recurrence or a complication (occlusion, abscess, ureteral compression with dilation of excretory cavities).

The tumor markers requested in this context are most often ACE, CA 19-9 and CA125. A blood level of ACE has been described in PGM associated with both malignant and benign tumors. These markers, and especially CA 19-9, can be useful in monitoring the disease and diagnosing recurrences [9].

There has long been a debate about the origin of PMP: appendicular or ovarian. Such a controversy existed because a simultaneous attack of these two locations was found in most women. Thanks to immunohistochemistry and molecular engineering, it is accepted that the origin is appendicular in approximately 90% of cases [10]. In 2002, Cornell *et al.,* studied the expression of the MCU 2 receptor on peritoneal mucinous cells, their

positivity confirmed that the primary tumor was an appendicular mucinous tumor.

According to Ronnett, Young and Prayson, ovarian mucinous tumors of low malignancy discovered during PGM are almost always secondary localizations of an appendicular tumor, especially when they are bilateral (in 32 to 80% of cases). When they are unilateral, they can be primary tumors responsible for PMP and are most often located on the right. They are large and often have implants or nodules on their surface [11]. Our patient had a large mucinous tumor of the left ovary without appendicular involvement.

This case is exceptionally found in the literature. There are essentially two types of treatment for PMP: multiple surgical debulking and cytoreductive surgery with perioperative intraperitoneal chemotherapy: hyperthermic intraperitoneal chemotherapy with or without immediate postoperative intraperitoneal chemotherapy [11]. The goal of "debulking" is to remove as much gelatin and tumor formations as possible. Symptomatic recurrences present in the form of intestinal obstruction, abdominal pain or abdominal distension, and are treated by a new "debulking" [12]. Each reoperation becomes more and more difficult.

Cytoreduction surgery (CCR) consists of resecting any visible tumor formation by performing peritonectomy procedures. Right hemicolectomy and hysterectomy with bilateral salpingo-oophorectomy in women are recommended. The goal of CCR is to resect any visible tumor leaving only tumor nodules less than 2.5mm in diameter, so that perioperative intraperitoneal chemotherapy is effective. In our patient, we opted for the "debulking" technique with evacuation of the maximum amount of gelatin without hyperthermic intraperitoneal chemotherapy because of its unavailability in our hospital.

CONCLUSION

Peritoneal pseudomyxoma is most often of appendicular origin, but an ovarian origin remains probable. In the absence of early treatment, the prognosis of this disease remains poor. Improving prognosis requires early diagnosis of mucinous tumors even before the stage of ascites.

COMPETING INTERESTS

The authors declare no conflict of interest.

Authors' Contributions

All authors participated in writing and editing the article. All authors have read and approved the final version of the manuscript.

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