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Case Report

Gelatinous peritoneal disease secondary to appendiceal mucinous adenocarcinoma: a case study and review of the literature

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Abstract

Pseudomyxoma peritonei also referred to as gelatinous ascites, is a rare disorder, described for the first time by R. Wyerth in 1884. It is characterized by the presence of mucous disseminated throughout the peritoneal cavity generally arising from the rupture of an appendicular mucocele. Pseudomyxoma peritonei can be asymptomatic, discovered during a laparotomy. The most common symptom is abdominal distension associated with diffuse abdominal pain. An abdominal CT scan is the most specific diagnostic tool. It shows pathognomonic signs of gelatinous ascites. Mucinous neoplasms of the appendix are the most frequent cause of pseudomyxoma peritonei accounting for 90% of cases. Pseudomyxoma peritonei needs to be considered as a borderline malignant disease because of its inevitable persistence and progression without an adapted therapeutic approach: cytoreductive surgery combined with perioperative intraperitoneal chemotherapy in specialized centers. The principal prognostic factors are the prior surgical history, the completeness of cytoreduction and especially the histopathologic grade.

We report the case of pseudomyxoma peritonei secondary to appendiceal mucinous adenocarcinoma.

Introduction

Pseudomyxoma Peritonei (PMP) is a rare tumor characterized by the dissemination of mucus in the peritoneal cavity excreted by adenomatous tumor cells that have invaded the peritoneum [1]. Its incidence is 2 cases per million inhabitants with a female predominance. This invasion is the last step in a process that begins with the rupture of a localized mucinous tumor. The tumors at the origin of PMP are generally appendiceal mucoceles, more rarely ovarian mucinous cystadenoma and cystadenocarcinoma [2,3]. Its scarcity and its unusual clinical manifestations remain diagnostic and therapeutic challenges. The pseudomyxoma peritonei can remain asymptomatic, discovered during a laparotomy. The CT scan is the most specific diagnostic means. Treatment is based on the combination of cytoreduction surgery and Hyperthermic Intraperitoneal Chemotherapy (HIPEC). Improving the prognosis requires early diagnosis and appropriate therapeutic management [4,5].

We report the case of a pseudomyxoma peritonei on appendiceal mucinous adenocarcinoma revealed by huge gelatinous ascites responsible for pelvic organ prolapse and diaphragmatic injury.

Case report

We report the case of a 65-year-old woman, with no pathological history. The beginning of the symptomatology goes back 4 months by the installation of a rapidly progressive abdominal distension with the impaired general condition. The physical exam found a huge distended abdomen with a belly circumference of 120cm (Figure 1), with diffuse dullness related to abundant ascites, associated with stage 3 genital prolapse, the lymph node examination was normal. Paracentesis found a gelatinous viscous liquid with a protein level of 53g/l. A thoraco-abdominal-pelvic CT showed a dense peritoneal effusion occupying all the compartments of the abdomen and the pelvic floor, responsible for scalloping on the hepatic and

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splenic parenchyma and repression of the intestine loops. The appendix appeared normal and the ovaries were not explored with no deep lymph nodes (Figure 2). Regarding tumor markers, Carcinoembryonic Antigen (CEA) was very high at 705 ng/ml, Carbohydrate Antigen (CA) 19-9 and CA 125 were normal.

The decision was to perform an exploratory laparotomy. Surgical exploration revealed copious *amounts* of mucinous *ascites* with the presence of three masses with gelatinous contents at the appendicular level, left ovary, and left fallopian tube and the presence of a 6cm diaphragmatic rupture (Figures 3,4). The surgical gesture consisted of the aspiration of 18L of gelatinous liquid, an appendectomy carrying away the first mass, and a left adnexectomy carrying away the two masses (ovary + left fallopian tube) with the suture of the diaphragmatic injury. The postoperative course was simple. The anatomopathological study of the surgical specimen was in favor of mucinous adenocarcinoma, in which the appendicular origin is the most probable classified G2 of AJCC pathological stage, with secondary involvement of the fallopian tube and left ovary and the peritoneum at the immunohistochemical study. The decision was to perform a right hemicolectomy and refer the patient for hyperthermic intraperitoneal chemotherapy. Unfortunately, the patient has been lost to follow-up since the surgery.

Figure 1: Enormous abdominal distension in relation with an abundant gelatinous ascites.

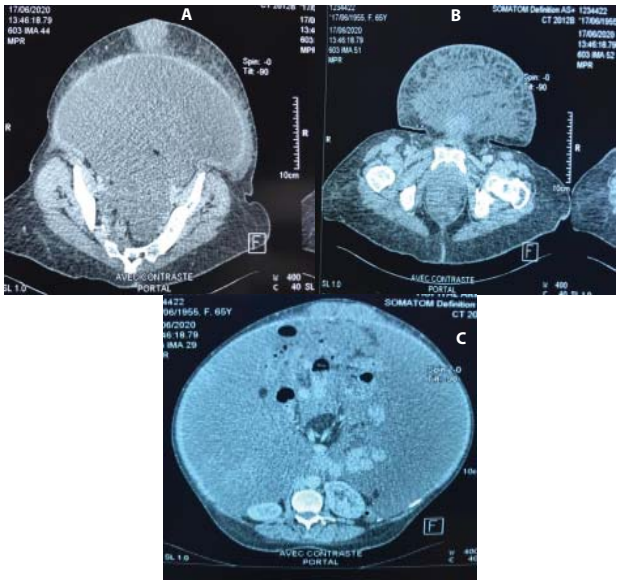

Figure 2: (A,B,C): CT scans revealing dense peritoneal effusion occupying all compartments of the abdomen and the pelvic floor, responsible for scalloping of the hepatic and splenic parenchyma and repression of the intestinal loops.

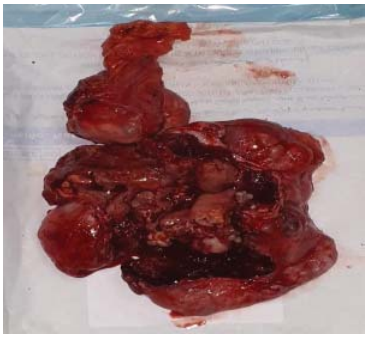

Figure 3: Surgical exploration showing gelatinous ascites..

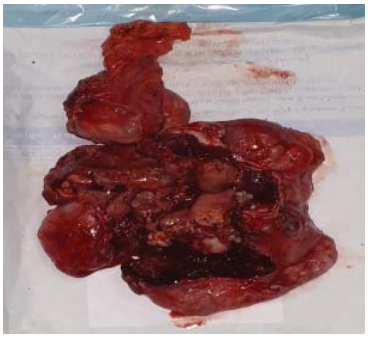

Figure 4: Operating specimen of appendectomy with left adnexectomy.

Discussion

Pseudomyxoma Peritonei (PMP) or gelatinous peritoneal disease corresponds to a rare anatomo-clinical entity. Its incidence is estimated at 2 cases per million inhabitants per year and 2 cases per 10,000 laparotomies [6]. The average age of onset is 46 years with female predominance with a sex ratio of one man for two women [3-7].

Without prejudging its origin, the PMP is characterized by an effusion of variable abundance, of viscous or mucinous aspect in the peritoneal cavity, corresponding to multifocal mucinous epithelial implants, the fundamental histological element is the presence of extracellular mucin in the peritoneal cavity, which may be associated with more or less well differentiated mucinous epithelial cells whose degree of malignancy is variable [8,9]. First described by R. Wyerth in 1884, it described the accumulation of extracellular mucin in the peritoneal cavity with the presence of an ovarian mucinous tumor [10]. Then in 1901, Frankel associated it with the presence of an appendiceal mucocele [11].

There has long been a debate about the origin of PMP: appendiceal 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 appendiceal in approximately 90% of cases, secondary to a mucinous tumor of the appendix (LAMN) ruptured into the peritoneum [7,12]. Other causes of PMP can be an ovarian tumor (teratoma) [13], a tumor of the cervix [14], a tumor of the urachus, an appendicular, colonic, or pancreatic adenocarcinoma (cystadenoma, cystadenocarcinoma and rare cases papillary and mucinous

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intraductal tumor (TIPMP)). It has been demonstrated through immunohistochemical studies on the CK7, CK20 and HAM-56 antigens that the origin of pseudomyxoma was most often appendiceal and not ovarian [15]. This has recently been proved by molecular biology data concluding that the over-expression of the MUC-2 gene in the pseudomyxoma would be the consequence of the presence of gram-negative bacteria (resulting from the perforation of the appendix). This overexpression is correlated with the density of germs and a poor prognosis [4]. In addition, the study of mutations in the K-ras gene and the loss of alleles at the level of chromosomes 18q, 17p, 5q and 6q are found in pseudomyxoma peritonei and are not found in true borderline tumors of the ovary [3]. Ovarian mucinous tumors, thought to cause pseudomyxoma, can give rise to peritoneal tumor implants called peritoneal carcinomatosis, but not true pseudomyxoma. The only primary ovarian tumors with pseudomyxomatous dissemination would be mature cystic teratomas, probably linked to the existence of a gastrointestinal contingent in these embryonic tumors [16].

Clinically, the symptomatology is non-specific. Pseudomyxoma peritonei (PMP) can be revealed by late clinical signs with the general condition that is often good. Thus the diagnosis is rarely made before laparotomy [17]. The revealing signs are multiple, dominated by the progressive and isolated distended abdomen. The other signs are mainly related to the impact of the peritoneal gelatinous disease on the digestive tract (transit disorders, subocclusive syndromes) and/or the urinary tract [18]. A study by Esquivel and Sugarbaker carried out on 217 patients found that the clinical presentations were 27% suspected appendicitis, 23% progressive abdominal distension and 14% revealing an inguinal hernia [19]. Unfortunately, the discovery of PMP is most often fortuitous and made after a laparotomy for suspected appendicitis or ovarian cancer. The surgical act committed is then most often insufficient and makes the second intervention more difficult. Nevertheless, imaging before any surgery is highly recommended to allow a preoperative diagnosis and to plan the appropriate surgical treatment.

Biology is not useful for diagnosis, but tumor markers (CEA, CA 19.9, CA125) are used to detect early recurrences. They are high in most patients and useful for evaluating the effectiveness of chemotherapy. When an increase in one of the three markers is observed, it is often earlier than the appearance of morphological abnormalities, especially when they were increased preoperatively. This justifies their systematic dosage during monitoring. Nevertheless, 20% of PMPs do not lead to an increase in markers. This dosage is therefore not sufficient [19,20].

The radiological diagnosis of pseudomyxoma is based on the identification of three lesions: mucinous ascites and their characteristics, nodular peritoneal implants if they are visible, and the primary tumor, which is only exceptionally visualized. The radiological elements suggestive of pseudomyxoma are scalloping of the liver due to hepatic compression by the gelatinous mass, partitioned ascites, and peritoneal effusion accompanied by curvilinear calcifications [2]. It should be noted that this scalloping can be observed even when the volume excreted is very low [21].

Computed Tomography (CT) and magnetic resonance imaging (MRI) of the abdomen is required for the assessment of resectability. 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 complication (occlusion, abscess, ureteral compression with dilation of excretory cavities). The extent of the disease and the possibility of achieving complete cytoreduction remain difficult to assess on the imaging assessment. In particular, one of the limiting factors for complete resection is the diffuse involvement of the small intestine, not visible on imaging examinations. Abdominal exploration by laparoscopy makes it possible to evaluate the possibilities of complete resection and to analyze the hail [7].

The treatment of pseudomyxoma peritonei is not yet standardized and there is no data in the literature to establish clear conclusions. Nevertheless, complete excision by cytoreductive surgery followed by hyperthermic intraperitoneal chemotherapy gives good results in terms of survival, and as such remains considered the "gold standard" treatment [22]. There are essentially two types of treatment for PMP: surgical debulking and cytoreductive surgery with perioperative intraperitoneal chemotherapy: hyperthermic intraperitoneal chemotherapy with or without immediate postoperative intraperitoneal chemotherapy [2]. The goal of "debulking" is to remove as much gelatin and tumor formations as possible. Cytoreductive surgery (CCR) consists of resecting any visible tumor formation by performing peritonectomy procedures [7]. No intraperitoneal recurrence has been reported for peritoneal pseudomyxoma of ovarian origin [9]. Even with radical treatment with complete cytoreductive surgery and HIPEC, approximately 25% of patients relapse, and the risk of recurrence is 3 times higher in the case of high-grade PMP. The majority of recurrences occur during the first 3 years, but some recurrences can be diagnosed very late (10 years), especially for low-grade forms [23].

Conclusion

Pseudomyxoma Peritonei is most often of appendiceal origin, but an ovarian origin remains probable. In the absence of early treatment, the prognosis of this disease remains poor. Significant progress has been made in recent years in understanding this rare disease that is PMP. Despite its name, PMP remains a fatal disease in the short or long term and must be treated appropriately. CCR with HIPEC seems to be the gold standard treatment and is preferred by most experienced centers.

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