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Case Report

Gelatinous Ascites in Gastric Adenocarcinoma

GK Bennani*, I Benelbarhdadi and FZ Ajana

Department of Diseases of the Digestive System, Medicine C, CHU IBN SINA-Souissi Med University V, Rabat, Morocco

***Corresponding Author:** GK Bennani, Department of Diseases of the Digestive System, Medicine C, CHU IBN SINA-Souissi Med University V, Rabat, Morocco.

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Abstract

Gelatinous disease of the peritoneum or pseudo-peritoneal mixing is a very rare condition. Its origin is mainly appendicular (70 - 80%) and not only ovarian but other. Sporadic causes were described in the literature we add an observation of a patient who presents a gelatinous ascites whose etiology found is an adenocarcinoma cardia which to our knowledge has not been reported.

Keywords: *Gelatinous Ascites; Gastric Adenocarcinoma*

Introduction

Gelatinous disease of the peritoneum or pseudo-peritoneal mixing is a very rare condition. Its origin is mainly appendicular (70 - 80%) and not only ovarian but other.

Case Report

It is an observation of Mr. BM aged 65 years, with no pathological history, who was hospitalized in our medical clinic for edemato-ascitic syndrome who took charge with no previous pathology which is taken care of at the, it is from the Ibn Sina University Hospital in Rabat Morocco for evolving since 1 year ago the abdominal examination finds a distended abdomen like an amée, phibian belly with umbilical hernia, palpation finds a checkerboard, abdominal ultrasound showed immobile ascites with septations. Ultrasound-guided puncture confirmed gelatinous nature of ascites.

The biological assessment showed an inflammatory syndrome with accelerated sedimentation rate, hypergammaglobulinemia.

The abdominal CT scan showed significant heterogeneous ascites with a mass effect on the liver with no detectable tumor lesion.

The barium enema was normal with opacification of the appendix which appeared radiologically normal. Colonoscopy was normal oeso gastroduodenal fibroscopy showed an ulcerated polyploid formation located at the level of the cardia and whose anatomopathological results revealed a tubular adenocarcinoma infiltrating the gastric mucosa. The chest CT scan was normal.

At the end of this assessment, a gelatinous ascites associated with an adenocarcinoma of the cardia was retained. The patient was operated on in the surgical clinic after laparotomy, there were large, hard budding formations containing cysts filled with gelatin. The surgery was only palliative and the excision of the cardia tumor could not be performed given the extent of the carcinosis.

The anatomopathological results of the tumor blocks have a peritoneal tissue largely dissociated by pools of mucus comprising flaps of well differentiated glandular epithelium without mitosis. Cytoplasm are abundant with eosinophils and filled with mucus. We therefore concluded with a pseudo grade I peritoneal myxoma of the sugar baker classification.

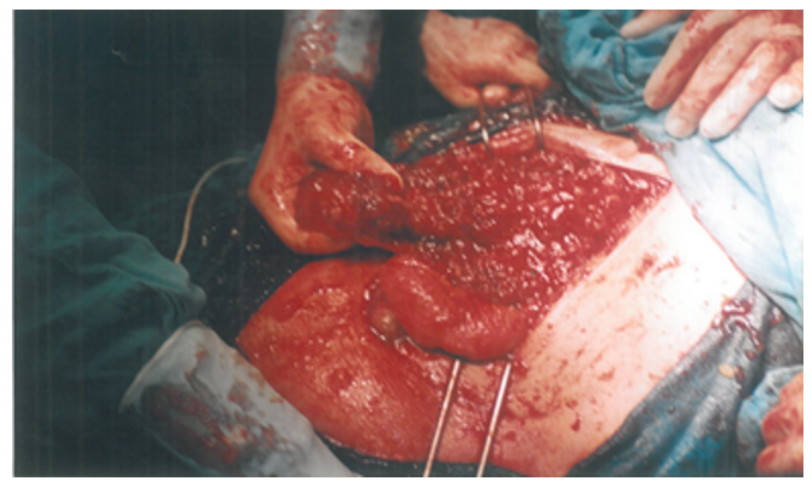

Figure 1: *Macroscopic appearance of gelatinous ascites.*

Discussion

Gelatinous or more precisely mucinous ascites is an essentially clinical term, imprecise, which is why it is currently called pseudo peritoneal myxoma (PP) [1]. The first case was described by Rokitansky in 1842 [1]. In 1884 Wertn described its association with a mucinous carcinoma of the ovary [1]. Franckel in 1901, reported the first case of pseudo peritoneal myxoma associated with an appendix tumor [2].

It is a very rare disease whose frequency is evaluated at 2 per 10,000 laparotomies [3], seeming to interest twice as much the man as the woman [2,3]. Etiopathogenically, gelatinous ascites was formerly considered to be secondary in the majority of cases to an ovarian tumor (45 to 60%) or to a tumor of the vermiform appendix (30%) [4,5]. Bifocal origin (appendix and ovary is frequent [4-6] but recent contributions from immunohistochemistry, genetic analysis by gene amplification (PCR) have concluded that most: pseudo peritoneal myxomas had an origin appendicular, therefore monoclonal [7,8].

Thus, after these molecular biology data, the majority of PP would be of appendicular origin. (70% to 80%), after rupture of either a uni adenoma or a mucinous adenocarcinoma. However, other tumors may be involved: certain mucinous tumors ruptured in the peritoneum, notably borderline tumors of the ovary and mucinous cystic tumors of the pancreas [1]. Other organs or other tumors have been reported in the literature but sporadically: certain carcinomas or colloids of colorectal [9], gastric or pancreatic [10], vesicular or biliary [11], adnexal (tubal) origin [12] and even mammary [13] and pulmonary [14].

Our observation reports a case of PP most probably secondary to cardiac carcinoma which, to our knowledge, has not been described in the literature. Unfortunately, we could not explore the appendix intraoperatively given the extent of the carcinosis but we were able to rule out the possibility of an appendicular tumor on the data of the barium enema where the radiological aspect of the appendix was normal.

Despite the knowledge of these etiopathogenic data, the pathophysiology of gelatinous ascites remains very poorly understood. Several hypotheses have been suggested [3,5]:

- The mechanical theory which would explain the large quantity of mucin in the peritoneum by rupture of the tumor but the mechanism of recurrence after removal of the primary tumor remains obscure.
- The theory of metaplasia of peritoneal cells in the presence of mucin.
- Metastatic theory or the theory of attenuated malignancy where PP is secondary to a malignant tumor.

On the histological level, a recent and interesting classification has distinguished two entities [15]:

- Disseminated peritoneal mucinous adenomatosis (AMPD): Characterized by peritoneal lesions made of abundant mucin containing an scarce epithelium and devoid of mitosis or cellular atypia.
- Peritoneal mucinous carcinosis (CPM): Characterized by peritoneal lesions made of abundant mucin but with a very abundant epithelium and architecture and cytology of carcinoma. Intermediate forms between these two forms can be seen.

AMPD is the most common (60%), most often secondary to a mucinous adenoma of the appendix.

Sugarbaker., *et al*. [16] proposed a grading from 0 to 3:

- Grade 0 corresponds to an acellular mucin corresponding to AMPD.
- Grade 1 is characterized by epithelial tumor cells without mitosis or atypia.
- Grade 3 corresponds to real tumor blocks with cells in a kitten ring, cellular atypia and a high mitotic index.
- Grade 2 is intermediate between grade 1 and 3.

In fact grades 1, 2 and 3 are real cancers corresponding to different degrees at the CPM.

These different classifications have a prognostic interest.

The clinical pictures are not specific [1,17]. Most frequently (40%) it is a progressive abdominal distension related to ascites. The abundance of this ascites can compress the other viscera at the origin of other functional signs such as dyspnea, vomiting, constipation. This ascites can be diffuse with percussion or in checkerboard (omental cake) with palpation of abdominal nodules or indurations. Digital rectal examination is fundamental, it shows a bulging cul-de-sac, sometimes firm under tension and sensitive. Other circumstances may be revealing [1]: a pelvic (ovarian) or abdomino-pelvic tumor (20%); an acute appendicitis chart (10%); during a cure for an inguinal hernia (15%); during the assessment of infertility in women (10%); or incidental discovery during an imaging examination (5%).

The abdominal ultrasound shows either free ascites or immobile and septate ascites, this is the case for our patient.

The abdominal computed tomography (CT) is of great interest because the density of mucin is close to that of fat. It shows large, often heterogeneous hypodense areas under the anterior abdominal wall, under the cupolas as well as in the lateral gutters and the pelvis. A scalloping appearance frequently covers the liver and spleen. Sometimes, this shell compresses these organs and strongly deforms their contours with a "thumbprint" appearance. This examination can sometimes refer to the original tumor (cystic mass, especially in the right iliac fossa) [18].

Magnetic resonance imaging seems to be able to help in the diagnosis of PP but there are few studies [19].

Digestive clouding, in particular barium enema, can help the etiological diagnosis, especially of the appendix, by showing an absence of injection of the appendix by the opaque product, a caecal repression or a notch of the cecal bottom [5]. In our case, the appendix was opacified and normal in appearance.

The ascites puncture when it brings back a gelatinous liquid is pathognomonic of gelatinous ascites, it is the case of our patient. It may not bring anything back at all when the liquid is too thick. This puncture carries a theoretical risk of mucoid or carcinomatous parietal graft [17].

The assay of tumor markers, ACE and Ca19-9, which are greatly increased during CPM, is useful for subsequent surveillance [1,17].

The standard treatment is surgical [7,20]. It consists in operating and re-operating on demand these patients, as soon as the discomfort linked to the volume of the disease requires it at the start, then due to occlusive phenomena thereafter. As much tumor and gelatin should be removed as possible without taking any major surgical risk. The appendix should be removed in principle when possible, for histological analysis [1]. The removal of the initial lesion, when possible, is performed. This tumor reduction surgery is followed by an inevitable progression of PP and after a certain number of reoperations, it becomes more and more difficult and risky due to fibrosis.

Sugarbaker., *et al*. [21] have introduced a new therapeutic concept: ultra-radical surgery which consists in surgically removing (extensive peritonectomies) or destroying all visible lesions by electrofulguration.

This ultra-radical surgery combined with immediate post-operative intraperitoneal chemotherapy (CIPI) done continuously for five days has shown its interest in patient survival unlike systemic chemotherapy [22]. This CIPI has the advantage of allowing the use of very high local concentrations when there is little systemic toxicity [1]. Its effectiveness is potentiated by the addition of hyperthermia between 42° and 43°C [23]. This intraperitoneal chemo-hyperthermia (CHIP) has a great efficacy on residual microscopic disease [24]. This aggressive loco-regional treatment seems to be the reference treatment or at least the "gold standard" of PP treatments because survival is better compared to conventional treatments. It is recommended both for AMDP where the disease is purely peritoneal, without lymphatic or metastatic damage and both for CPM [1].

Adjuvant radiotherapy has not shown its interest [25].

The prognosis of PP depends on:

- Histological type,
- The type of surgery performed, complete or incomplete,
- Whether or not to add a CIPI or CHIP.

But overall survival at five years is between 50% and 70% with ultra-radical surgery when possible. Addition of a CIPI or CHIP appears to improve survival outcomes. Recurrence is as common as after tumor reduction surgery but seems to appear later [22]. The usual causes of PP death are digestive obstruction or jaundice by tumor compression [1].

Conclusion

Peritoneal pseudomyxomas (PP) are very rare, mainly of appendicular origin. Other tumors can be the cause but sporadically, as our case of PP secondary to a carcinoma of the cardia. The standard treatment is ultra-radical surgery, when possible, followed by ultra-peritoneal chemo-hyperthermia. This treatment seems to improve the prognosis for survival and delay recurrences.

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