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Case Report

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Mysterious Gelly Belly: a Case Report of Pseudomyxoma Peritonei and Literature Review.

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Abstract

Pseudomyxoma peritonei (PMP) is a rare clinical condition defined as extensive intraperitoneal spread of mucus associated with a variety of mucinous tumors. Although appendix has usually been implicated as the primary site, some reports found no cause. This case also describes a PMP with no identifiable primary site. A 52-year-old male presented with an abdominal distension evolving for 3 months associated with diffuse abdominal pain, imaging techniques objective intra peritoneal mucoid materials with septated ascites but it failed to identify the primary site. Exploratory laparotomy with Biopsy confirmed PMP but also failed to found the original site.

Keywords: Pseudomyxoma Peritonei; Imaging; Ascites.

Introduction

Pseudomyxoma Peritonei (PMP) is a descriptive term, and not a histopathological diagnosis, illustrating a clinical syndrome related to accumulation in the peritoneal cavity (and / or pelvic) mucinous material, Described for the first time by Rokitansky in 1842, the term peritoneal pseudomyxoma was imposed in 1884 by Werth. Sometimes called "gelatinous disease of the peritoneum", it is a rare condition, the incidence is estimated at 1 to 2 cases per million inhabitants and per year [1, 2].

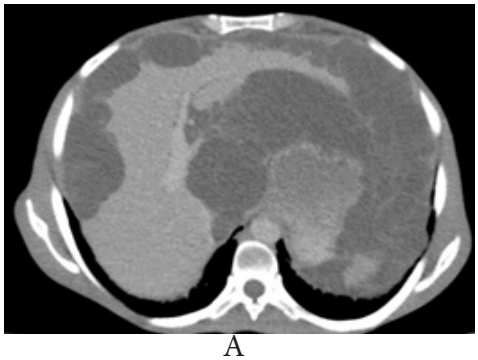
PMP is more commonly seen in women who usually present with increasing abdominal girth and this tends to be related to underlying ovarian lesions, which are usually mucinous tumors. Though uncommon in men, these cases are virtually all associated with a lesion in the appendix [3, 4] Other possible primary sites include colorectum, gallbladder, pancreas, urachus, urinary bladder, breast and lung, but these are uncommon, Primary site of origin was unknown in some cases [5, 6].

Case Report

A 52-year-old man, with no particular pathological history, who presents for abdominal distension evolving since 3 months associated to diffuse abdominal pain and deterioration in general condition. Clinical examination objective a conscious patient, hemodynamically stable, with a normal blood pressure at 130/70 mmHg. Examination of the abdomen shows significant abdominal distension secondary to massive ascites with umbilical hernia, without detectable abdominal mass.

Regarding the blood investigations, we observed elevated level of white blood cells (13000/mm3) and C reactive protein (75mg/l), the other routine biochemical analysis were in normal range.

Abdominal ultrasound shows a massive peritoneal effusion, echogenic and septated. Computed Tomography (CT) revealed a massive peritoneal effusion of the peritoneal cavity, omentum and mesentery, accompanied by scalloping of visceral surfaces, particularly the liver and pancreas, associated with multiple calcified septas (Figure 1).



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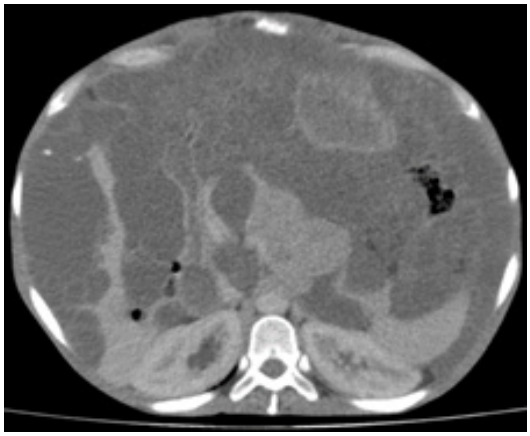

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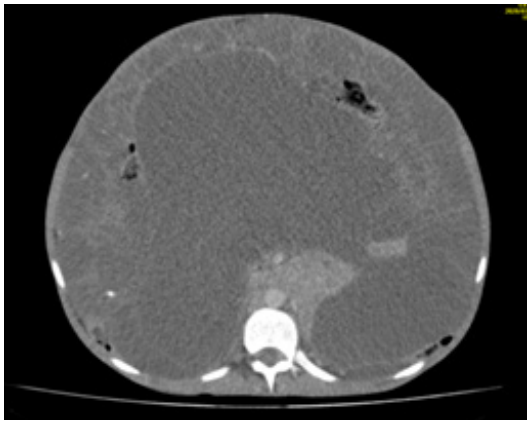

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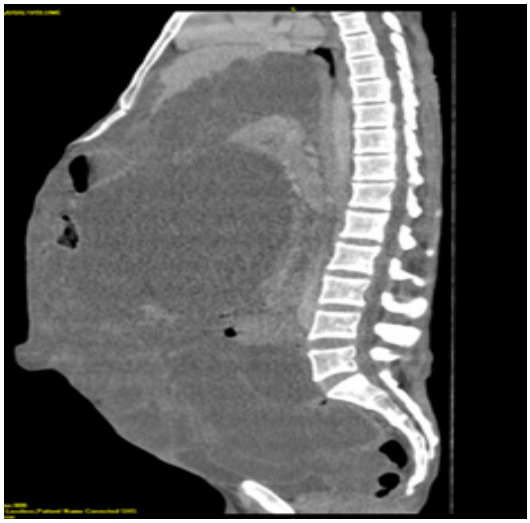

D

Fig 1: Abdominal contrast-enhanced computed tomography in axial (A, B and C) and sagittal (D) sections showing a massive peritoneal effusion, accompanied by scalloping of visceral surfaces, particularly the liver and pancreas (red arrows), with multiple calcified septas (bleu arrows) and multifocal discrete nodules (green arrows).

The patient was referred to a surgical oncologist for treatment. During exploratory laparotomy, peritoneal cavity was filled with semi liquid gelatinous mucoid materials septated into several cysts. Multiple adhesions were noted between intra-abdominal and intra pelvic structures. Biopsy was taken from implants on greater omentum and peritoneal thickening. Appendix looks normal in the ileo caecal region. Peritoneal cavity evacuated and closed after extracting 10 liters of gelatinous ascites (Figure 2). Patient recovered uneventfully and declared outgoing on the eighth post-operative day.

Histological examination of biopsies revealed peritoneal tissue dissociated by areas of mucus without epithelial cells. The cytological study of gelatinous ascites shows a mucous and acellular liquid without specific or suspect cells.

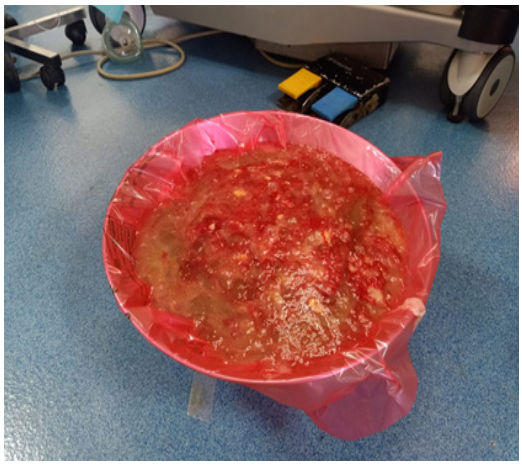

Figure 2: 10 liters of gelatinous ascites extracted during exploratory laparotomy.

Discussion

Pseudomyxoma peritonei is a rare clinical condition defined as extensive intraperitoneal spread of mucus associated with variety of mucinous tumours. The incidence of PMP is about 1–2 per million population [2]. Low incidence forecloses the possibility of large-scale studies, and as a result, there is broad diversity in the definitions, pathology, site-of-origin theories, treatment protocols, and prognosis for this disease [7].

This disease presents a great diagnostic challenge to clinicians. Because of its idle nature and unspecific clinical symptoms, PMP is inclined to be frequently misdiagnosed or diagnosed at advanced stages with generalized peritoneal tumor implants, fistula formation and adhesions are common. In this advanced stage, abdominal symptoms caused by partial or complete bowel obstruction are the main complaints [8].

Ronnet proposed a classification into 3 histological grades according to the degree of malignancy: grade 1 or DPAM (Disseminated Peritoneal Adenomyosis), the most frequent, characterized by peritoneal lesions consisting essentially of mucus, poor or devoid of cells and atypia (84% 5-year survival); grade 3 or PMCA (Peritoneal Mucinious Carcinomatosis) with characteristics close to adenocarcinomatous colorectal carcinosis (7% 5-year survival); and grade 2 or PMCA ID (intermediate grade) of hybrid architecture and intermediate prognosis (50% 5-year survival). In fact, the exact classification of the pseudomyxoma for one patient is

complex because several grades of pseudomyxoma can coexist in the peritoneal cavity (leading to the hypothesis of an evolutionary sequence " adenomucinosis - adenocarcinoma"). We tend in any case to no longer consider PMP as a benign pathology, whatever its grade because its evolution is always long-term towards to a lethal outcome [9-11].

Cross-sectional imaging especially computed tomography (CT) is the modality of choice as it allows relatively accurate localisation and quantification of PMP. The results of computed tomography revealed characteristic findings of PMP such as multiple complex cystic masses of low attenuation in the peritoneum, which may have rims of calcifications, characteristic scalloping of the liver (and occasionally splenic) margins, omental thickening, omental 'cake' and peritoneal implants, septated 'pseudo' ascites and varying degrees of compression of the visceral organs and structures [12]. CT imaging can detect mucocele, which is a significant prognostic factor, allowing surgeons to take the necessary precautions to avoid perioperative rupture and the consequent peritoneal seeding. Even in cases of early detection of rupture of a mucocele, a more tailored and radical approach could be taken, reducing the chances of reoperation with unfavorable prognoses [13]. CT can also be used to follow-up and re-evaluate patients [12].

The differential diagnosis includes primary or secondary peritoneal tumours, which may also present with scalloped liver margins and septated ascites.10 Pancreatitis with ascites and pancreatic pseudocysts may also form part of the differential diagnosis, although the presence

Of pancreatic abnormalities may allude to this diagnosis. Infective causes such as pyogenic peritonitis, widespread echinococcal disease and TB peritonitis, may be considered but the clinical presentation of these patients as well as the presence of other imaging findings such as liver abscesses or micro-abscesses (in the case of TB) may also be present [12].

Our patient had an unknown origin pseudomyxoma peri¬tonei, which is a rare cause of ascites. The appendix was presumed, at the beginning, to be the site of origin for pseudomyxoma peritonei in our case, but this was dismissed during laparotomy. As PMP is in the majority of cases secondary to an appendicular mucocele or less frequently to gastrointestinal or adnexal tumors in women, its primary peritoneal origin remains a diagnosis of elimination. This kind of presentation continues to be an issue of debate among oncologists interested in research on pseu¬domyxoma peritonei.

The evolution of treatment strategies of PMP remains debated though the current mainstay of the treatment remains surgical extirpation of the lesion. Repeated cytoreductive surgical debulking procedures including resections of the tumor implants, omentum and obstructive bowel are common due to recurrence of the disease. A study by Draco et al showed that cytoreductive surgery with intraperitoneal hyperthermic perfusion permitted complete tumor removal, and this study confirmed the efficacy of this combined treatment in terms of improved long-term survival and better local control of the disease [14]. Indeed, the current strategy of treatment includes cytoreductive surgery combined with intraoperative hyperthermic intraperitoneal chemotherapy (mitomycin at 42 degrees C). The aim is to avoid entrapment of tumor cells at operative sites and to destroy small residual mucinous tumor nodules [15, 8].

Considered for several years as a "border-line" pathology, PMP must be reconsidered as a malignant pathology because of its long-term lethal evolution; major prognostic factors are stage of local invasion throughout the peritoneal cavity, histological grade of causative lesion and completeness of cytoreduction [16, 17].

Conclusion

Although PMP is a relatively rare entity, radiologists must be aware of its possible imaging findings, common locations, and possible patterns of recurrence. The origin of the primary tumor should also be investigated, although it may remain unknown in rare situations.

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