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Benign multicystic peritoneal mesothelioma (BMPM) presenting with ambiguous symptoms: A rare case report

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ABSTRACT

Introduction: Benign multicystic peritoneal mesothelioma (BMPM) represents a very rare clinical entity, with only 130 registered cases in the medical literature, therefore it is usually overlooked from the list of differential diagnoses. The treatment consists of surgery and other authors suggest complementing it with hyperthermic intraperitoneal chemotherapy.

Presentation of case: A 25-year-old multiparous female experienced periodic abdominal discomfort for two weeks. She developed constipation, urinary urgency, and irregular menstruation. Family history was remarkable for endometrial and breast cancer. Abdominal examination revealed a palpable mass. Abdominal ultrasound and computed tomography scan identified the multicystic appearance of the mass. The diagnosis was unclear, therefore exploratory laparotomy was performed, which revealed multiple grape-like clusters of cysts that were excised immediately. BMPM was diagnosed based on the pathology report. Eventually, the follow-up did not reveal any recurrence.

Discussion: Mesothelial tumors include three pathological entities, including Benign multicystic peritoneal mesothelioma (BMPM). BMPM is an uncommon neoplasm and has a high recurrence rate after surgery. BMPM consists of clear cysts that take the shape of a grape-like cluster. Clinically, BMPM resembles a tangible abdominal mass and it is challenging to be diagnosed, due to its numerous differential diagnoses.

Conclusion: The definitive diagnosis of intraperitoneal cystic masses is usually challenging. Therefore, BMPM -although very rare- should always be thought of when dealing with an intraperitoneal cystic mass, especially in women in the reproductive years. In our case cytoreductive surgery solely was sufficient to achieve a disease free follow up, however, further studies regarding treatment and follow-up are required.

1. Introduction

Benign multicystic peritoneal mesothelioma (BMPM[1]) is a very rare benign cystic tumor arising from the peritoneal mesothelioma and it mostly affects women in reproductive age [2]. BMPM should be kept in mind when confronting a case of peritoneal cysts. Surgery is the mainstay treatment with the consideration of hyperthermic intraperitoneal chemotherapy as adjuvant therapy [3,4] Recurrence is common [5].

We report a rare case of a 25-year-old female with BMPM. This case has been reported in line with the SCARE criteria [1].

2. Case presentation

A 25-year-old female, gravida 2, para 2 presented to the clinic with asymmetric abdominal distention and occasional abdominal discomfort for two weeks. The pain increased gradually and spread from the right iliac fossa to the right hypochondrium and the right thigh. The patient had irregular menstruations and menorrhagia since the last delivery; therefore, she used oral contraceptive pills to regulate the menstrual cycle. Fatigue, nausea, anorexia, and urinary urgency were also reported. Furthermore, the patient developed constipation, which failed to

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improve on laxatives. The patient's obstetric history was unremarkable and consisted of two vaginal deliveries. Past medical and surgical history was also unremarkable, except for a moderate uterine pain as a result of IUD insertion after the second birth. She had a family history of breast and endometrial cancer. She is not a smoker nor alcoholic. The patient also mentioned a fourth degree of consanguinity between her parents.

The patient appeared well and was hemodynamically stable. Mild pallor was noted, but the vital signs were normal. On Abdominal examination, we inspected a right-sided distention without engorged veins and palpated a tender mass. The physical examination of other systems was otherwise unremarkable.

Abnormal laboratory results included elevated platelet count (425 × 109/L), decreased hematocrit (35%) and hemoglobin (11.5 g/dL), which indicated anemia. However, the white blood cell count was within normal range. Additional assays were performed, which revealed a negative echinococcus antibody test and a significant increase in CA 19.9 and CA 125 tumor markers levels, with values of 69.59 and 64.08 U/ml respectively.

Abdominal ultrasound (US) showed a large pelvic mass extending to the upper borders of the umbilical region, with a multicystic appearance and multiple thick septations measuring 25 × 15 cm. The uterus, liver, and spleen were normal and homogeneous. A small amount of free fluid was also noted in cul-de-sac. Computed tomography (CT) scan revealed an irregular multi-loculated cystic-appearing mass, located in the abdomen and pelvis with dimensions of 25 × 10 cm (Fig. 1). Para-aortic lymph nodes were not enlarged. Given these findings, we considered right ovarian mucinous cyst, appendiceal mucinous adenocarcinoma, hydatid cyst type CE2 and pseudomyxoma peritonei as a potential diagnosis.

Based on these findings the diagnosis was unclear, so the general surgery specialist with the gynecology specialist performed a laparotomy. Intraoperative inspection revealed multiple grape-like clusters of cysts (Fig. 2).

The clusters of cysts appeared clear, smooth-walled and were diffuse, which made the resection process challenging. Eventually, the mass and the free-floating cysts were carefully harvested and removed after releasing the adhesions with the surrounding structures. A specimen was sent for pathological evaluation. Microscopic examination revealed a single layer of mesothelial cells lining the cysts and varying from flattened to cuboidal, with inflammatory cell infiltration in the stroma (Fig. 3).

Multicystic benign mesothelioma was confirmed in the pathology report.

The postoperative period was uneventful. On five months follow-up, the patient presented to the clinic, both physical exam and US showed no clinical evidence of disease recurrence.

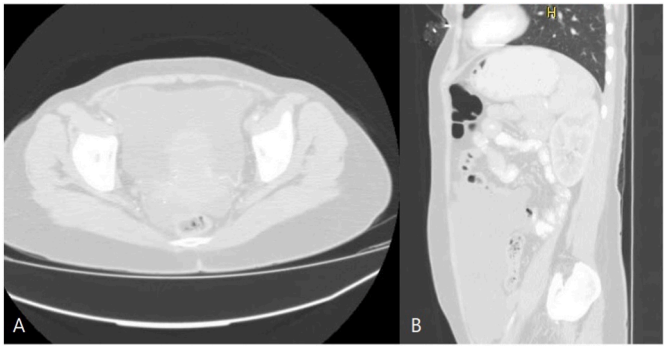

Fig. 1. (A) Axial and (B) sagittal computed tomography (CT) images show an irregular cystic-appearing mass, measuring 25 × 10 cm.

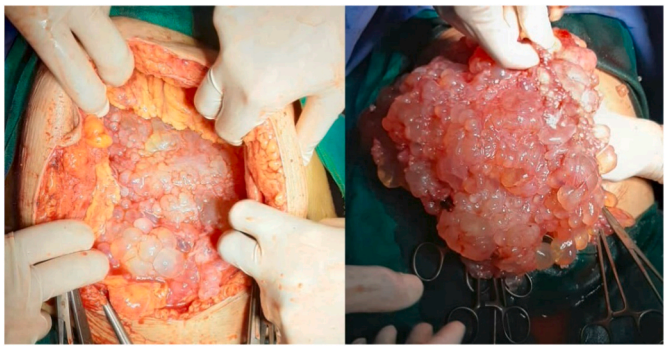

Fig. 2. The gross appearance of the tumor shows multiple grape-like clusters of cysts.

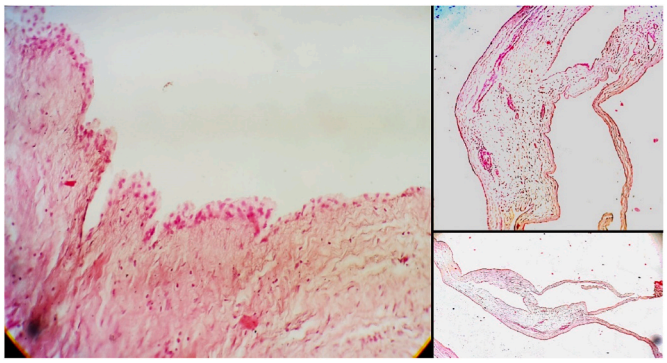

Fig. 3. Microscopic examination, using Hematoxylin and Eosin (H&E) Staining, demonstrated a single layer of mesothelial cells lining the cysts and varying from flattened to cuboidal.

3. Discussion

There are three pathological forms of mesothelial tumors of the peritoneum with variable prognoses. The first one is a benign adenomatoid tumor, which has a low rate of recurrence after surgical eradication, the second form is malignant peritoneal mesothelioma, which has a poor prognosis, and the last one is benign multicystic peritoneal mesothelioma (BMPM) [5]. The pathogenesis of the preceding diseases could be classified into either a neoplastic proliferation or a reactive proliferation of the mesothelium due to prior abdominal surgery, pelvic inflammatory disease, or endometriosis [6,7].BMPM is a rare benign tumor with distinguished regional expansion and a high potential for recurrence after surgery [5]. The etiology of BMPM is unexplored, but there is no connection between its occurrence and asbestos exposure, unlike malignant mesothelioma [7]. It occurs predominantly in females of reproductive age. The mean age of presentation is 37 years (ranging from 15 to 92 years). The medical literature reveals that only 17% of cases occur in men [8].

BMPM is characterized by multiple or solitary fluid-filled, thin-walled adherent cysts with a high preference for the pelvic peritoneum. In general, cysts are clear and organized in grape-like clusters separated by fibrous tissue, which often reveal acute and chronic inflammation, moreover mild to moderate cytological atypia is frequently observed [6,8]. According to several reported cases, the presence of cysts in the Douglas pouch (the lowest area of the peritoneum) is almost certain [3,9]. However, cysts can be free-floating in the peritoneal cavity. The intracystic fluid components vary from clear to blood-tinged, mucinous, or gelatinous in some cases [6,8]. The volume of the cysts often ranges from 1mm to 200mm [8]. Histologically, BMPM appears as mesothelial proliferation with multiple cell layers, papillary projections, and tubules. Occasionally, Squamous cell metaplasia may occur [6,7].

The most common complaint of BMPM is chronic pain but it varies from asymptomatic to acute abdomen [7]. On clinical examination, BMPM appears as a palpable mass with distension of the abdomen [6,8].

As we mentioned in our case, the preoperative diagnosis of BMPM is difficult; several differential diagnoses must be considered, such as cystic lymphangioma, endometriosis, and cystic epithelial neoplasms of the ovaries and peritoneum including pseudomyxoma peritonei. The most complex differential diagnosis is cystic malignant mesothelioma. In our case; due to the lack of diagnostic materials, our diagnosis depended on hematoxylin and eosin stain which is considered the best morphological test to distinguish between benign or malignant in the pathological examination according to King et al. [10]. In addition, the positive results of CA-125 have been observed in both benign and malignant mesothelioma. The most common usage of CA-125 is for marking the ovarian adenocarcinomas, however, it has not been considered as a particular antigen [11]. This recommends that mesothelial cells are effective in CA-125 combination and excretion [11]. This explains the positive results of CA-125 in our patient.

Walz et al. [12] indicated that CA-19-9 is normally found in glandular epithelium and its neoplastic counterparts. Therefore, the presence of elevated CA-19-9 in our patient does not ascertain the neoplasm.

A systematic review by King et al. concluded that the best morphological test to distinguish between benign or malignant pleural disease is the pathological examination with hematoxylin and eosin stain [10].

The UltraSound (US) is always a part of the diagnostic process in a suspected abdominal mass, which typically reveals an anechoic multiloculated cystic mass. Computed tomography (CT) and Magnetic Resonance Imaging (MRI) may be used as complementary investigations in making the preoperative diagnosis [13]. All preoperative diagnostic modalities were unclear in our case, so we were obliged to perform an open laparotomy.

Therapeutic protocols of BMPM consist of cytoreductive surgery, but some authors suggest associating hyperthermic intraperitoneal chemotherapy to reduce the high risk of recurrence [3,4].Regarding our case, we suggest considering the cytoreductive surgery approach as the most suitable and sufficient treatment and as complete as possible while avoiding cysts rupture –which is thought to be origin of postoperative recurrence.

Since BMPM is very rare there is lack of evidence regarding its treatment follow-up, therefore we find it essential to report this case with long uneventful follow-up period to inform other clinicians about our management and to keep this diagnosis in mind preoperatively when confronting a similar case. We still recommend further studies in this domain.

4. Conclusion

Every physician should keep BMPM -despite its rarity-in his list of differential diagnoses when spotting a multicystic peritoneal mass, especially in a female in her reproductive years. Though cytoreductive surgery solely while avoiding cysts rupture lead to an uneventful follow-up in our case, we still recommend further research regarding treatment and follow-up.

Patient perspective

The patient participated in the treatment decision and she was satisfied with the results of the treatment. Her perspective on this treatment was to get rid of the uncomfortable mass without complications.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

Not required for case reports at our hospital. Single case reports are exempt from ethical approval in our institution.

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Declaration of competing interest

The authors declare that they have no conflict of interest.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2020.12.022.

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