CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 74 (2020) 218–221

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Benign multi-cystic peritoneal mesothelioma of the porta hepatis

Hassan Aziz, Michelle Hao, Alam Merchant, Daniel Alfson, Chris Foran, M. Raashid Sheikh*

Department of Surgery, Division of Hepatobiliary Surgery, Keck School of Medicine, University of Southern California Medical Center, Los Angeles County, Los Angeles, CA, United States

ARTICLE INFO

Article history:
Received 22 May 2020
Received in revised form 23 August 2020
Accepted 24 August 2020
Available online 29 August 2020

Keywords:
Benign multi-cystic peritoneal mesothelioma of the porta hepatis

ABSTRACT

INTRODUCTION: Our case report describes a patient where multiple laparoscopies five years from initial presentation of symptoms were performed prior to laparotomy for benign multi-cystic peritoneal mesothelioma (BMPM), which has not been documented.
PRESENTATION: A 61-year-old woman presented with years of chronic abdominal pain. Computerized tomography (CT) demonstrated a multi-cystic mass near the porta hepatis, and ultrasound was concerning for contained gallbladder perforation. Fine needle aspiration (FNA) demonstrated benign ductal epithelial cells in a background of mucin and bile without the presence of malignant cells. During laparotomy, a cystic mass attached to the porta hepatis seen emanating from the small bowel mesentery, and additional small cystic lesions through the abdomen were removed. The specimen, measuring $26 \times 18 \times 8$ cm, showed multi-loculated cysts filled with serous fluid.
DISCUSSION: BMPM is a rare neoplasm of mesothelioma cells originating from serosa of viscous organs. BMPMs appear as cystic structures with thin walls containing mucinous/gelatinous fluid. Microscopic features include a lack of invasion and no increased cellularity in the stroma, with or without inflammation (Myers & Babiker, 2018). It is postulated to be either a reactive or neoplastic process. There is no gold-standard treatment for BMPM. Our case is unique in the sense that our patient required several surgical biopsies before final diagnosis could be made.
CONCLUSION: This case highlights the difficulty of diagnosing BMPM and differentiating it from malignant diseases that can present similarly and can be associated with significantly worse prognosis. Defined management strategies have yet to be demonstrated.

1. Case summary

A 61-year-old Hispanic woman presented with complaints of several years of chronic, dull abdominal pain. Her past medical history is significant for hypothyroidism, hyperlipidemia, gastritis, gastroesophageal reflux, and former use of tobacco. She had no relevant surgical history. Computerized tomography (CT) demonstrated a multi-cystic abdominal mass located near the porta hepatis (Fig. 1) along with multiple low-attenuating peritoneal nodules.

Initial workup with endoscopic ultrasound could not localize the source of the mass, but it was visualized as separate from the pancreas, bowel, gallbladder, and bile ducts. Ultrasound findings were also concerning for a contained gallbladder perforation. Fine needle aspiration (FNA) sampling the mass near the gallbladder demonstrated benign ductal epithelial cells in a background of mucin and bile without the presence of malignant cells. The differential diagnosis at this point included a cystic neoplasm versus a contained, perforated gallbladder. Further imaging evaluation was planned. A subsequent CT in the following months showed near complete resolution of the multi-cystic mass near the porta hepatis. However, there was interval growth of the mesenteric nodules from the presenting CT, suspicious for pseudomyxoma peritonei, as well as a newly dilated appendix.

A diagnostic laparoscopy was performed. Intraoperative findings reported a grossly normal appendix and gallbladder, as well as absence of any fluid collection or cyst in the area of the porta hepatis. The appendix was removed, and a biopsy of a peritoneal nodule was obtained. On final pathology, the appendix was unremarkable, and the peritoneal nodule was determined to be a mesothelial cyst.

Further workup included a colonoscopy, which was normal. A transvaginal ultrasound was concerning for endometrial hyperplasia, although she has never had abnormal uterine bleeding and was not followed by gynecology-oncology. Subsequently, the patient experienced worsening abdominal discomfort, and surveillance CT imaging continued to show an increase in the size and number of peritoneal nodules.

* Corresponding author.
E-mail address: MohdRaashid.Sheikh@med.usc.edu (M. Raashid Sheikh).

https://doi.org/10.1016/j.ijscr.2020.08.047

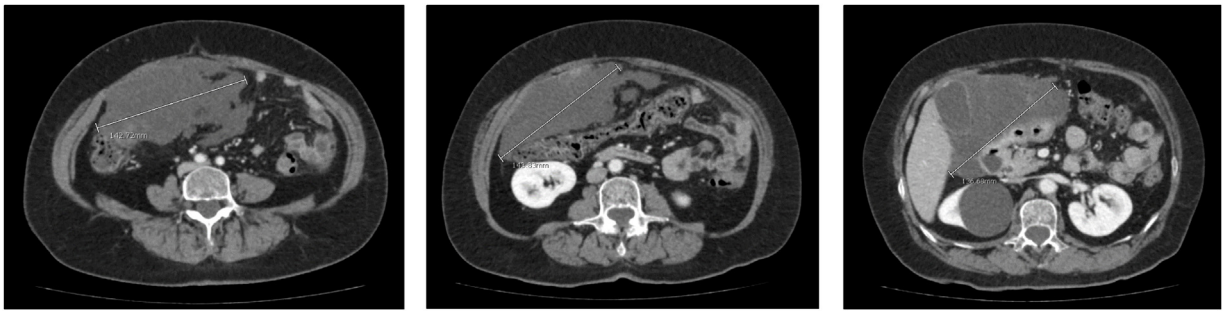

Fig. 1. Axial CT imaging showed a large multi-cystic intra-abdominal mass in the right abdomen spanning from the porta hepatis to the right lower quadrant.

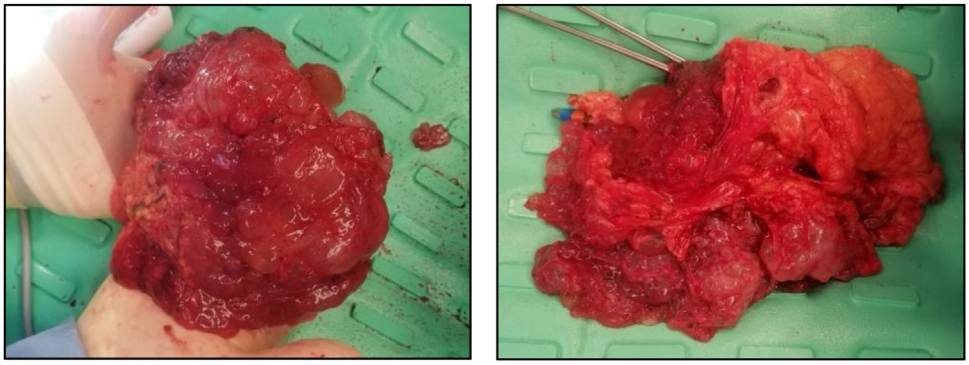

Fig. 2. Multi-loculated cystic surgical specimen measuring 26 × 18 × 8 cm filled with serous fluid.

A second diagnostic laparoscopy with peritoneal biopsy was then performed given the patient's worsening symptoms and imaging findings. Intraoperative findings reported diffuse mucinous and gelatinous material throughout the abdomen. Bilateral ovaries and the gallbladder appeared normal. Biopsy of a peritoneal nodule revealed a peritoneal inclusion cyst without malignant cells.

Given the benign findings on surgical pathology with the two previous operations, she continued to be treated with conservative management. However, as her abdominal pain worsened in subsequent years, exploratory laparotomy and surgical debulking were performed for therapeutic benefit and tissue diagnosis.

During laparotomy, a large cystic mass seen emanating from the small bowel mesentery and involving the porta hepatis, gallbladder, stomach, and duodenum. A smaller, multi-loculated cystic mass was also found in the lesser sac. Additional, innumerable small cystic lesions through the abdomen were removed. Grossly, the abdominal mass specimen, measuring 26 × 18 × 8 cm, showed multi-loculated cysts filled with serous fluid (Fig. 2). On immunohistochemistry, tissue from the large cystic mass stained positive for calretinin, CK4/5, CK7, and D2-40 (Figs. 3–5). The final diagnosis was benign multi-cystic peritoneal mesothelioma (BMPM).

2. Discussion

Benign multi-cystic peritoneal mesothelioma (BMPM) is a rare neoplasm of mesothelioma cells originating from serosal linings of viscous organs. Approximately 150 cases have been reported in the literature. The first case was described in 1979 by Mennemeyer and Smith [1]. Its incidence is estimated at 0.15/100,000 annually [2]. BMPMs usually occur in women of reproductive age and have been associated with inflammation (endometriosis, pelvic inflammatory disease), previous abdominal surgeries, and cadmium exposure. Only seventeen percent of cases have been reported in men [3]. Most cases occur in the peritoneal viscera, but two cases arising in the pleura and single cases arising in the pericardium, spermatic cord, and tunica vaginalis have been described [3]. There is no association between asbestos exposure and benign mesothelioma. Patients may present with abdominal and pelvic pain or mass. In most cases, the lesions may be found incidentally during laparotomy.

Grossly, BMPMs appear as grape-like cystic structures with thin walls containing mucinous or gelatinous fluid. Microscopic features of benign mesothelioma include a lack of invasion and no increased cellularity in the stroma, with or without inflammation [2]. Characteristically, the cysts are lined with a single layer of flat mesothelial cells alternating with cuboidal cells with hobnail features and fibrovascular stroma between cysts. Vascular congestion in the stroma may also be observed [3]. While malignant mesothelioma may also present as a cystic mass, histology of benign mesothelioma lesions lacks cellular atypia and increased mitotic counts [4]. Immunohistochemistry markers of benign mesothelioma include desmin, calretinin and D2-40 [2].

The etiology of BMPM is unclear. It is postulated to be either a reactive or neoplastic process. Risk factors including pelvic inflammatory disease, endometriosis, and previous abdominal surgeries suggest BMPM is a reactive lesion of peritoneal mesothelial cells in response to chronic irritation. Associations favoring a neoplastic process include the high likelihood of recurrence and a slow but progressive growth of untreated lesions. The role of female sex hormones is also unclear in the pathogenesis of BMPM. In a case series of 14 lesions, 2 cases tested positive for PR and/or ER [5,6].

The differential diagnosis includes lymphangioma, pseudomyxoma peritonei, malignant peritoneal mesothelioma and cystic adenomatoid tumor. Other differentials include cystic forms of endosalpingosis, endometriosis, Müllerian cysts involving the retroperitoneum, and cystic mesonephric duct remnants.

Recurrence rates even after complete surgical debulking are very high, ranging from 33 to 50% [4]. There has only been one death reported in the literature of a patient due to local tumor effects after refusal of resection for 12 years [6]. It is extremely rare

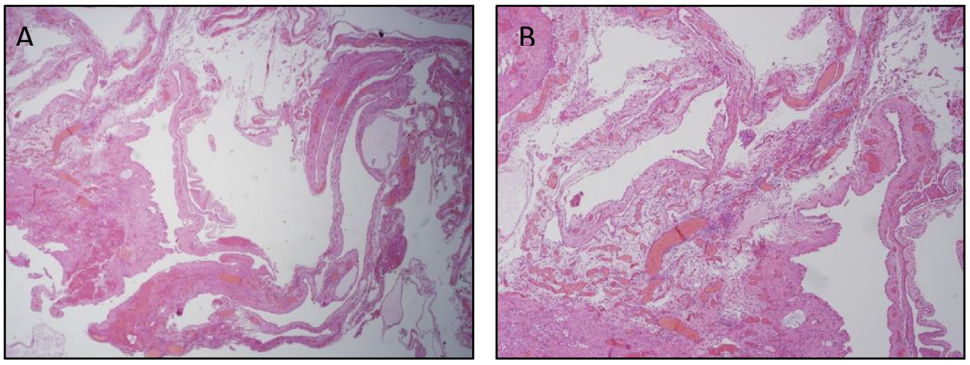

Fig. 3. Multi-cystic peritoneal mesothelioma with vascular congestion, HE staining ×20 (A) and ×40 (B).

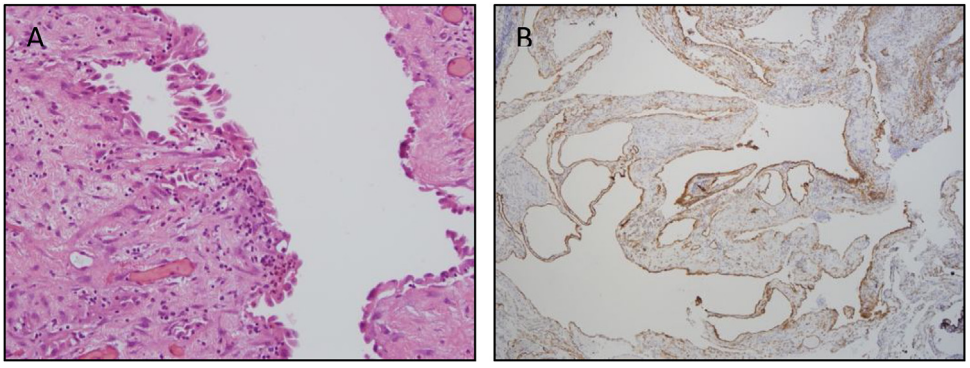

Fig. 4. (A) Cystic wall with fibrovascular stroma lined by flat to cuboidal mesothelial cells, HE staining ×200. (B) D2-40 immunohistochemical staining.

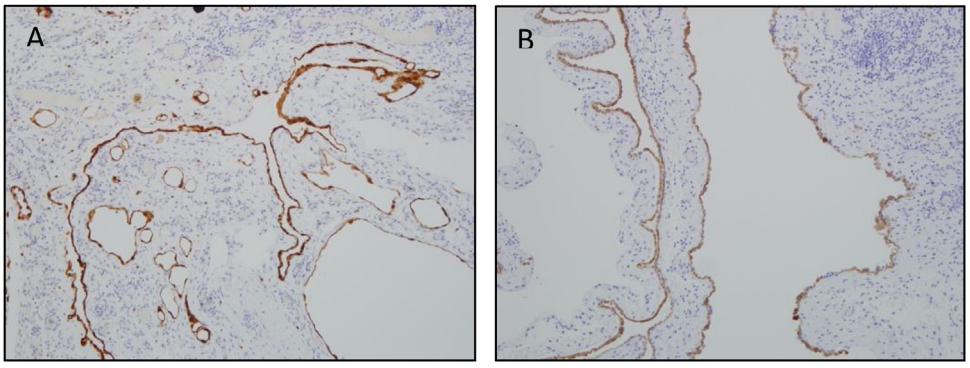

Fig. 5. (A) Calretinin immunohistochemical staining. (B) CK 5/6 immunohistochemical staining.

for a benign mesothelioma to progress to malignant mesothelioma, with only two reported cases in the literature [3]. There is currently no gold-standard treatment for BMPM, but two case series on the results of extensive peritonectomy followed by HIPEC (hyperthermic intraperitoneal chemotherapy) are reported in the literature. The first series reported 1 out of 5 patients with recurrence and the second series reported 2 out of 5 patients with recurrence. Follow up time ranged from 6 to 69 months in the first series and 3–102 months in the second series [7].

3. Conclusion & case summary continued

Our patient's case was unique in the sense that the initial, loculated cystic mass near her porta hepatis appeared to spontaneously regress in the span of two months, as detected by follow-up CT. Yet, subsequent imaging studies performed for continued abdominal pain re-demonstrated the cystic mass. Also, initial endoscopic ultrasound suggested gallbladder perforation though the gallbladder was found to be intact on laparoscopy. The ultrasound findings of the gallbladder may have represented primary lesions of BMPM or secondary changes from an inflammatory response instead. Taken together, these regressions and growths may suggest that both reactive and neoplastic processes were components in this patient's disease. A major learning point from this study is to be able to recognize BMPM lesions or changes from inflammatory responses and how they differ from gallbladder perforation on ultrasound of the abdomen, specifically in the gallbladder, given the context.

Because our patient was a higher risk candidate for debulking surgery, multiple laparoscopies spanning five years form initial presentation were performed prior to laparotomy. This case highlights the difficulty of diagnosing BMPMs and differentiating it from

malignant diseases that can present similarly and that can be associated with significantly worse prognosis. Given the rarity of this disease, defined management strategies have yet to be demonstrated with solid evidence. Our case is unique in the sense that our patient ultimately required several surgical biopsies before final diagnosis could be made.

After discussion with the medical oncologists at our facility, the patient elected to follow recommendations for continued surveillance of symptoms and CT imaging as needed. She will be followed as an outpatient given the high risk of recurrence. Should symptoms recur and if transformation to malignant pathology is identified on pathology, HIPEC may be considered.

Considerations to the SCARE 2018 criteria were taken into account when writing this manuscript [8].

Declaration of Competing Interest

The authors report no declarations of interest.

Funding

None.

Ethical approval

NA.

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.

Author's contribution

Dr Aziz – wrote the manuscript.
Dr Sheikh – concept and review.
Dr Foran – data collection.
Alam Merchant – editing.

Registration of research studies

1. Name of the registry: NA.
2. Unique identifying number or registration ID: NA.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): NA.

Guarantor

Dr Sheikh.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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