Case Report

The rare occurrence of unifocal peritoneal mesothelioma: a case report, literature review, and future directions

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Background: Mesothelioma is a rare, aggressive disease originating from mesothelial cells and carries a poor prognosis. Mesothelioma may arise from the pleura, pericardium, or peritoneum. Peritoneal mesothelioma (PM) usually spreads in a diffuse manner; however, a localized unifocal form of PM may occur. Literature on unifocal mesothelioma remains scarce.

Case Description: Herein, we highlight a case of localized epithelioid PM in an 81-year-old gentleman with the unique challenges faced during management. The pelvic mass was 7 cm, well-circumscribed, and hyper-vascular with fibrous attachments to the abdominal wall. The patient had a peritoneal cancer index (PCI) of 4 on initial diagnostic laparoscopy. Diagnosis was confirmed by histology. Resection of the mass with a partial omentectomy was performed. Months later, the patient developed recurrence detected on follow-up imaging in the peri-splenic region. The patient underwent cytoreductive surgery (CRS) and heated intraperitoneal chemotherapy (HIPEC) for 60 minutes using mitomycin C and cisplatin followed by an uneventful recovery. Our case report is followed by a review of literature on disease pathophysiology, treatment options, and recently promising immunotherapy approaches.

Conclusions: CRS and HIPEC remains the standard treatment regimen for patients with PM. Nonetheless, a more nuanced approach might be indicated in specific patients with localized unifocal PM. Disease distribution and burden may impact the decision on surgical management in selected patients.

Keywords: Epithelioid mesothelioma; peritoneal malignancy; cytoreductive surgery (CRS); heated intraperitoneal chemotherapy (HIPEC); mesothelin; case report

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Introduction

Mesothelioma is a rare malignancy of mesenchymal origin, most commonly arising from the pleura (1). Approximately 15% of mesothelioma originates in the peritoneum, known as primary malignant peritoneal mesothelioma (PM), leading to diffuse spread within the abdominal cavity. Secondary peritoneal metastasis may also occur from dissemination of gastrointestinal or gynecologic malignancies (2). A solitary mass-like appearance of PM

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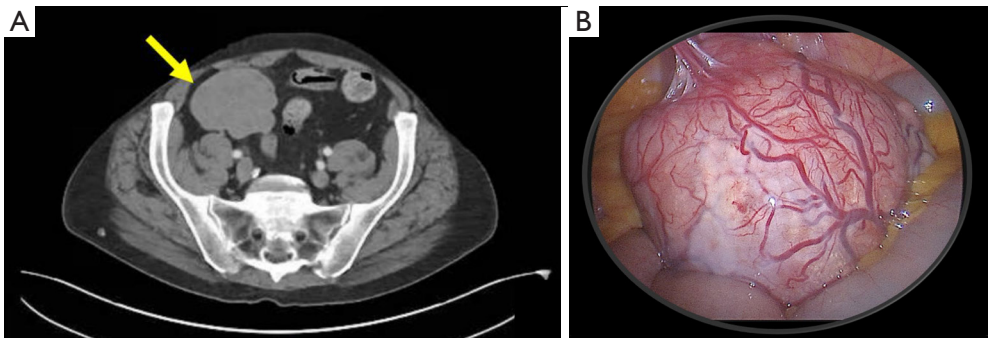

Figure 1 Radiologic and gross imaging of initial mass. (A) Computed tomography showing a mass in the right lower quadrant mass measuring 5.5 cm × 7.9 cm × 7.3 cm (yellow arrow). (B) Intra-operative 7 cm mass consistent with the patient's pre-operative imaging.

originating from the abdominal wall is not a common presentation and may lead to a misdiagnosis, ultimately delaying appropriate therapy.

In this case report we showcase an atypical presentation of peritoneal mesothelioma as a unifocal, well-circumscribed mass originating from the parietal peritoneum, in contrast to the more common presentation of diffuse PM with more widespread parietal and visceral peritoneal involvement. We present this case in accordance with the CARE reporting checklist (available at https://jgo.amegroups.com/article/view/10.21037/jgo-24-266/rc).

Highlight box

Key findings

- Unifocal, localized peritoneal mesothelioma (PM) is rare, without specialized treatment guidelines.

What is known and what is new?

- Although different treatment modalities have been established for diffuse PM to include cytoreductive surgery and heated intraperitoneal chemotherapy, systemic cytotoxic chemotherapy, immunotherapy, and targeted molecular therapy; the benefit of each remains unclear for patients with unifocal PM.
- A nuanced surgical and systemic approach should be tailored for patients with localized PM and based on individualized clinical presentation.

What is the implication, and what should change now?

- A high level of suspicion for PM should be maintained when evaluating a patient with an intrabdominal mass. Imaging modalities, histopathology, and tumor markers may aid in the diagnosis.
- In addition to standard surgical treatment, novel immunotherapy regimens are currently being evaluated in clinical trials.

Case presentation

Initial presentation

An 81-year-old male with a history of hypertension, coronary artery disease was found to have a well-circumscribed mass in the abdomen after computed tomography (CT) imaging was obtained for acute-onset abdominal pain. Family history is notable for a mother with diffuse abdominal malignancy of unknown origin and a son with rectal cancer. Of note, the patient was exposed to secondhand asbestos during childhood from his father who worked in a shipyard. The CT scan exhibited a mass in the right lower quadrant measuring 5.5 cm × 7.9 cm × 7.3 cm (*Figure 1A*). Pathology from a percutaneous biopsy showed morphological and immunohistochemical (IHC) features of epithelioid mesothelioma. A diagnostic laparoscopy was performed for evaluation of peritoneal disease burden and definitive operative planning. The laparoscopy was most notable for a 7-cm mass consistent with preoperative imaging, without any additional parietal peritoneal abnormalities upon survey. Empiric four-quadrant peritoneal biopsies were harvested. The mass was well-circumscribed, arising from the right lower quadrant parietal peritoneum and loosely adherent to the underlying mesentery with grossly hyper-vascular features (*Figure 1B*). At the end of the case, a peritoneal cancer index (PCI) of 4 was noted. Pathology of the four-quadrant peritoneal biopsies returned as fibromuscular tissue with benign mesothelial cell proliferations.

Index operation

Subsequent resection of the mass was performed along with

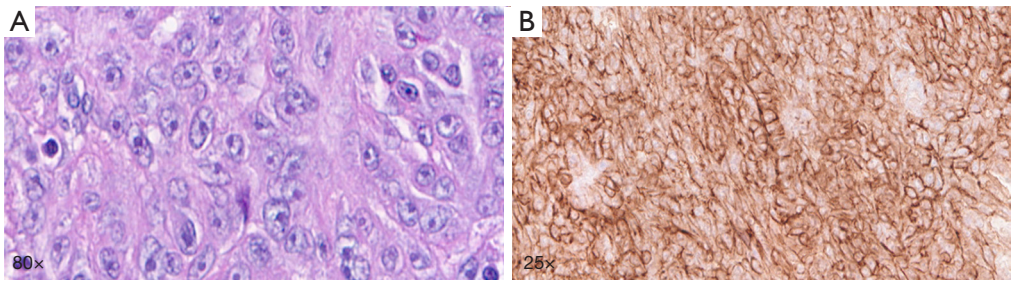

Figure 2 Histopathology of resected mass. (A) Hematoxylin and eosin cross-sectional representative image of the resected mass demonstrates the characteristic features of epithelioid mesothelioma cells with large nucleoli, eosinophilic cytoplasm, and papillary structures. (B) Immunohistochemistry of tissue slides demonstrates mesothelin (>99%, 3+).

adjacent partial omentectomy. The patient recovered well and was discharged on post-operative day 3. Pathology displayed malignant epithelioid mesothelioma measuring 10 cm × 7.5 cm × 5 cm without tumor cells in the omental specimen. Histopathology showed cells in sheets with eosinophilic cytoplasm, pleomorphic vesicular nuclei, and large nucleoli. IHC stains were positive for calretinin, mesothelin (>99%, 3+), and Wilm's tumour-1 (WT-1). In some cells, the BRCA-1 associated protein 1 (BAP1) expression was lost with weak nuclear staining (*Figure 2*).

Next-generation genomic sequencing reported two pathological small nucleotide variants (SNV): an *NF2* R341 mutation with a variant allele frequency (VAF) of 61% and *TP53* R175H mutation with a VAF of 62%. There were no reportable RNA fusion transcripts.

Peritoneal recurrence and definitive cytoreductive surgery (CRS)/HIPEC

Follow-up CT imaging after 5 months demonstrated two small peri-splenic lesions not seen previously. It was decided to observe the evolution of these lesions over time. Interval growth of these two peri-splenic lesions with no additional masses were demonstrated on imaging 3 months later (*Figure 3A,3B*). Exploration indicated a PCI of 4, with two peri-splenic masses and punctate masses along the residual omentum concerning for mesothelioma (*Figure 3C,3D*). The patient then underwent CRS, splenectomy, and completion omentectomy with heated intraperitoneal chemotherapy (HIPEC) using cisplatin and mitomycin C for 60 minutes. The completeness of cytoreduction (CC) score was CC-0, indicating no remaining residual disease. Recovery was uneventful, and he was discharged on post-operative day 9.

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for the publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

Epidemiology and pathophysiology

Peritoneal mesothelioma is a rare and invasive disease defined by the malignant transformation of the mesothelium, often with diffuse involvement of the peritoneal surfaces. The annual incidence diagnosed in the United States is approximately 800 cases, with the median age at diagnosis of 63 years (3). Median overall survival is 53 months among patients treated with complete or near-complete CRS and HIPEC (CC-0 or CC-1) (4). Although etiology is not entirely understood, pleural mesothelioma is strongly associated with asbestos exposure. However, the association of asbestos with peritoneal mesothelioma is less established (5). Furthermore, other factors have played a role in the development of pleural mesothelioma, including industrial pollutants and minerals such as silica dust, thorium, and mica, but the association with PM development has not been associated with duration and time of exposure to these materials (6). Our patient reported secondhand exposure to asbestos from his father who worked at a shipyard. It is unclear if this should be considered one of the contributing factors to his development of PM.

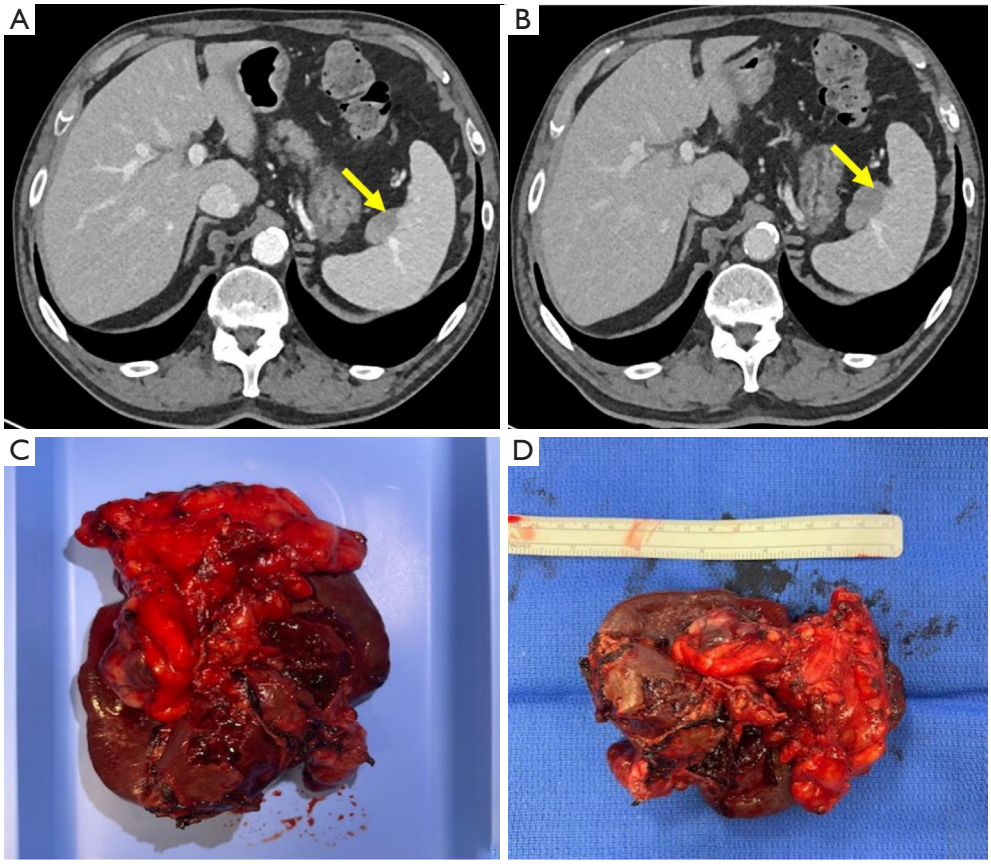

Figure 3 Radiologic and gross imaging of recurrent mass. Computed tomography shows the peri-splenic recurrence at 5 months (A) and at 8 months (B) post-index operation (yellow arrows indicating mass close to hilum). (C,D) Gross intraoperative images of spleen.

Genetics

Different mutations are associated with the development of mesothelioma. The *BAP1* gene is involved in DNA repair, and germline or somatic mutations are often seen in peritoneal mesothelioma. Other somatic genetic mutations include *CDKN2A*, *NF2*, and *TP53*. Families with *BAP1* mutations are at a higher risk of developing mesothelioma with autosomal dominant inheritance, which paradoxically is frequently associated with substantially improved prognosis due to more indolent disease biology (7). The *BAP1* tumor predisposition syndrome is also characterized by uveal and cutaneous melanoma, atypical intradermal tumors, and an increased incidence of renal cell carcinoma (1). Our patient's tumor had somatic mutations in *NF2* and *TP53*. The *NF2* inactivating mutation occurs through truncation of the protein, observed in 20% of peritoneal mesotheliomas. On the other hand, the mutation in *TP53* occurs in the DNA binding domain and has been identified in 7.1% of peritoneal mesotheliomas (8).

Clinical symptoms and histologic findings

Onset is insidious with nonspecific clinical manifestations, such as nausea or change in bowel habits. Some patients have abdominal distention, abdominal pain, intestinal obstruction, and ascites, which usually indicate more advanced disease. Approximately 8% of cases are incidentally diagnosed (9). Our patient presented to the emergency department for right lower quadrant abdominal pain, a vague and nonspecific symptom, without other signs or symptoms.

The World Health Organization (WHO) has histologically classified peritoneal mesothelioma as epithelioid, sarcomatoid, and biphasic subtypes (10). Roughly 75–90% of PM has an epithelioid histology, associated with the best prognosis and therefore the strongest predictor of survival (11,12). This patient had

a localized, well-circumscribed epithelioid PM without evidence of gross parietal or visceral peritoneal spread on initial abdominal exploration.

Localized PM can be solid and/or cystic with an irregular morphology and may invade adjacent structures. Metastases may occur by local infiltration or hematological or lymphatic seeding (13). The characteristic diffuse spread is seen as nodular and irregular thickening of the visceral and parietal peritoneum, adnexa, and omentum. Unifocal or localized peritoneal mesothelioma is scarcely reported in the literature as it almost always presents as multifocal disease. This multifocal presentation frequently involves diffuse peritoneal disease with a seeding appearance, often with ascites. Unifocal or localized presentation has been reported in several case reports, but no definitive guidelines exist regarding management in these instances (14,15). In fact, the presentation of localized mesothelioma often prompts a suspicion of alternative neoplastic diagnoses, such as gastrointestinal stromal tumor or desmoid fibromatosis.

Tumor markers

Serologic markers associated with peritoneal mesothelioma include CA-125, alpha-fetoprotein, carcinoembryonic antigen (CEA), mesothelin, and osteopontin. Yet, these markers are not used as a standard of care practice for diagnosis or treatment response. Furthermore, an elevated CA-125 in women with peritoneal malignancy is often assumed to be secondary carcinomatosis from gynecologic origin. Of note, the baseline value of mesothelin can be an independent predictor of overall survival, with sensitivity of 50% and specificity of 95% (16). When the preoperative levels of these tumor markers are not elevated, a complete surgical resection is achieved in 97% of patients (17). Clinicians variably use these tumor markers to identify progression or detect recurrence after surgical resection. The CA-125 in our patient was not elevated on follow-up.

Diagnostics

Radiology has a crucial role in supporting surgical decision making. The initial and oftentimes preferred imaging modality is CT. This is likely due to the short acquisition time, reproducibility, and feasibility. A CT scan can demonstrate the presence of ascites, omental thickening, and cystic or solid components of the disease (18). CT scans may also be used to assess the radiographic PCI and disease burden on the abdominal wall, mesentery, and small bowel in order to predict ability to optimally cytoreduce disease. Although routinely used for surgical planning, CT scans do have limitations, including poor sensitivity in identifying nodules that are less than 0.5 cm and relative underestimation of radiographic PCI compared to PCI scoring at laparoscopy or laparotomy (19). Magnetic resonance imaging (MRI) may have higher sensitivity and better preoperative PCI prediction capability when compared to CT imaging, as gadolinium contrast and diffusion-weighted imaging often allow for better identification of peritoneal tumors (20,21). However, sensitivity of MRI imaging is more protocol- and institution-dependent than CT. Fluorine-18 fluorodeoxyglucose (^{18}F-FDG)-positron emission tomography-contrast-enhanced CT (PET/CT) has aided in more accurate differentiation between benign lesions of the peritoneum versus malignant peritoneal mesothelioma. The sensitivity and sensitivity of detecting peritoneal lesions is reported to be as high as 87% and 86%, respectively (22). However, detection of lesions requires enough cellular mass to register increased radiotracer uptake. Newer radiotracers such as fibroblast activation protein inhibitor (FAPI) PET/CT may enhance sensitivity and specificity of peritoneal implant detection (23).

Immunohistochemistry

Histologic analysis is essential for diagnostic confirmation. Biopsies are obtained by percutaneous image-guided biopsy, laparoscopy, or laparotomy. Ascites cytology has little diagnostic utility and can often be misleading although is important in prognostic stratification. IHC staining is usually positive for calretinin, vimentin, cytokeratin 5/6 (CK 5/6), and WT-1 (24). Mesothelin is rarely used in current IHC staining modalities due to its high expression in normal tissue; however, it can be used to differentiate between epithelioid and sarcomatoid variants as it is rarely expressed in sarcomatoid mesothelioma (25). Similarly, nuclear loss of BAP1 is a common discriminatory feature for malignant mesothelioma regardless of germline mutation status. This patient's tumor demonstrated characteristic histologic and IHC features of epithelioid mesothelioma (*Figure 2*).

Treatment

The treatment of PM is challenging, often approached using a combination of different modalities including

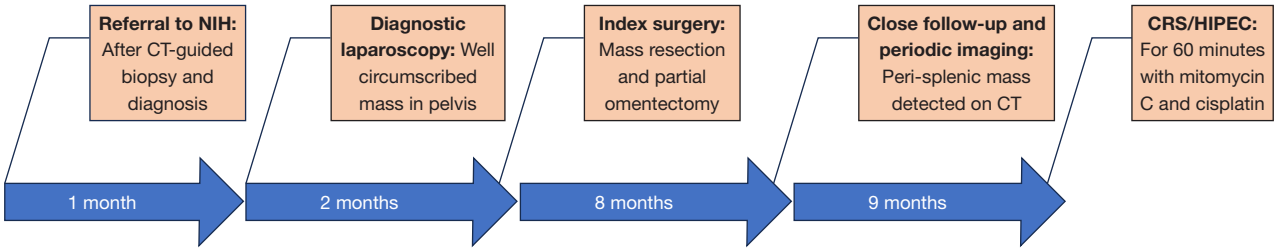

Figure 4 Treatment timeline. Timeline from diagnosis, surgical treatment, follow-up, to subsequent surgery. NIH, National Institutes of Health; CT, computed tomography; CRS, cytoreductive surgery; HIPEC, heated intraperitoneal chemotherapy.

systemic cytotoxic chemotherapy, CRS combined with HIPEC, immunotherapy, and targeted molecular therapy. Chimeric antibody receptor T-cell (CAR-T) therapy against the mesothelin surface antigen has also evolved as a treatment modality for both pleural and peritoneal mesothelioma in treatment-refractory settings after first- and second-line therapies (26). Radiotherapy has little role except for palliative treatment of bulky foci.

CRS and HIPEC

Locoregional treatment of the peritoneal cavity with CRS and HIPEC is commonly employed as a standard treatment for selected patients with peritoneal mesothelioma. The overall objective of CRS is to eliminate all grossly visible tumors, when possible (CC-0), or to cytoreduce disease foci down to no greater than 2.5 mm (CC-1). Following CRS, high-dose chemotherapy is delivered to treat microscopic foci of disease as well as any small residual tumors. Platinum-based regimens, namely cisplatin, are most effective and perfused at a supraphysiologic temperature of 41 to 43 ℃, which enhances cytotoxicity. This can be performed using either an open (coliseum) or a closed technique. Long-term outcomes after CRS and HIPEC for diffuse PM include 5-year overall survivals ranging from 47% in a large multi-institutional US cohort to 61% in a smaller Canadian cohort (27,28). Most patients demonstrated uni-cavitary disease with epithelioid histology, which is the basis for National Comprehensive Cancer Network guidelines recommending CRS and HIPEC (29). Timing is of the essence when it comes to referral for CRS-HIPEC eligible patients. The highest average life expectancy after initial diagnosis for patients eligible for CRS-HIPEC who received surgery, had delayed surgery, and who did not receive surgery were 5.24-, 4.80-, and 2.11-year overall survival respectively (30). In addition, sarcomatoid and biphasic histological subtypes have an aggressive infiltrative growth pattern leading to a potentially rapid disease progression. This in turn may impact the PCI from the time of the initial diagnostic laparoscopy (10). Despite the advantages of performing a diagnostic laparoscopy and its relative low complication rate technical limitations exist in the form of incomplete visualization of mesenteric, retro-hepatic, retro-splenic, and retroperitoneal peritoneum impacting PCI assessment (31).

It should be clarified that recommendations for CRS and HIPEC in the management of peritoneal mesothelioma apply almost universally to diffuse PM, whereas specific guidelines regarding HIPEC in the management of unifocal peritoneal mesothelioma do not exist. These cases should be managed with patient-specific factors in mind, incorporating the risks of HIPEC with the benefits of potentially mitigated peritoneal disease recurrence.

In our index procedure we opted to have a more nuanced approach to his unifocal lesion by excluding HIPEC. Detailed counselling occurred with the patient considering his older age, personal preference for a less extensive surgery, lack of gross peritoneal spread, and negative biopsies from our empiric four-quadrant peritoneal biopsies on diagnostic laparoscopy. In addition, chemotherapy may have enhanced morbidity without certain benefit. There was clear agreement with the patient that HIPEC would be part of the plan should he have future recurrence. Careful monitoring for recurrence was then obtained. It is unclear why the patient developed a rather remote recurrence compared to the original disease focus tracking from the original spot to omentum to splenic hilum (*Figure 4*). Future directions of investigation include to evaluate and risk-stratify patients molecularly for the potential benefit of HIPEC in patients with localized unifocal peritoneal mesothelioma.

Systemic chemotherapy

The overall efficacy of cisplatin or carboplatin monotherapy is historically poor, even in combination with gemcitabine (32). A subsequent phase III clinical trial showed an increase in median overall survival of 12.1 months in patients who revived pemetrexed combined with cisplatin when compared to patients who received cisplatin alone at 9.3 months (33). This combination, with the addition of a monoclonal antibody bevacizumab, has been associated with a statistically significant overall survival (P=0.017) (34).

Immunotherapy

Immunotherapy has recently emerged as a promising approach with demonstrable effect in PM treatment (35). As with earlier regimens, data indicating efficacy of checkpoint inhibitors in peritoneal mesothelioma originates from the numerous clinical trials in pleural mesothelioma, including the landmark CheckMate 743 trial where dual checkpoint therapy outperformed chemotherapy with an overall survival benefit of 18.1 *vs.* 14.1 months [hazard ratio (HR) =0.74, P=0.002] in previously untreated, unresectable pleural mesothelioma (36). For this reason, dual checkpoint therapy with ipilimumab (anti-CTLA4) and nivolumab (anti-PD-1) is an approved first-line regimen for peritoneal mesothelioma and is preferred for biphasic and sarcomatoid histologic variants, likely due to the fact that these subtypes are less likely to respond to chemotherapy (29).

More recently, efforts have been made to deploy CAR-T cells in the management of treatment-refractory pleural and peritoneal mesothelioma, albeit with only modest activity (26). Anti-mesothelin CAR-T cell therapy trials have demonstrated some efficacy in mesothelioma (37). CAR-T cell therapy continues to evolve, more recently with the development of T-cell receptor fusion construct (TRuC) regimens, as do efforts to develop anti-mesothelin adoptive cell transfer treatment approaches.

Other targeted therapies

The most common alternative targeted therapy utilized in malignant peritoneal mesothelioma is the anti-angiogenic monoclonal antibody, bevacizumab (anti-VEGF). This agent is most often used in combination with pemetrexed and platinum-based chemotherapy and its utility in PM has been extrapolated from clinical trials in pleural mesothelioma. Although the MAPS study demonstrated an overall survival benefit of 18.8 *vs.* 16.1 months (HR =0.77, P=0.017) for bevacizumab combined with pemetrexed and cisplatin compared to pemetrexed and cisplatin alone in previously untreated malignant pleural mesothelioma, the clinical significance and correlative benefit for peritoneal primaries remains uncertain (38). Other targeted therapies have limited utility in peritoneal mesothelioma.

Conclusions

Our case demonstrates a rare presentation of unifocal peritoneal mesothelioma. The diagnosis of a solitary peritoneal mass should prompt consideration of mesothelioma as part of a broader differential. As treatment of the peritoneal cavity with CRS and HIPEC is commonly employed as a standard treatment, the management of unifocal PM should be tailored to the clinical presentation and patients' characteristics. Further studies and corresponding treatment guideline updates are needed.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://jgo.amegroups.com/article/view/10.21037/jgo-24-266/rc

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Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://jgo.amegroups.com/article/view/10.21037/jgo-24-266/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised

in 2013). Written informed consent was obtained from the patient for the publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

References

1. Hung YP, Dong F, Torre M, et al. Molecular characterization of diffuse malignant peritoneal mesothelioma. Mod Pathol 2020;33:2269-79.
2. Dahdaleh FS, Sherman SK, Witmer HDD, et al. Potential evidence of peritoneal recurrence in Stage-II colon cancer from the control arm of CALGB9581. Am J Surg 2022;224:459-64.
3. Gregory SN, Sarvestani AL, Blakely AM. Malignant peritoneal mesothelioma literature review: past, present, and future. Dig Med Res 2022;5:29.
4. Helm JH, Miura JT, Glenn JA, et al. Cytoreductive surgery and hyperthermic intraperitoneal chemotherapy for malignant peritoneal mesothelioma: a systematic review and meta-analysis. Ann Surg Oncol 2015;22:1686-93.
5. Zona A, Fazzo L, Minelli G, et al. Peritoneal mesothelioma mortality in Italy: Spatial analysis and search for asbestos exposure sources. Cancer Epidemiol 2019;60:162-7.
6. Gao Y, Mazurek JM, Li Y, et al. Industry, occupation, and exposure history of mesothelioma patients in the U.S. National Mesothelioma Virtual Bank, 2006-2022. Environ Res 2023;230:115085.
7. Maccaroni E, Giampieri R, Lenci E, et al. BRCA mutations and gastrointestinal cancers: When to expect the unexpected? World J Clin Oncol 2021;12:565-80.
8. Kim JE, Kim D, Hong YS, et al. Mutational Profiling of Malignant Mesothelioma Revealed Potential Therapeutic Targets in EGFR and NRAS. Transl Oncol 2018;11:268-74.
9. Sun L, Li C, Gao S. Diffuse malignant peritoneal mesothelioma: A review. Front Surg 2023;9:1015884.
10. Kim J, Bhagwandin S, Labow DM. Malignant peritoneal mesothelioma: a review. Ann Transl Med 2017;5:236.
11. Pezzuto F, Vimercati L, Fortarezza F, et al. Evaluation of prognostic histological parameters proposed for pleural mesothelioma in diffuse malignant peritoneal mesothelioma. A short report. Diagn Pathol 2021;16:64.
12. Galateau-Salle F, Churg A, Roggli V, et al. The 2015 World Health Organization Classification of Tumors of the Pleura: Advances since the 2004 Classification. J Thorac Oncol 2016;11:142-54.
13. Cortés-Guiral D, Hübner M, Alyami M, et al. Primary and metastatic peritoneal surface malignancies. Nat Rev Dis Primers 2021;7:91.
14. Saisho K, Fujiwara S, Anami K, et al. Localized biphasic malignant peritoneal mesothelioma presenting as a rectal tumor. Clin J Gastroenterol 2020;13:308-15.
15. Sugarbaker PH. Malignant peritoneal mesothelioma presenting as a mass in a Spigelian hernia. Report of a case. Int J Surg Case Rep 2020;68:239-41.
16. Cheng X, Gou HF, Liu JY, et al. Clinical significance of serum CA125 in diffuse malignant mesothelioma. Springerplus 2016;5:368.
17. van der Bij S, Schaake E, Koffijberg H, et al. Markers for the non-invasive diagnosis of mesothelioma: a systematic review. Br J Cancer 2011;104:1325-33.
18. Park JY, Kim KW, Kwon HJ, et al. Peritoneal mesotheliomas: clinicopathologic features, CT findings, and differential diagnosis. AJR Am J Roentgenol 2008;191:814-25.
19. Salo SAS, Lantto E, Robinson E, et al. Prognostic role of radiological peritoneal cancer index in malignant peritoneal mesothelioma: national cohort study. Sci Rep 2020;10:13257.
20. Low RN, Barone RM, Lucero J. Comparison of MRI and CT for predicting the Peritoneal Cancer Index (PCI) preoperatively in patients being considered for cytoreductive surgical procedures. Ann Surg Oncol 2015;22:1708-15.
21. Chandramohan A, Shah N, Thrower A, et al. Communicating imaging findings in peritoneal mesothelioma: the impact of 'PAUSE' on surgical decision-making. Insights Imaging 2021;12:174.
22. Dubreuil J, Giammarile F, Rousset P, et al. The role of 18F-FDG-PET/ceCT in peritoneal mesothelioma. Nucl Med Commun 2017;38:312-8.
23. Yang T, Ma L, Hou H, et al. FAPI PET/CT in the Diagnosis of Abdominal and Pelvic Tumors. Front Oncol 2022;11:797960.
24. Husain AN, Colby TV, Ordóñez NG, et al. Guidelines

for Pathologic Diagnosis of Malignant Mesothelioma 2017 Update of the Consensus Statement From the International Mesothelioma Interest Group. Arch Pathol Lab Med 2018;142:89-108.
25. Ordóñez NG. Application of immunohistochemistry in the diagnosis of epithelioid mesothelioma: a review and update. Hum Pathol 2013;44:1-19.
26. Castelletti L, Yeo D, van Zandwijk N, et al. Anti-Mesothelin CAR T cell therapy for malignant mesothelioma. Biomark Res 2021;9:11.
27. Deban M, Taqi K, Knapp GC, et al. Cytoreductive surgery (CRS) and heated intraperitoneal chemotherapy (HIPEC) for peritoneal mesothelioma: Canadian practices and outcomes. J Surg Oncol 2023;128:595-603.
28. Valenzuela CD, Solsky IB, Erali RA, et al. Long-Term Survival in Patients Treated with Cytoreduction and Heated Intraperitoneal Chemotherapy for Peritoneal Mesothelioma at a Single High-Volume Center. Ann Surg Oncol 2023;30:2666-75.
29. NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Guideline Mesothelioma: Peritoneal 2024 © National Comprehensive Cancer Network, Inc. 2024. All rights reserved. Available online: https://www.nccn.org/guidelines/guidelines-detail?category=1&id=1513
30. Rouhi AD, Choudhury RA, Hoeltzel GD, et al. Delayed CRS-HIPEC Is Associated with Decreased Survival in Patients with Malignant Peritoneal Mesothelioma: A Markov Decision Analysis. J Gastrointest Surg 2023;27:995-7.
31. Yurttas C, Überrück L, Nadiradze G, et al. Limitations of laparoscopy to assess the peritoneal cancer index and eligibility for cytoreductive surgery with HIPEC in peritoneal metastasis. Langenbecks Arch Surg 2022;407:1667-75.
32. Lurvink RJ, Villeneuve L, Govaerts K, et al. The Delphi and GRADE methodology used in the PSOGI 2018 consensus statement on Pseudomyxoma Peritonei and Peritoneal Mesothelioma. Eur J Surg Oncol 2021;47:4-10.
33. Vogelzang NJ, Rusthoven JJ, Symanowski J, et al. Phase III Study of Pemetrexed in Combination With Cisplatin Versus Cisplatin Alone in Patients With Malignant Pleural Mesothelioma. J Clin Oncol 2023;41:2125-33.
34. Zalcman G, Mazieres J, Margery J, et al. Bevacizumab for newly diagnosed pleural mesothelioma in the Mesothelioma Avastin Cisplatin Pemetrexed Study (MAPS): a randomised, controlled, open-label, phase 3 trial. Lancet 2016;387:1405-14.
35. Raghav K, Liu S, Overman M, et al. Clinical Efficacy of Immune Checkpoint Inhibitors in Patients With Advanced Malignant Peritoneal Mesothelioma. JAMA Netw Open 2021;4:e2119934.
36. Baas P, Scherpereel A, Nowak AK, et al. First-line nivolumab plus ipilimumab in unresectable malignant pleural mesothelioma (CheckMate 743): a multicentre, randomised, open-label, phase 3 trial. Lancet 2021;397:375-86.
37. Beatty GL, Haas AR, Maus MV, et al. Mesothelin-specific chimeric antigen receptor mRNA-engineered T cells induce anti-tumor activity in solid malignancies. Cancer Immunol Res 2014;2:112-20.
38. Bibby AC, Maskell NA. Current treatments and trials in malignant pleural mesothelioma. Clin Respir J 2018;12:2161-9.

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