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## Case Report

### A Forgotten Disease – Peritoneal Mesothelioma in an Older Adult

Hsi-Yang Chen<sup>a,d</sup>, Wei-Chen Lin<sup>b,c,d</sup>, Ming-Jen Chen<sup>b,c,d</sup>, Chia-Yuan Liu<sup>b,d</sup>, Chen-Wang Chang<sup>b,c,d\*</sup>

<sup>a</sup> Division of Pulmonary and Critical Care Medicine, Department of Internal Medicine, MacKay Memorial Hospital, Taipei, Taiwan, <sup>b</sup> Division of Gastroenterology, Department of Internal Medicine, MacKay Memorial Hospital, Taipei, Taiwan, <sup>c</sup> MacKay Junior College of Medicine, Nursing and Management, Taipei, Taiwan, <sup>d</sup> MacKay Medical College, New Taipei City, Taiwan

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#### SUMMARY

Mesothelioma is a rare malignancy often observed in the pleura of patients exposed to asbestos. Peritoneal mesothelioma is a rare malignancy accounting for less than 20% of all mesothelioma cases. Many older adults might have been exposed to asbestos in the past; therefore, they could still develop mesothelioma later. Due to its rarity and nonspecific presentation, peritoneal mesothelioma is often overlooked.

We present the case of a 64-year-old man who had been exposed to asbestos for two year 40 years ago because he lived in an asbestos factory. He visited our emergency room because of acute abdominal pain for one day. Abdominal computed tomography showed many ascites with an irregularly thickened peritoneum, and ascites without portal hypertension were suspected. Esophagogastroduodenoscopy, small-bowel capsule endoscopy, and colonoscopy revealed no organic lesions. Diagnostic laparotomy revealed uniform epithelioid tumor cells with eosinophilic cytoplasm and a predominant tubular pattern infiltrating the peritoneal stroma. Therefore, peritoneal mesothelioma was diagnosed, and the patient received cytoreduction plus hyperthermic intraperitoneal chemotherapy (HIPEC) with a stable condition.

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## 1. Introduction

Mesothelioma is a malignancy with an inferior prognosis due to delayed diagnosis and its dissemination in the peritoneum and mesentery, which makes curative resection extremely difficult. Geographic and temporal variations in diagnostic practices confound the epidemiology of peritoneal mesothelioma.<sup>1</sup> In Taiwan, asbestos consumption was high in the 1980s. The age-standardized incidence rate of malign pleural mesothelioma from 2009 to 2013 was 0.28 per 100,000 person-years in males and 0.11 per 100,000 person-years in females.<sup>2</sup> Peritoneal mesothelioma is a rare malignancy accounting for less than 20% of all mesothelioma cases. Most patients did not undergo a thorough examination until ascites developed. Typical clinical presentation includes mild abdominal pain and ascites. Other symptoms, including asthenia, weight loss, anorexia, and abdominal mass, were reported in less than 50% of all cases at initial presentation.<sup>3</sup>

## 2. Case report

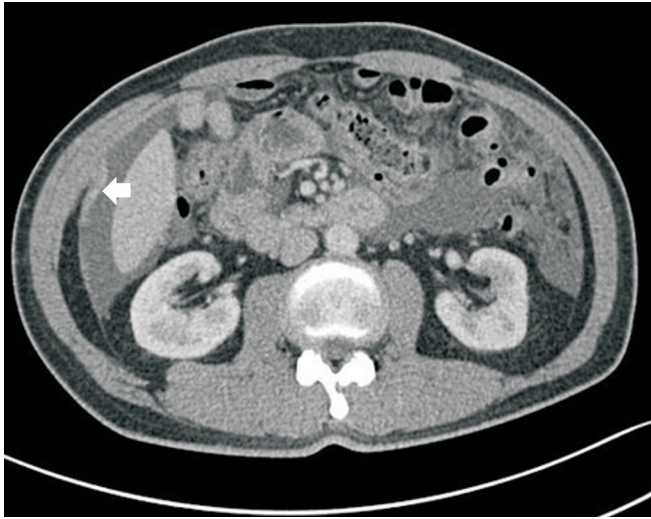
A 64-year-old man presented with hypertension that did not require medical treatment and a history of *Helicobacter pylori* infection post eradication fifteen years ago and visited our emergency room because of periumbilical pain for one day. He stated that this periumbilical pain had been on and off for three years. The pain was dull, numerical rating scale of 3–5/10, usually self-limiting, and re-

sponded well to analgesics. Therefore, he visited the emergency room several times over the past three years. He had a normal appetite and denied involuntary body weight loss, difficulty swallowing, or change in bowel habits. Blood tests revealed no abnormalities. Abdominal computed tomography showed a moderate amount of ascites, with some parts of the peritoneum being irregularly thickened (Figure 1). Paracentesis showed the ascites were yellow and cloudy in appearance, with a high specific gravity of 1.028. The white blood cell was 5,760/CMM, comprising 87% of lymphocytes and 0% of neutrophils. The data for ascites albumin, total protein, sugar, LDH, and amylase were unavailable because the ascites were too thick for laboratory examination. Peritoneal carcinomatosis and TB peritonitis were initially diagnosed. The ascites cytology test revealed atypical cells only. Therefore diagnostic laparoscopy was performed and revealed multiple nodular lesions in the omentum, peritoneum, right abdominal wall, and subdiaphragmatic area (Figure 2). A biopsy of the omentum and peritoneum showed peritoneum tissue with invasive epithelioid mesothelioma composed of malignant mesothelial cells with a predominant tubular pattern that infiltrated the peritoneal stroma. Therefore, invasive epithelioid mesothelioma was diagnosed (Figure 3). Tracing back his history, he lived in an asbestos factory for two years when he was in his twenties. The patient underwent cytoreduction and HIPEC using cisplatin and mitomycin. He tolerated the treatment well and was discharged after he could eat.

## 3. Discussion

Peritoneal mesothelioma diagnosis is challenging, despite its

\* Corresponding author. Division of Gastroenterology, Department of Internal Medicine, MacKay Memorial Hospital, No. 92, Sec. 2, Chung-Shan N. Road, Taipei, Taiwan.  
E-mail address: [mky378@yahoo.com.tw](mailto:mky378@yahoo.com.tw) (C.-W. Chang)



**Figure 1.** Abdominal computed tomography showed a moderate amount of ascites, with some part of the peritoneum being irregularly thickened (arrow).



**Figure 2.** Diagnostic laparoscopy found multiple nodular lesions on the surface of the peritoneum.

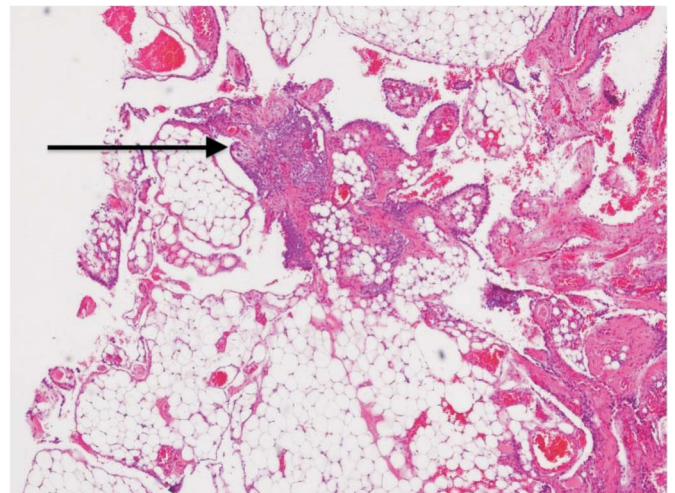
characteristic symptoms. In a multicenter study of 81 patients, the most common presentations were ascites (77%) and abdominal pain (66%), followed by asthenia (43%), weight loss (32%), anorexia (30%), abdominal mass (30%), fever (22%), diarrhea (17%), and vomiting (15%). However, only 51% of patients had a positive peritoneal fluid cytology for mesothelioma. Laparoscopy and laparotomy were the main diagnostic procedures, performed in 49% and 44% of cases, respectively. On average, peritoneal mesothelioma was diagnosed 4 to 6 months after the initial presentation.<sup>3</sup>

Peritoneal mesothelioma is diagnosed by exploratory laparotomy, according to the PSOGI/EURACAN clinical practice guidelines.<sup>4</sup> The guidelines mentions that core needle biopsy is acceptable, while fine needle aspiration and ascites cytological examination are both non-diagnostic for several reasons. Cytological examination of ascites can reveal atypical cells.

Mesothelioma has an overall poor prognosis, with a median survival of 4–18 months after diagnosis. However, this is partly because it is most often diagnosed at a later stage.

The completeness of cytoreduction (CCR) was assessed using the CCR score. The index that quantifies the extent of residual disease at the end of the procedure is classified into 4 categories: CCR0 (no macroscopic residual disease); CCR1 (residual tumours less than 2.5 mm); CCR2 (residual tumours between 2.5 mm and 2.5 cm); CCR3 (residual tumours greater than 2.5 cm). CCR0 and CCR1 represent complete cytoreduction, while CCR2 and CCR3 are designed as incomplete cytoreduction. Yan et al. detailed a case series demonstrating that the median survival was 94, 67, 40, and 12 months for CCR 0, 1, 2, and 3, respectively.<sup>5</sup> Magge et al. also reported a median overall survival of 56.7 months in those with a complete cytoreduction and 7.4 months in those with an incomplete cytoreduction.<sup>6</sup>

Early detection of the disease is important. Asbestos has been banned since the early 1990s, and people exposed to it are mostly older adults. Asbestos exposure history should be checked in older patients presenting with abdominal pain or ascites of unknown origin for the early diagnosis of peritoneal mesothelioma. This raises the question of whether there is an optimal protocol for screening the population at risk of developing peritoneal mesothelioma. Nonetheless, it is difficult in the first place to identify the population at risk because only 33–50% of patients diagnosed with peritoneal mesothelioma reported knowing prior exposure to asbestos.<sup>1</sup> Also, none of the imaging studies have shown reliable sensitivity to detect



**Figure 3.** The black arrow points out where the mesothelioma grows beyond the normal mesothelium lining into the adjacent adipose tissue.

peritoneal mesothelioma.

Despite the lack of reliable screening tools for peritoneal mesothelioma, the PSOGI/EURACAN clinical practice guidelines for peritoneal mesothelioma recommends that individuals with current or prior history of asbestos exposure should undergo a screening program with an abdominal ultrasound every year to improve early detection of peritoneal mesothelioma.<sup>4</sup> However, the level of evidence is low, and only 10 of 27 experts (37%) agreed with this consensus. Considering that abdominal ultrasound is relatively non-invasive and has a lower cost than other imaging studies, screening with abdominal ultrasound annually should still be considered.

Cytoreduction plus hyperthermic intraperitoneal chemotherapy (HIPEC) remains the standard treatment of choice according to the PSOGI/EURACAN clinical practice guidelines for peritoneal mesothelioma.<sup>4</sup> The standard treatment with better-quality evidence is cytoreduction plus HIPEC. Diffuse malignant peritoneal mesothelioma is characterized by widespread dissemination of chemoresistant metastatic nodules within the peritoneal cavity. The rarity of systemic lesions suggests that diffuse malignant peritoneal mesothelioma is a locoregional disease, making it a model for the development of comprehensive locoregional radical approaches, such as

the combination of complete cytoreductive surgery with HIPEC. This treatment is associated with the best long-term outcomes and is recommended by international guidelines in selected patients.<sup>7</sup>

The main prognostic factors are the histological features and adequacy of cytoreduction. Epithelioid mesothelioma has a better prognosis than sarcomatoid or biphasic mesothelioma. The median survival of patients with the epithelial type was 63 months, whereas those with a combination of biphasic and sarcomatoid types had a median survival of 16 months. CCR0/1 has a better outcome than CCR2/3: median survival was 94, 67, 40, and 12 months for CCR 0, 1, 2, and 3, respectively.<sup>8</sup> Inoperable patients may benefit from palliative chemotherapy with the regimen of platinum-based chemotherapy, but these data are difficult to interpret due to their rarity and the lack of randomized control studies.<sup>9,10</sup>

#### 4. Conclusion

Peritoneal mesothelioma is rare, but its incidence rate could still rise in the upcoming decade, considering that gradual asbestos removal from the environment takes time and effort. Many older adults might have been exposed to asbestos sometime in the past; therefore, they could still develop mesothelioma later. Because of its non-specific clinical symptoms, peritoneal mesothelioma can be misdiagnosed as functional ileus, chronic constipation, or simply neglected.

#### Conflicts of interests

None of the authors have any conflicts of interest to declare.

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None.

#### Authors' contributions

I declare that I have participated in the preparation of the article "A Forgotten Disease – Peritoneal Mesothelioma in An Elderly". Hsi-Yang Chen wrote this article. Lin WC, CY Liu and Chen MJ con-

ducted the literature review. Chang CW supported this work by performing a critical reading of the manuscript and supervising the final editing. All authors read and approved the final manuscript.

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