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Tumor Discovery

CASE REPORT

Sarcomatoid mesothelioma presenting as a mediastinal mass: A case report

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Abstract

Sarcomatoid mesothelioma (SM) is a subtype of mesothelioma, a deadly cancer strongly related to asbestos exposure. Herein, we present the case of a 50-year-old female who presented with back pain and non-specific symptoms for approximately 9 months. The diagnostic investigation revealed a large, centrally necrotic mass in the thorax that appeared to arise from the mediastinum and surrounded vital structures, including the superior vena cava. A biopsy of the thoracic mass revealed SM. The patient developed hematemesis, and emergent endoscopy revealed a gastric ulcer composed of malignant SM cells, a rare finding in pleural mesothelioma. Her disease was too advanced for surgical resection and conventional therapy. She tragically died weeks after diagnosis. The findings of this report highlight the varied presentation of SM to underscore the importance of early diagnosis and treatment of patients in improving overall survival.

Keywords: Mediastinal mass; Dyspnea; Back pain; Sarcomatoid mesothelioma; Lung cancer

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1. Introduction

Sarcomatoid mesothelioma (SM) is a subtype of mesothelioma, a rare cancer arising from mesothelial surfaces of tissues – most commonly the pleura – and is directly related to asbestos exposure.[1] Asbestos-related diseases have long latency periods, so consequences of exposure tend to be detectable approximately 40 years after initial contact. Due to this, many patients first present in their fifth or sixth decades of life with late-stage disease, often with signs of progressing respiratory failure, making treatment challenging.[2,3] Mesothelioma is classified into three main subtypes based on histology: epithelioid, sarcomatoid, and biphasic. SM is the least common but most aggressive of these classes, with the lowest median survival time of approximately 3.5 – 7.4 months.[2,4,5] Herein, we report a case of advanced SM with an atypical presentation due to a heavy burden of locally invasive disease and extrathoracic metastases. This case report aims to highlight the importance of early diagnosis and treatment in patients with SM with the aim of improving their overall survival.

2. Case presentation

A 50-year-old female with diabetes presented with acute-on-chronic right-sided upper back pain. Nine months before presentation, the patient experienced intermittent right-sided back pain without an identifiable trigger. Subsequently, non-productive cough and dyspnea on exertion developed 4 months before presentation. Her symptoms progressed, and she developed progressively worsening anorexia and nausea, as well as experiencing 20 pounds of unintentional weight loss over 2 weeks, all of which prompted her to seek medical consultation and her admission to the hospital. Fevers, chills, and chest pain were absent. She was a non-smoker with no history of secondhand smoke exposure. According to her, multiple family members had succumbed to an unknown lung cancer.

Physical examination revealed tachycardia and decreased breath sounds over the middle and lower lobes of the right lung. Routine hematology was notable for mild neutrophilic leukocytosis and thrombocytosis. Additional metabolic and infectious testing was unrevealing. A chest radiograph showed a large right-sided pleural effusion with compressive atelectasis. Computed tomography (CT) of the chest redemonstrated the effusion and revealed a 14.5 × 6.9 cm centrally necrotic right hemithorax mass arising from the mediastinum with invasion into the adjacent anterior chest wall and compression of the superior vena cava and right interlobar and upper lobe bronchi (Figure 1).

The patient developed hypoxemia. Subsequently, a chest tube was placed, and cytologic examination revealed an exudative pleural effusion with reactive mesothelial cells. Bacterial, fungal, and acid-fast bacilli cultures were negative. A biopsy of the mediastinal mass showed a spindle cell neoplasm composed of large, highly atypical pleomorphic spindle cells with irregular hyperchromatic nuclei. Immunohistochemical tests showed that the biopsied mass was positive for D2-40, calretinin, and WT-1, revealing SM. On further investigation of environmental exposures, the patient and her family reported prior residence next to a large asbestos plant in Central America with frequent exposure to friends and family employed at the plant, often with asbestos fibers on their clothing.

Given the proximity of the mass to key thoracic structures, it was deemed unresectable, and the patient was to start chemotherapy at discharge. However, she suddenly developed large-volume hematemesis and an emergent endoscopy was immediately conducted, revealing a malignant gastric ulcer, which was confirmed on biopsy to be metastatic SM (Figure 2). Shortly thereafter, she developed a pulmonary embolism with right heart strain and respiratory failure. Her condition progressed rapidly to obstructive shock, ultimately causing her death.

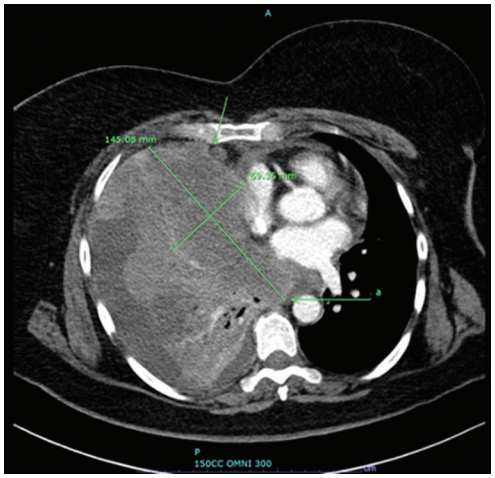


Figure 1. Computed tomography of the thorax in the axial plane shows a 14.5 × 6.9 cm mass arising from the mediastinum, which invades the anterior chest wall and causes significant superior vena cava stenosis.

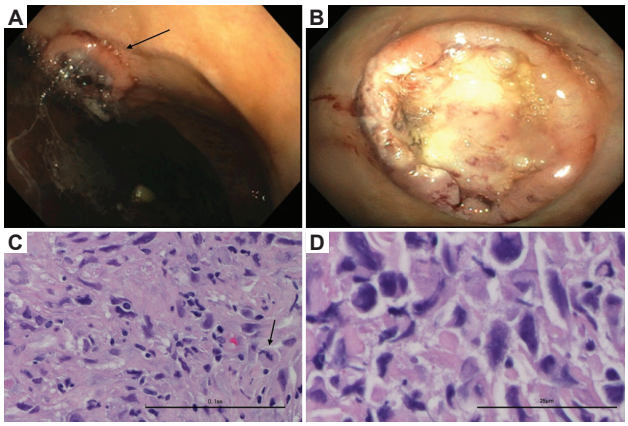


Figure 2. Pleural mesothelioma is complicated by a metastatic gastric ulcer. (A) Gastric body ulcer visualized endoscopically (black arrow). (B) Enhanced view of the ulcer. (C) Hematoxylin-eosin (H&E) staining of biopsied gastric cancer specimens shows cells with pleomorphism with the absence of normal gastric parenchyma. Mitotic figures are seen (black arrow). Scale bar: 0.1 mm (×10 magnification). (D) Higher-magnification view of H&E-stained malignant cells with enlarged irregular nuclei. Malignant cells are positive for pancytokeratin, D2-40, WT1 (in a subset of cells), CAM5.2, CK5&6 (rare cells), and calretinin (in a small subset of cells), but they are negative for P63, S100, beta-catenin, desmin, EMA, MOC31, and CD34. The histology and immunostaining pattern are consistent with the histopathologic features of metastatic mesothelioma. Scale bar: 0.025 mm (×40 magnification).

3. Discussion

Globally, the incidence rate of mesothelioma has declined over the last few decades following nationwide bans on asbestos in many countries, including the United States and most European countries.[1,6] However, asbestos production

and commercialization still occur widely in Asia and many Central and South American countries.[7-9] In a highly globalized world with accessible travel, it is critical for physicians to recognize and manage mesothelioma, even if asbestos production is limited in their home nation.

Initial symptoms of SM are often non-specific, most commonly cough and dyspnea. Many patients are incidentally found to have pleural effusions on imaging. Chest wall or back pain are less commonly described in SM, though they were presenting features in our case.[3,10] Much of the morbidity and mortality associated with SM is due to local disease progression and invasion of critical structures in the mediastinum.[2] Previously believed to be rare, tumor dissemination in extrapleural and extrathoracic sites is common, occurring in 87.7% and 55.4% of patients, respectively.[11] In lung cancer, however, metastasis to the gastrointestinal tract is exceptionally uncommon, noted in only ~0.5% of cases.[12] In pleural mesothelioma specifically, metastasis to the gastrointestinal lumen in the absence of peritoneal disease is rare, with <20 cases reported in the literature thus far. Most of these cases were caused by epithelioid mesothelioma, and three resulted from SM. Only one case reported gastric ulcers from metastatic SM,[13] similar to our case.

Challenges in diagnosing SM include difficulty in differentiating it from other spindle cell neoplasms and inconsistent expression of the common tumor markers used in immunohistochemistry.[13,14] On biopsy, SM is characterized by spindle-shaped cells and a lack of the typical glandular structures seen in other types of mesothelioma.[15] However, SM may mimic other neoplasms depending on the anatomic site involved, namely sarcomatoid carcinoma and intraabdominal synovial sarcoma, which share a similar histomorphology.[13] Common mesothelial markers for immunohistochemical studies include calretinin, D2-40, WT-1, and cytokeratin. For SM specifically, staining is typically positive for cytokeratin and calretinin, but there have been rare cases with negative calretinin expression, making diagnosis additionally challenging.[14,16]

Management of SM centers around surgical resection of the tumor and chemotherapy. For resectable tumors, surgery lessens the tumor burden, relieving dyspnea and reducing the occurrence of pleural effusions.[14] Neoadjuvant or adjuvant chemotherapy with cisplatin and pemetrexed is the only Food and Drug Administration-approved combination for the treatment of pleural mesothelioma and remains the sole treatment modality for patients in the United States who are either not candidates for surgery or have unresectable disease.[2,14] Globally, immune checkpoint inhibitors, such as nivolumab, and anti-angiogenic drugs, such as bevacizumab, have shown efficacy against malignant pleural mesothelioma.[17] Unfortunately, Mansfield *et al.*[18] found that compared to patients with other subtypes, fewer patients with SM have complete and partial responses to treatment, with response rates recorded at 13.9% and 21.9%, respectively. Novel research into alternative treatment modalities, including trimodal therapy with surgery, chemotherapy, and adjuvant radiotherapy, as well as molecular targeted therapy, is needed to improve outcomes for patients with SM.[14]

4. Conclusion

SM is the rarest main form of mesothelioma, a disease that has remained unknown to the public for a long time due to international governmental policies limiting asbestos production and use. The long latency period since exposure, non-specific symptom manifestation, and challenges in diagnostics lead to the majority of patients presenting with late-stage disease upon diagnosis. During the terminal stage of the disease, local invasion of the tumor has typically become too extensive for surgical resection, and conventional chemotherapy options have proven unsuccessful in managing SM. Therefore, clinicians must have high diagnostic suspicion for mesothelioma, particularly SM, when encountering patients originating from asbestos-producing regions to ensure prompt diagnosis and treatment and to optimize patient outcomes.

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Conflict of interest

The authors declare no conflicts of interest.

Author contributions

Conceptualization: Elizabeth Chiang, Alec B. Rezigh
Investigation: Elizabeth Chiang, Alec B. Rezigh
Methodology: Elizabeth Chiang
Writing – original draft: Elizabeth Chiang
Writing – review & editing: All authors

Ethics approval and consent to participate

The manuscript and patient data were written and collected in accordance with the declaration of Helsinki and our institutional ethics guidelines.

Consent for publication

We have obtained informed verbal consent from the patient for the publication of this report. The authors have

obtained permission to reprint any figures or tables that were initially printed elsewhere, should they be required.

Availability of data

Data are available from the corresponding author upon reasonable request.

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