A Case of Systemic Lupus Erythematosus with Malignant Pleural Mesothelioma in a 42-year Woman

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ABSTRACT

Systemic lupus erythematosus (SLE) is an autoimmune connective tissue disease characterised by inflammation. Malignant pleural mesothelioma (MPM) is a highly invasive malignant tumor derived from pleural mesothelial cells. Here, we report a case of SLE with MPM. A 42-year woman with no exposure to asbestos presented with severe left chest pain. Initially, we diagnosed her with SLE because of the clinical manifestations and high antinuclear antibody titer. Finally, a diagnosis of MPM was made, based on pleural biopsy. Her condition was under control after one cycle of chemotherapy and oral methotrexate. However, three years later, she was admitted with dyspnea, mild orthopnea, and tachycardia, and died one month later after discontinuing treatment. MPM is rare, and MPM with SLE is even rarer. We should pay attention to pleural effusion when diagnosing SLE. If possible, a pleural biopsy should be performed to reduce misdiagnosis and missed diagnosis.

Key Words: Pleural effusion, Systemic lupus erythematosus (SLE), Mesothelioma.

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INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune connective tissue disease characterised by inflammation. The presence of multiple autoantibodies and multi-system involvement are two major clinical features of SLE. Malignant pleural mesothelioma (MPM) is a highly invasive malignant tumor derived from pleural mesothelial cells that accounts for 0.02-0.04% of all malignant tumors.^{1,2} Its morbidity has recently increased.³ Here, we report a case of SLE with MPM in a 42-year non-smoking woman with no history of exposure to asbestos.

CASE REPORT

A 42-year non-smoking woman was admitted to the hospital because of a 10-day history of left chest pain. The patient had not been exposed to asbestos. She had experienced intermittent bilateral knee joint pain for more than 10 years, bilateral interphalangeal joint pain for more than 2 years, and occasional sensation of dry mouth for more than 2 months. Her blood tests and blood cell count were unremarkable, except for an above normal erythrocyte sedimentation rate (68 mm/1st h), higher neutrophil percentage (88.1%), raised C-reactive protein (97.72 mg/L), and mild anemia (hemoglobin 82 g/L). Blood test results were also positive for anti-nuclear antibodies (Table I).

Chest contrast-enhanced computed tomography (CT) showed enlargement of the mediastinum and bilateral axillary lymph nodes, small nodules in the subpleural lobe of the right lung, left pleural effusion, left lower lobe atelectasis, and right lobe, upper lobe tongue and lower lobe lesions (Figure 1 a, b).

A moderate left-sided pleural effusion, a small amount of pericardial effusion, slight splenomegaly, fluid in the pelvic cavity, bilateral hypoechoic lymphadenopathy of the neck, and axilla and enlarged lymph nodes in the groin were also observed. She underwent ultrasound-guided thoracentesis. Pleural fluid was yellow and exudative. Analysis of the pleural fluid revealed the following: specific gravity: 1.020; Rivalta test: positive (++) ; total number of nucleated cells: 4795×10⁶/L (46.8% neutrophils, 53.2% lymphocytes, 54% macrophages, 3% eosinophils, 1% basophils); total protein: 43.6g/L; and lactic dehydrogenase (LDH): 288 u/L. Gram stain, culture, and cytology of the aspirated fluid were negative.

Tracheoscopy showed acute inflammatory changes in the tracheobronchial mucosa, with negative gram stain, culture, and cytology. Medical thoracoscopy revealed multiple nodules and thickening of the parietal pleura (Figure 2a). Pathological results of the pleural biopsy suggested MPM. Immunohistochemistry was positive for calretinin, CK (L), CK 5/6, vimentin and Ki-67 (+10%), weakly positive for WT-1, and negative for napsinA, p63, TTF-1 and desmin. Combined with the morphology, these findings were consistent with MPM (Figure 2b).

The patient received only one cycle of pemetrexed/cisplatin. Prednisone acetate and hydroxychloroquine were administered orally to control SLE.

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Figure 1: Contrast-enhanced computed tomography (CT) of the thorax on initial admission (a, b). ACT scan of her thorax 3 years later (c, d).

Figure 2: (a) Thoracoscopy revealing multiple nodules and thickenings of the parietal pleura; (b) Pathological results of pleural biopsy suggested pleural mesothelioma.

Table 1: Anthropometric measures and laboratory tests during the hospital and clinical evaluations.

Findings	
Neutrophil percentage [50%-70%]	88.1%
Hemoglobin (g/l) [110-150]	82
Erythrocyte sedimentation rate (ESR, mm/1 st hour) [0-20]	68
C-reactive protein (CRP, mg/dl) [0-8]	97.72
Anti-nuclear antibodies (ANA)	1/3200
Anti-double-stranded deoxyribonucleic acid antibodies	+
Anti-Smith antibodies	+
SS-A antibody	+
SS-B antibody	-
snRNP/Sm antibody	+
Anti-mitochondrial M2 antibody	+
Anti-nucleosome antibody	+
Ribosome P protein antibody	+
Anti-ro52 antibody	+

Square brackets indicate the normal levels of each variable.

Approximately three years later, she was admitted with dyspnea, mild orthopnea, and tachycardia. Chest contrast-enhanced CT showed massive pericardial effusion and bilateral pleural effusion, nodules in the lower lobe of the right lung (Figure 1c, d). Echocardiography showed marked pericardial effusion. She opted to discontinue treatment and died one month later.

DISCUSSION

We analysed this case of MPM and SLE in a patient who had not been exposed to asbestos.

The current classification criteria for SLE are those recommended by the American Society of Rheumatology.⁴ SLE can be diagnosed after excluding infection, tumors and other connective tissue diseases. Patients may present with knee joint pain, interphalangeal joint pain, dry mouth, serous cavity effusion, pleural effusion, pericardial effusion, anemia, lymphopenia, high antinuclear antibody titer, anti-dsDNA positivity and Sm antibody positivity.

Symptoms of MPM mainly include chest pain, chest tightness, dyspnea, cough, fever, and shoulder and back pain.⁵ The imaging findings include: diffuse pleural thickening, multiple pleural nodules, and pleural effusion. However, symptoms and imaging findings lack specificity. Pleural biopsies can be used for pathological diagnosis. Our patient had chest pain and dyspnea. Thorax CT indicated pleural effusion, and thoracoscopy indicated multiple nodules in the parietal pleura. The pathological results suggested a mesodermal tumor; and immunohistochemical stains were consistent with MPM.

The patient was diagnosed with SLE with MPM. There are no such reports in China, and there are two cases of pericardial malignant mesothelioma misdiagnosed as SLE.^{6,7} Could the diagnosis of MPM with SLE in this case be established easily? Or was the diagnosis of SLE a mistake? Unlike the previously reported cases, the anti-nuclear antibody titer in our patient was 1:3,200, and both the anti-dsDNA antibody and anti-Sm antibody were positive, making the diagnosis of SLE certain.

MPM is a rare malignant neoplasm of mesothelial lined cavities; and MPM presenting with SLE is even rarer. Its pathogenesis is still unclear. Determining whether there is any association between MPM and SLE requires more cases.

We should pay attention to pleural effusion when diagnosing SLE. If possible, a pleural biopsy should be performed to reduce misdiagnosis and missed diagnosis.

CONFLICT OF INTEREST:

The authors declared no conflict of interest.

PATIENT'S CONSENT:

Written informed consent was obtained from the patient for analysis and publication of personal clinical data and accompanying images.

AUTHORS' CONTRIBUTION:

YL: Mainly responsible for the conception and writing of articles. JT, XL, WC, LF: Mainly responsible for revising the work.

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