Treatment and outcome of a patient with pleural unilocular cystic mesothelial proliferation

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Abstract
Pleural cystic mesothelial proliferation is an extremely uncommon disease arising in the mediastinal pleura. Usually, the cysts are incidentally found and preoperative diagnosis is difficult due to their atypical appearance. To date, only seven cases of pleural mesothelial proliferation have been reported, and only one patient with unilocular has been reported. Here, we report a case of a 55-year-old woman with a mediastinal pleural unilocular cystic mesothelial proliferation that was completely resected by video-assisted thoracoscopic surgery. After 2 years of follow-up, the patient had no evidence of recurrence. In conclusion, our case report may contribute to the understanding of pleural unilocular cystic mesothelial proliferation.

KEYWORDS
chest, mesothelial proliferation, pathology, pleura, VATS

INTRODUCTION
Cystic mesothelial proliferation is a rare disease, generally considered rooted in peripheral mesothelial cells. This disease occurs commonly in the peritoneum of young women and rarely in the pleura.1 To date, only seven cases have been reported in the literature. Here, we report an extremely rare case of pleural unilocular cystic mesothelial proliferation in an adult patient that was successfully resected by video-assisted thoracoscopic surgery (VATS). To our knowledge, this is the second case report about pleural unilocular cystic mesothelial proliferation which has been resected by VATS.

CASE REPORT
A 55-year-old female patient was admitted to our hospital with a 3-week history of shortness of breath and chest tightness, without other accompanying symptoms. No obvious abnormalities were found in physical examination. The laboratory blood tests were normal. She was not exposed to asbestos and had no history of tuberculosis. The computed tomography (CT) scan showed cystic hypodensity shadow in the right superior mediastinum with a diameter of about 5.4 cm. CT value of the cystic hypodensity is about 12 HU, without reinforcement. No other lesions were detected (Figure 1). Endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) was performed, and ultrasound showed a low-density mass with non-vascular structure. After positioning, puncture biopsy and negative-pressure aspiration were performed, and a total of about 20 ml of light yellow clear liquid was extracted. No positive tumour cells were found in cytology (Figure 2A,B).

After sufficient preoperative examination and preparation, the mediastinal mass resection by VATS was performed. Video-assisted thoracoscopy showed a single, unilocular, thin-walled, translucent cyst with a size of 6 × 5.5 × 2 cm located in the right upper mediastinum. The cyst with a complete envelope was completely excised and no fluid spilled (Figure 2C,D). The patient had an uneventful post-operative course and was discharged on post-operative day 7.

Gross findings showed that the inner wall of the cyst was smooth and 0.1 cm thick (Figure 3A). Microscopy examination revealed that the cyst was lined by a single layer of flattened and cuboidal cells. Immunohistochemistry results showed that the cells were positive for AE1/AE3, vimentin, calretinin, D2-40, WT1 and CK5/6. The Ki67 positivity rate was approximately 1% (Figure 3B–H).
FIGURE 1  Radiological findings. The computed tomography showed cystic hypodensity shadow in the right superior mediastinum. (A) Axial view, (B) coronal view and (C) sagittal view.

FIGURE 2  Findings in endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) and video-assisted thoracoscopic surgery. (A) Ultrasound findings in EBUS-TBNA progress. (B) Extracted liquid by EBUS-TBNA. (C) Video-assisted thoracoscopy revealed a single, unilocular, thin-walled, translucent cyst beneath the parietal pleura. (D) The surgically resected specimen was $6 \times 3.5 \times 2$ cm in size.
The pathological diagnosis was cystic mesothelial proliferation. During 2 years of follow-up, the patient was free of symptoms and without any evidence of recurrence on physical and radiologic examination.

**DISCUSSION**

The mesothelial origin of the lesion was first confirmed in 1979 by electron microscopy. Before that, the lesion has been given many names including ‘multicystic peritoneal mesothelioma’, ‘inflammatory cysts of the peritoneum’, ‘post-operative peritoneal cysts’, ‘benign papillary peritoneal cystosis’ and so on. This variation in terminology is attributed to the controversy as to whether the lesion is benign or malignant and to the fact that there is no consistent aetiology. About its biological behaviour, Weiss and Tavassoli suggested that the lesion is a true neoplasm and not a mesothelial reaction. Their hypothesis is based on the lesion progression seen in one case in their series resulting in death and the mixture of cystic and epithelial-like pattern. Other authors suggested that the multicystic mesothelial proliferation (MMP) represented reactive mesothelial proliferative lesions. The inflammatory component, fibrosis, oedema and vascularity of the stroma are usually observed in reactive lesions, and DNA analysis further supports this evaluation as all the cases are euploid. De Rosa et al. investigated the clinicopathological findings of five cases with MMP and revealed that immunohistochemistry showed positive staining for cytokeratin and epithelial membrane antigen, and focal positivity for vimentin and carcinoembryonic antigen, and DNA analysis identified euploid cell populations in all cases. Clinical and pathological data support the hypothesis that MMP represents a reactive mesothelial proliferation and not a neoplastic process. The immunohistochemistry of this patient was consistent with the above results and also revealed that MMP is a reactive mesothelial proliferation, the only difference in this patient is that the cyst was unilocular.

The aetiology of this disease is still not known. Mesothelial cells may be damaged by many inciting factors such as foreign fibres and dusts and inflammatory mediators, resulting in hyperplastic and neoplastic changes. Asbestos exposure is a well-established cause for malignant mesothelioma. However, there is not any evidence of a certain association in benign MMP. Psathakis et al. reported the first reported male with pleural MMP. The case was diagnosed as pleural MMP 1 year after the treatment of tuberculous pleurisy. But it is unknown whether the tuberculous pleurisy had any aetiologic relation with the development of pleural MMP.

The pleural cystic mesothelial proliferation generally has no obvious clinical symptoms and is often found during physical examination. According to seven reported cases, half of them presented with no specific symptoms and were
found with abnormality in the chest imaging examination, yet the rest presented with chest pain or dyspnoea. But there are also patients with serious complications, such as hemothorax. The patient in our study has chest tightness and shortness of breath due to mass compression. The diagnosis of cystic mesothelial proliferation is difficult, and is confirmed mainly according to pathological results. However, it is difficult to obtain a definitive histological diagnosis before surgery. The standard surgical approach for resection has not yet been well established. The progress of VATS enabled us to make the diagnosis and treatment simultaneously.

This case reveals that the symptoms of pleural cystic mesothelial proliferation are untypical, and the diagnosis relies mainly on pathological outcomes. Due to the small number of reported cases, no risk factors affecting recurrence have been found, and it is not clear whether the prognosis of unilocular and multicystic is consistent. Complete resection and regular follow-up are necessary in any case.

**AUTHOR CONTRIBUTION**
Guoliang Zhang drafted the manuscript and Jing Li revised it. Rui Wang provided the clinical data. Xueying Qiao reviewed the literature. All authors read and approved the final manuscript.

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We thank the patient for giving consent to this case report.

**CONFLICT OF INTEREST**
None declared.

**DATA AVAILABILITY STATEMENT**
The data that support the findings of this study are available from the corresponding author upon reasonable request.

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