Benign Metastasizing Leiomyoma of the Lung with Spontaneous Pneumothorax

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Introduction

Benign metastasizing leiomyoma (BML) is a rare benign tumor, occurring commonly in patients with a history of enucleatic or total myomectomy for benign uterine leiomyoma. BML frequently presents with multiple asymptomatic lung lesions, and is usually diagnosed incidentally. A case of BML associated with pneumothorax is reported.

Case Description

A 47-year-old woman was admitted with recurrent pneumothorax. Preoperative chest computed tomography (CT) showed multiple lung nodules and cysts bilaterally. She had undergone enucleatic myomectomy 12 years earlier. Video-associated thoracoscopic biopsy was performed. Histopathologically, there were bulla-like dilated cystic changes, the walls of which showed spindle cell proliferation, causing pneumothorax. Hormone therapy was started after benign metastasizing leiomyoma resection; pneumothorax has not recurred in 7 months. Multiple residual lung nodules have decreased or disappeared on CT.

Abstract

A 47-year-old woman was admitted with recurrent pneumothorax. Preoperative chest computed tomography (CT) showed multiple lung nodules and cysts bilaterally. She had undergone enucleatic myomectomy 12 years earlier. Video-associated thoracoscopic biopsy was performed. Histopathologically, there were bulla-like dilated cystic changes, the walls of which showed spindle cell proliferation, causing pneumothorax. Hormone therapy was started after benign metastasizing leiomyoma resection; pneumothorax has not recurred in 7 months. Multiple residual lung nodules have decreased or disappeared on CT.

Keywords

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**Discussion**

BML was first described by Steiner in 1939.\(^1\) This is a rare tumor, with less than 200 cases documented in the literature, and it occurs in the pelvic cavity, lungs, lymph nodes, and, rarely, in the vertebrae or skull base.\(^2\)

BML is suggested by hematogenous lung metastasis of uterine leiomyoma, and there is a history of uterine fibroids in women of reproductive age. Because many cases of postoperative uterine fibroids are enucleated, operation appears to stimulate metastasis.\(^3\)

The prognosis of most cases of BML is good and depends on the degree of estrogen receptor expression.\(^4\) Some patients die as a result of respiratory insufficiency due to the pulmonary metastases.\(^4\) Tadashi et al reported a case of BML that decreased in size naturally, suggesting that menopause may play a role in its natural decrease in size.\(^5\)

There is no standard treatment, but the treatment option of choice is primary surgical resection or hormone treatment. After initiation of leuprorelin acetate therapy, total lung capacity increased by 10% (500 mL), forced vital capacity increased by 15% (300 mL), forced expiratory volume in the first second increased by 14% (270 mL), and diffusion capacity for carbon monoxide increased by 4% over 11 years of follow-up.\(^5\)
Cystic lesions were observed surrounding the uterine myoma cells in the present patient and, while rare, cases of BML presenting as cystic lesions have been reported, with pneumothorax arising in 1% of all BML cases. One proposed mechanism for the development of cystic lesions is gradual growth of the uterine myoma cells in the respiratory tract, which then expands as a result of breakdown of elastic fibers and air trapping, causing cystic lesions to develop. In the present patient, the mechanism of recurrent pneumothorax suggests that uterine myoma cells in the distal airway became check valves, cystic lesions were formed, and then the cysts ruptured.

Catamenial pneumothorax due to heterotopic endometriosis was initially suspected in the present case; however, pathological examination of the multiple dark red lesions observed intraoperatively in the visceral pleura of the right lung revealed no endometrial tissue. Despite these findings, the repeated occurrence of pneumothorax with menstruation and the elevated CA 125 levels in the pleural effusion fluid prevent coincidence of catamenial pneumothorax due to intrathoracic endometriosis from being completely ruled out. No diagnostic criteria have been established for catamenial pneumothorax. Pores, blueberry spots, and diaphragmatic lesions, such as white, depressed lesions, are observed in 78% of catamenial pneumothorax cases, and pulmonary pleural lesions are observed in 14 to 39% of catamenial pneumothorax cases. The present patient had no diaphragmatic lesions or pelvic endometriosis, and although CA 125 levels in the pleural effusion fluid were inexplicably high, elevated CA 125 levels are also found in benign diseases such as pleurisy, ultimately providing minimal evidence supporting a diagnosis of catamenial pneumothorax.

Regular follow-up is required in the present case due to the possibility of the BML growth.

References