



## Case Report

## Benign metastasizing leiomyoma with lung cystic lesions and pneumothoraces: A case report

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## A B S T R A C T

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Benign metastasizing leiomyoma (BML) affects women of the middle age, and is a rare disease. Typically, in BML, uterine leiomyoma, and lung nodes are found. In the literature, only 3 cases of large cysts have been reported. In our case, we describe a patient with uterine leiomyoma with lung cysts, and several pneumothoraces.

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## 1. Introduction

Benign metastasizing leiomyoma (BML) is a rare disease, with about 100 cases yet reported.<sup>1</sup> It was described for the first time in 1939 by Steiner.<sup>2</sup> Typically, the disease is diagnosed several years after surgery for uterine leiomyoma, in a women aged 30–64 years. Asymptomatic lung nodules ranging from a few millimeters to several centimeters are present. Up to now, only a very few cases of BML with bilateral cystic lesions have been reported.<sup>3</sup>

We describe a new case of a patient with a typical pattern of BML with bilateral cystic lesions, and recurrent pneumothoraces.

## 2. Case

A 55-year-old woman was referred to our outpatient clinic on December 2000 for pulmonary nodes. Mother of one healthy child, non-smoker, exposed to environmental tobacco smoke for 20 years, she had been a shoemaker for 7 years, then a saleswoman, and an administrative employee. Her previous history was marked by an uterine surgery (no more precise information) for uterine fibroma, when 22 years old, followed by a partial hysterectomy 10 years later, and a total one with bilateral oophorectomy, at age 47, for an important uterine myosis with endolymphatic peritoneal

stromal involvement. Partial hysterectomy pathology, at age 32, was characterized by the presence of typical dystrophic endometrium with a suspicion of transformation in a squamous cell carcinoma. Total hysterectomy pathology, at age 47, was characterized by an endolymphatic peritoneal stroma, and a uterine myosis. There were 7 nodules on the uterine piece, with no atypical or neoplastic cells. The endometrial mucosis showed tubular epithelium with several mitosis. There was also a proliferation of smooth muscle cells.

In the mean time, the patient experienced, from age 22 to 47, 6 pneumothoraces (2 right, and 4 left).

Finally, following a systematic chest X-ray done after a contact with a TB patient, a vertebral angioma was diagnosed, at age 52, on a spinal MRI.

The patient was referred to us for a spontaneously resolute stage III NYHA classification dyspnoea, with presence of multiple bilateral, round, well limited, homogeneous lung nodes (4–10 mm in size) associated, on CT scan, to bilateral cystic lesions more importantly on the left side (Fig. 1). Physical examination was normal. Serum  $\alpha$ 1 anti-trypsin level was normal. No finger clubbing was observed.

In view of the presence of bilateral lung nodes, a search for a primary carcinoma was carried out. Mammography, FDG-PET, and thyroid scintigraphy excluded gynaecological or thyroid neoplasia. Interestingly, no hypermetabolism was observed by FDG-PET at thoracic level. Lung function testing was as follows (% predicted): FVC: 1.94 l (68%), FEV<sub>1</sub>: 1.43 l (59%), and TLC: 3.77 l (78%), with following blood gases: PaO<sub>2</sub>: 74 mmHg, PCO<sub>2</sub>: 38 mmHg, and pH: 7.4.

In view of the presence of bilateral cystic lesions, and of the history of pneumothoraces, no transthoracic biopsy was carried out.

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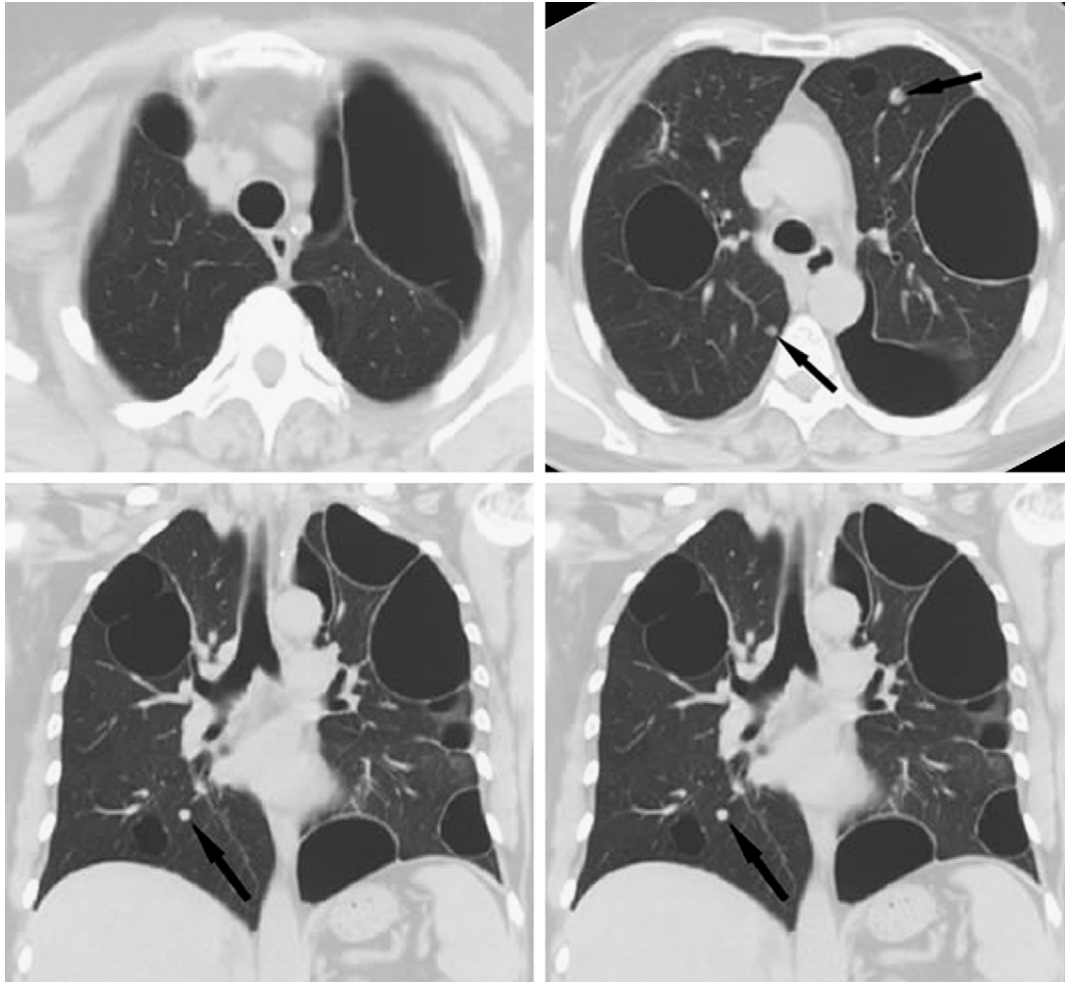


Fig. 1. CT scan with cystic lesions, and lung nodules, at age 60.

No sign of tuberous sclerosis was observed, and no familial history of this disease recorded.

Thus, if the diagnosis of BML was very likely, in view of age at disease onset, history of previous hysterectomy for uterine leiomyoma, typical uterus pathology, and presence of typical lung nodules, two clinical features were less typical: bilateral lung cystic lesions, and pneumothoraces.

Since diagnosis, a carefully follow-up has been carried out every year, with clinical examination, CT scan, and lung function testing. A total stability of the disease has been observed.

### 3. Discussion

BML is a rare disease associated with typical uterine leiomyoma.<sup>1</sup> Common sites of metastasis include pelvic lymph nodes, and lung.<sup>1</sup> This disease is characterized by a much less aggressive course than leiomyosarcoma, due to the benign nature of proliferating smooth muscle cells in leiomyoma.<sup>1</sup>

BML appears in premenopausal women with a history of uterine leiomyoma, and the metastases can appear from 3 months to 20 years after uterine surgery. This disease is usually asymptomatic, but sometimes, cough, chest pain, and dyspnoea have been observed. Typical radiologic findings show well-circumscribed bilateral non calcified pulmonary nodules ranging from a few millimeters to several centimeters. Large cystic lesions with multiple nodules have been reported in only 3 cases.<sup>3</sup> And, only one case of pneumothorax has

been, to our knowledge, reported in the literature; our case is the second. The cause of the pneumothoraces is unclear, but the rupture of cystic lesions may, at least partially, explain it.<sup>4</sup>

Leiomyoma histology is characterized by the presence of smooth muscle cell growth with oestrogen, and progesterone receptors present in approximately 70% cells.<sup>1</sup> In our observation, no histological material was available, and one of the authors (JMV), who is a lung pathologist, could not examine the material before for confirmation of the diagnosis.

The classical treatment of BML consists in pulmonary surgery, hormonal therapy (Tamoxifen, progesterone, aromatase inhibitors, luteinising hormone-releasing hormone analogue), or bilateral oophorectomy.<sup>5</sup>

The outcome is usually good in response to hormonal therapy, due to the presence of oestrogen, and progesterone receptors.<sup>1</sup> Since our patient was asymptomatic, did previously underwent bilateral oophorectomy, and her BML clinical features were stable, no specific treatment was prescribed.

In this observation, two clinical characteristics, lung cystic lesions, and pneumothoraces are not BML features, but both are observed in an another disease characterized by smooth muscle cell proliferation, namely lymphangioliomyomatosis (LAM).<sup>6</sup> Typical LAM patients are women of reproductive age, with a history of recurrent pneumothoraces associated with bilateral interstitial pattern that may be reticular, reticulo-nodular or miliary, and cysts.<sup>7</sup> LAM treatment is, basically, the same as BML.<sup>8</sup> Finally, two

distinct types of LAM have been reported: the sporadic type, and one associated with tuberous sclerosis due to a genetic mutation of genes TSC1 or TSC2.<sup>9</sup>

#### 4. Conclusion

This observation is the second case of BML diagnosed in a patient presenting lung cystic lesions, and pneumothoraces, both of these characteristics being more typical of LAM.

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