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Benign Pulmonary Metastasizing Leiomyoma

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Abstract

Benign metastasizing leiomyoma (BML) is a rare event that occurs in middle aged women with a history of uterine myomas. It is characterized by multiple leiomyomatous lesions in many tissues and organs, especially in the lungs. We report the case of 56 years old woman presenting chronic cough, hemoptysis and having a history of myomectomy for a uterine leiomyoma 6 years ago. Chest CT showed a right pulmonary process. The histological diagnosis was defined by bronchoscopy and biopsies. The surgery was not an option because of the bronchovascular extension of the tumor, so hormonal therapy was administrated to the patient. One year later, Chest CT showed that the tumor doesn't increase.

Keywords: Benign Metastasizing Leiomyoma, BML, Lung, Hormonal Treatment

Introduction

Benign metastasizing leiomyoma (BML) is a rare condition first described by Steiner in 1939 despite the universal belief of the time that benign neoplastic tumors do not metastasize [1]. Lungs are the most common location of uterine myoma metastasis, but other locations have also been described in this context, including the mediastinum, heart, trachea, esophagus, skin, skeletal muscles, deep soft tissue, breasts, liver, urinary bladder, retroperitoneal space, nervous system, and bones [1,2]. The majority of tumors are asymptomatic, they are therefore identified incidentally on routine chest X rays, however, certain of these tumors induce coughing, hemoptysis, dyspnea and decreased pulmonary function [2]. Due to the low morbidity and scarcity of reports on this condition, there is no consensus on which methods should be used to treat this disease.

Case Report

She is a 56 years old female patient, operated 6 years ago for uterine myoma, having since 10 months a dry cough. Physical examination is normal. Chest radiography objectified straight parahilar opacity, heterogeneous with fuzzy limits. The chest CT showed a right upper lobe tissue process measuring 40x32x16mm with right hilar lymphadenopathy (Figure 1).

To have a histological diagnosis, flexible bronchoscopy was performed objectifying stenosis of the ventral segmental bronchus of the right upper lobe.

The biopsies at this level showed spindle cells arranged in more or less elongate and beams with foci formation of palisade appearance structures. Immunohistochemical study showed positivity of tumor cells to anti AML and anti desmin and negative for hormone receptors, anti PS100, anti CD163 with a Ki 67 negative. This allowed us to retain the diagnosis of primary lung leiomyoma.

The surgery was not executed because the penalty is pneumonectomy for benign tumor. A monitoring was recommended but the patient lost sight of. She reconsulted a year after seeing the appearance of thoracic pain with hemoptysis evolving into a state of conservation of the general state.

Chest CT showed an increase in the volume of the mass (84x81x71mm) with compression of the bronchovascular structures without lesion at the abdominal level (Figure 2).

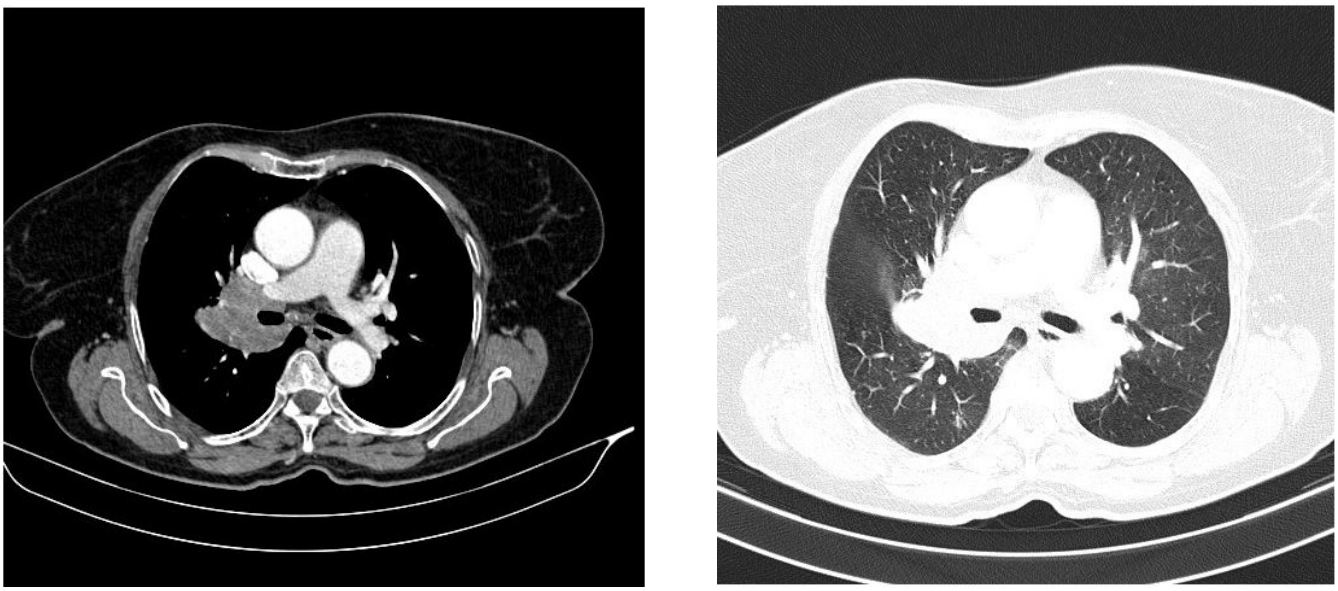

Figure 1: Chest CT shows a right upper lobe tissue process (**a**) mediastinal window and (**b**) lung window

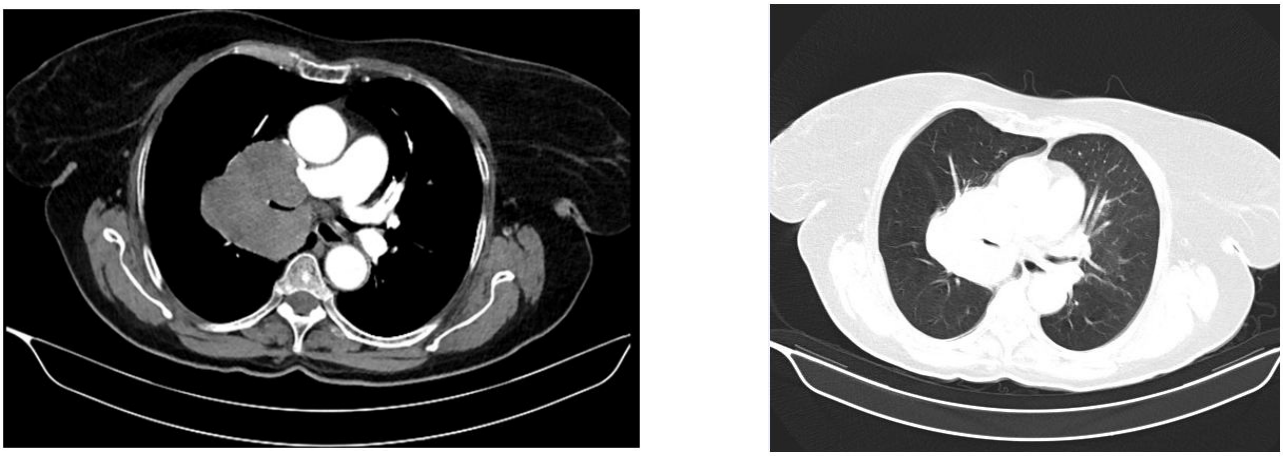

Figure 2: Chest CT done a year later showing an increase in the volume of the mass with compression of the bronchovascular structures (**a**) mediastinal window and (**b**) lung window

Given the evolution of the lesion, we doubted the initial diagnosis. Bronchial fibroscopy was redone with biopsy revealing the presence of smooth muscle cells expressing desmin, estrogen receptors and progesterone with a Ki 67 at 5% which was in favor of a benign metastasizing leiomyoma.

The file has been re-discussed: surgery is no longer possible. Hormonal treatment was retained. The patient was put on an aromatase inhibitor (letrozole: 1 tablet a day). Currently, she is one year of treatment with a clear clinical improvement: disappearance of hemoptysis and chest pain with persistent intermittent cough. On a CT scan, the tumor remained stable.

Discussion

BML is a rare condition which is a similar histological appearance with benign smooth muscle tumours but present at distant sites. Its most often diagnosed in women between the ages of 34 and 55 years; the mean age of diagnosis is approx. 47 years and it usually occurs years later after uterine leiomyoma surgery [1]. This condition is characterized by multiple leiomyomatous lesions in many tissues, organs and lungs are the most common site of the metastases. Benign metastasizing leiomyoma is rarely diagnosed and there have been over 150 cases reported in the literatüre [3].

The disease may be asymptomatic and diagnosed by chance during an imaging examination. It may also cause symptoms such as cough, dyspnea, and pain [1].

Radiographic findings most often reveal solitary or multiple well-circumscribed nodules, ranging from a few millimeters to several centimeters. Rare cases have been reported with a miliary pattern, cavitary lung nodules, interstitial lung disease and multiloculated fluid containing cystic lesions. For further examination a thorax CT can be performed to determine the exact

size and location of the lesions before the biopsy [2,4]. Previously, it's was known that there is a lack of 18 FDG uptake in PBML. However, a literature review concerning patients with PBML and 18 FDG PET/CT findings was conducted [5]. In total, 36 cases were identified in 30 reports. With regard to the 18 FDG PET/CT findings, the accumulation of 18 FDG varied significantly: A total of 25 patients (69.4%) were assigned to the no uptake group, 8 patients (22.2%) were assigned to the low uptake group and 3 patients (8.3%) were assigned to the high uptake group. However, only one patient out of those 3 had a tumor that demonstrated aggressive behavior [5].

It is indispensable to identify PBML from other pulmonary diseases such as benign or malignant tumor (low grade leiomyosarcoma), pulmonary lymphangioleiomyomatosis, tuberculoma, sarcoidosis, pneumoconiosis, lung collagen vascular disease, inflammatory pseudotumor, metastatic carcinoma of the lung and so on. That's why lung biopsy is indispensable and the standard diagnosis for PBML [6].

Pathological features of pulmonary BML are often of a benign nature, as observed in the present case. Absence of high cellularity coagulative tumor cell necrosis, cytological atypia and increased mitosis (>5 per 10 high powered fields) with a low Ki 67 index support the low proliferative state and benign nature of these tumors. Interlacing fascicles of smooth muscle cells lacking anaplasia or vascular inva¬sion, with the entrapped respiratory epithelium are revealed upon histological examination. A range of immunohistochemical markers, including desmin and muscle specific actin, are present to confirm the mesenchymal derivation of these tumors with smooth muscle differentiation. In addition, the presence of estrogen and progesterone receptors supports the derivation of BML from the uterus, which reinforces the use of treatment with hormonal agents [4].

Detailed cytogenetic and molecular studies on BML have demonstrated that telomere length is not a decisive factor in the development of the disease [1]. The studies also revealed specific anomalies regarding both the number of chromosomes (trisomy 12) and their structure (translocation, aberration 19q, deletion 22q) [1].

Currently, BMLs are considered to develop as a result of the monoclonal spread of uterine myomas through the circulatory system. Detecting the miR-221 marker may be helpful in differentiating myomas from sarcomas [1].

The pathogenesis of BML has yet to be defined. The presence of hormone receptors and susceptibility to antihormonal therapy suggests Müllerian origin [2,6]. Hypotheses include hormone-sensitive in-situ proliferation of smooth muscle bundles, hematogenous spread of benign smooth muscle cells from uterine leiomyoma, or surgically induced hematogenous spread during hysterectomy [2]. Surgical seeding intraoperatively seems less likely, based on cases in which BML was identified on preoperative imaging prior to hysterectomy. Some investigators have suggested that the lesions represent a low-grade, slow-growing and multifocal leiomyosarcoma [2,4]. However, histologic examination consistently fails to show features of malignancy, including an increased mitotic rate, necrosis and cytologic atypia [2].

Treatment methods for BML include surgical resection, hysterectomy and bilateral oophorectomy, administration of progestin's and aromatase inhibitors, and medical castration using luteinizing hormone releasing hormone analogs. Lesions that increase in size may require surgical resection to prevent potentially fatal complications such as massive hemoptysis. However, the effect of reducing the tumor burden through surgical palliation should be carefully evaluated. Smaller sub centimeter lesions can be followed by surveillance scans. Hormonal relative treatment is a commonly chosen therapy for BML when estrogen and progesterone receptors are identified on tumor histology. However, hormone therapy does not generate a response in all patients, and the side effects of flushes, fatigue and nausea can be aggravating to the patient. A combination of medical and surgical treatment would exert a synergistic effect and should be considered in the management of progressive and symptomatic lesions [1,3,4].

Conclusion

Although rare, BPML should be considered for reproductive-age women who have undergone hysterectomy for uterine leiomyoma and subsequently present with multiple, pulmonary nodules or cysts. Confirming smooth muscle origin and estrogen and progesterone receptor positivity via immunohistochemistry remains paramount to establishing the diagnosis and the cytogenetic studies seem to give us the most helpful findings in the differential diagnosis.

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