Benign Metastasizing Leiomyomatosis of the Lungs Presenting a Miliary Pattern

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Benign metastasizing leiomyomatosis is a very rare and significantly interesting pathology of the lungs. It is a challenge to clinicians when presenting a miliary pattern in preoperative radiologic imaging because it could be any other interstitial disease or infectious in etiology such as miliary tuberculosis. We report a case of innumerable tiny nodular densities spread evenly throughout both lungs in a patient with history of hysterectomy for a fibroid uterus.


Pulmonary benign metastasizing leiomyomatosis (BML) is a confusing clinicopathologic definition. A few hundred cases have been reported in the medical literature worldwide. They are solitary or multiple lung nodules metastasizing from a fibroid uterus. We recently encountered a miliary pattern of presentation of this disease. To our knowledge, this observation has not been reported since Lipton and colleagues [1] first described it in 1987.

The patient is a 41-year-old white woman with a long-standing history of infertility and seven attempts at in vitro fertilization. She eventually had one full-term pregnancy and a Caesarian section in 2006. She had lumpectomy for a ductal carcinoma in situ of the left breast and chest radiotherapy in 2009, followed by supracervical hysterectomy without salpingo-oophorectomy for a fibroid uterus a year later. She is negative for BRCA 1 and 2 gene mutation. She had been taking tamoxifen.

The patient was asymptomatic. Results of a preoperative laboratory workup, which included complete blood count, electrolytes, and hepatic and pulmonary function tests, were normal. A routine chest roentgenogram showed a tiny nodular density in the right upper lobe, but a chest computed tomography scan with contrast revealed innumerable tiny densities spread almost evenly in both lungs (Fig 1) suggesting a miliary pattern. A Mantoux test result was negative.

A right video-assisted thoracoscopy showed numerous reddish 2-mm to 3-mm nodules on the lung parenchyma scattered mainly along the margins of the middle lobe and upper margin of the lower lobe with nonspecific pattern. The visceral pleura was smooth and did not have signs of puckering (Fig 2). There was no effusion, and the mediastinum showed no lymphadenopathy.

The multiple wedge specimens showed BML exhibiting a miliary spread pattern. Multiple nodules scattered in the lung parenchyma associated with bronchioles consisted of a monomorphic population of spindle cells without atypia or appreciable mitotic activity (Fig 3). Immunostains demonstrated spindle cells positive for estrogen receptor, progesterone receptor, desmin, and smooth-muscle actin. Ki-67 showed low proliferation. No acid-fast bacilli were seen on a direct fluorochrome-stained smear.

A month after the lung operation, she underwent robotic-assisted bilateral salpingo-oophorectomy and fulguration of pelvic endometriosis. In a short-term follow-up, she has been symptom-free.

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Comment
In 1983 Martin [2] proposed a classification for smooth-muscle tumors in the lung. This addressed the confusion in diagnosing these tumors that histologically appear benign but that metastasize and prove to be clinically malignant, called “metastasizing fibroleiomyoma of the uterus” by Steiner [3] in 1939.

These lung tumors are categorized as (1) leiomyomatosis, which includes benign metastasizing leiomyoma, lymphangioleiomyomatosis (lymphangioleiomyomatosis), leiomyomatosis peritonealis disseminata, and intravenous leiomyomatosis, (2) metastasizing leiomyoma in men and children, and (3) multiple pulmonary fibroleiomyomatous hamartoma.

The case of BML we report presented histologically like a pulmonary fibroleiomyomatous hamartoma, but after correlating this finding with a history of uterine leiomyoma and hysterectomy, the treatment and prognosis became less benign. Because BML has been associated with hormonal imbalance, bilateral oophorectomy and hormonal therapy were indicated, which have been reported to regress the pulmonary tumors [4]. Regression of BML has also been observed with progestin withdrawal [5], treatment with the luteinizing hormone-releasing hormone analog goserelin [6], and pregnancy or postpartum [7].

In approximately 100 BML cases reported in the literature worldwide, these tumors presented as single or multiple pulmonary nodules and masses, sometimes fluid-filled cysts, in one or both lungs. Lipton and colleagues [1] reported a case of miliary interstitial nodular pattern presentation in both lungs. A similar pattern of presentation of BML is also reported in this case.

The pathogenesis of BML is unclear, although it has been postulated that antegrade metastasis through venous channels can occur to extraterine organs, such as the heart and the lungs, during surgical manipulation of a uterine leiomyoma [8]. Although the patient we report with miliary presentation is posthysterectomy, cases of BML, including the patient reported by Lipton and colleagues [1], have been discovered even before resection of a uterine leiomyoma [4].

In summary this unusual miliary presentation provides interesting diagnostic challenge, because the chest roentgenogram or computed tomography scan could also suggest an infectious, interstitial disease such as miliary tuberculosis or sarcoidosis, which would require a different course of treatment. Although we have not demonstrated regression of the pulmonary lesions in this report, repeat chest computed tomography in a long-term follow up is certainly warranted.

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References