

A Diffuse Pulmonary Presentation of Benign Metastasizing Leiomyomas

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Abstract

Benign metastasizing leiomyomas of the lung remain an infrequently encountered pulmonary pathology with poorly understood pathogenesis and unclear treatment algorithm. They may manifest a spectrum of clinical symptoms and have multiple proposed etiologies. Critical to this diagnosis is adequate tissue biopsy, which additionally may facilitate treatment based on hormonal receptor status. Complete surgical resection of solitary pulmonary lesions is advocated with targeted endocrine therapy as a proposed means of treating disseminated disease.

Long term follow up of these patients is critical to our understanding of the risk of recurrence, malignant transformation and natural history of this disease.

Case Report

This patient is a forty one year old lady who presented with the primary complaint of shortness of breath worse with physical activity. She additionally reported pleuritic chest pain and occasional orthopnea. Her past medical history was pertinent only for menorrhagia associated with uterine leiomyomas. These symptoms prompted a total abdominal hysterectomy at thirty-one years of age. The surgical pathology at that time was consistent with a uterine leiomyoma. The patient's shortness of breath gradually worsened over a twelve-month period prompting her presentation. She had no exposure to, or history of cigarette smoking.

Initial evaluation demonstrated stable hemodynamics with no evidence of jugular venous distension, lower extremity edema or abnormal heart sounds. Auscultation of the chest revealed clear lung fields with no decreased breath sounds, crackles or ronchi. Her chest x-ray demonstrated a peri-hilar Right Lower Lobe (RLL) mass with numerous scattered pulmonary nodules (Figure 1). The cardiac silhouette appeared normal, no effusions or pneumothoraces were present. These findings prompted further evaluation with a Computed Tomography (CT) scan of the thorax. This demonstrated a 4.2 cm × 3.2cm dominant mass in the RLL with diffuse scattered pulmonary nodules varying in sizes up to 1.1 cm (Figure 2). Echocardiography revealed a preserved left ventricular ejection fraction and no valvular or myocardial abnormalities.

The patient was admitted to the hospital for further evaluation of her findings. Bronchoscopy with associated endobronchial washings and transbronchial needle biopsy was non-diagnostic. An open lung biopsy was performed to evaluate the RLL lesion.

A 4 cm mini-posterolateral thoracotomy incision was used to



Figure 1: Chest X-ray demonstrating a right peri-hilar lesion.



Figure 2: CT imaging demonstrating scattered pulmonary nodules measuring up to 1.1cm along with a dominant right lower lobe mass measuring 4.2cm x3.2cm.

gain access to the right hemi-thorax. More than 15 palpable nodules measuring 1- 2 cm were present in the right lung. Wedge biopsy of a 2 cm right lower lobe lesion was performed with a rim of surrounding lung parenchyma. The lesion was well defined, pearly white, firm and appeared to have the propensity to be enucleated from the surrounding parenchyma. A chest tube was placed and the right hemi-thorax was closed.

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Histologic examination of the lesion demonstrated a well-circumscribed spindle cell nodule. There were interfasciculating bundles of spindle cells that were cytologically bland and without evidence of mitoses. Immunohistochemical stains confirmed the biopsy specimen to be a benign leiomyoma with a Ki-67 proliferation index of less than 5%, and estrogen and progesterone receptor positivity. The lesion was also diffusely immunoreactive with smooth muscle antigen, desmin and CD-56 (Figure 3-5).

The patient was discharged home on postoperative day four after removal of her thoracostomy tube. She opted for a trial of anti-hormonal therapy with Tamoxifen. This was started in consultation with a medical oncologist. Her repeat imaging will be obtained after completion of 3 months of therapy. She has been followed for a total of four months and has had no worsening of her symptoms.

Discussion

First described by Steiner in 1938, benign metastasizing leiomyomas remain a relatively rare and poorly understood pathologic process occurring mostly in women of reproductive age [1]. Multiple cases have been described with variations in presentations following myomectomies or hysterectomies ranging from several months to decades. Its incidence is unknown and the initiating agent prompting such a remarkable evolution in the natural progression of this disease is unclear.

Grossly these lesions are pearly white, benign appearing masses

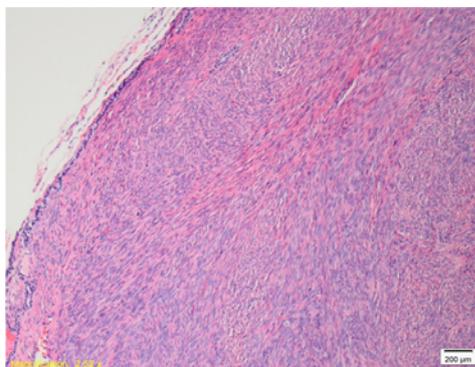


Figure 3: Hematoxylin and eosin stained slide demonstrating the well-circumscribed spindle cell nodule.

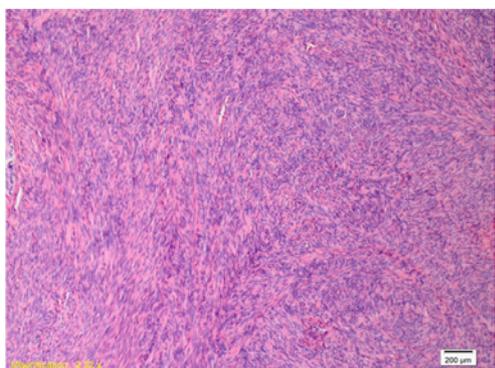


Figure 4: Interfasciculating bundles of spindle cells with moderately high cellularity; however these cells are cytologically bland and no mitoses are evident.

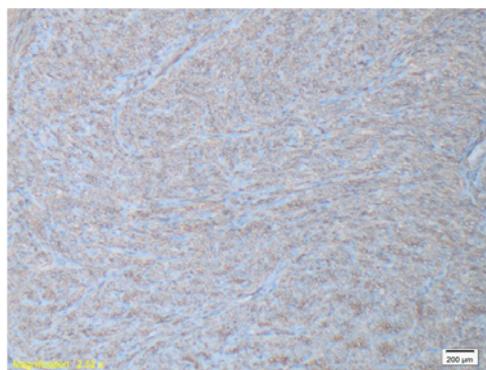


Figure 5: Smooth muscle stain is diffusely positive.

located outside of the uterus. Their proximity to the uterus is ubiquitous and they may occur in the adjacent regions or may be found in anatomically remote sites such as the thorax. They are highly differentiated histologically and have whorled trabeculations with mitotic inactivity or isolated mitoses (usually less than 5 mitoses per high powered field) [2]. In addition they characteristically do not have nuclear pleomorphism and necrosis is usually absent. These features along with the absence of pseudocyst formation differentiate them histologically from their malignant counterparts – the leiomyosarcoma [1-4]. There is no reported spontaneous transformation to leiomyosarcomas.

Multiple theories exist to explain the pathogenesis of benign metastasizing leiomyomas. These include intraperitoneal seeding at the time of uterine myomectomy or hysterectomy, lymphatic or hematogenous spread, hormonal stimulus and coelomic metaplasia as a result of transformation of mesothelial mesenchymal cells under hormonal stimulus [2-6]. Regardless of the means by which these tumors arise, their behavior in many ways mimics the behavior of endometriosis. It is probably more likely that a combination of the proposed theories govern their development and proliferation [2].

In the case we presented, hematogenous or lymphatic spread would account for the diffuse, bilateral manifestation of these lesions.

Clinically, patients are often asymptomatic or have incidentally found lesions on imaging studies or at the time of surgery for an unrelated disease process. However, non-specific pain, related to their anatomic location may occur; and rarely with increased tumor burden, are pulmonary symptoms such as chest pain and dyspnea on exertion present.

Radiographically, x-rays, ultrasonography, computed tomography scans (CT), magnetic resonance imaging (MRI) and F-fluorodeoxyglucose Positron Emission Tomography (F-FDG PET) will demonstrate these lesions. On contrasted CT scans, enhancement similar to the uterine myometrium will occur. T2 weighted MRI images demonstrate isointense to hypointense appearance with enhancement when compared to myometrium [6]. In contrast to leiomyosarcomas these lesions have isometabolic activity on F-FDG PET compared to hypermetabolic activity. Often times due to their sub-centimeter size these lesions may be undetected on routine imaging [6].

Comparison of the uterine and extra-uterine lesions often will demonstrate similar if not identical features. Of importance is the need to review the uterine pathology to exclude features of a leiomyosarcoma.

Once the diagnosis is established, resection of large solitary pulmonary masses may improve symptomatology and provide cure [2,3,6]. Recurrence following excision of a single lesion has not yet been described in the literature. In the case of diffuse lesions as we presented, accurate histopathologic identification and the determination of estrogen or progesterone receptors is crucial. Anti-hormonal therapy to estrogen and progesterone may lead to tumor regression or end further proliferation thereby stabilizing limiting symptoms or providing cure for this indolent disease [2,7].

While proliferation has been noted during estrogen and progesterone therapy, and regression has been noted after pregnancy and menopause, it is not clear whether bilateral salpingo-oophorectomy would be worthwhile to pursue especially since the majority of these cases have been documented in women of child bearing age [2,5]. There has been a reported case of recurrence after such therapy [7]. Additional proposed therapies include embolization and chemotherapy [4,7]. Long term follow up is imperative to improve our understanding of this pathologic process.

Reference

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