

CLINICAL VIGNETTE

Benign Metastasizing Leiomyoma Presenting as Pulmonary Masses

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Case Presentation

A 64-year-old female with hypertension and asthma was referred to the emergency department for elevated blood pressures noted by her dentist prior to routine extraction. She also reported several months of chronic cough and dyspnea on exertion which had been treated with antibiotics and steroids without improvement. She denied fever, chills, night sweats, weight loss, or chest pain. She had no history of smoking.

Exam was notable for mild tachypnea and trace expiratory wheezing bilaterally. Chest x-ray noted “Large rounded opacities within the right middle and lower lungs” and “moderate right pneumothorax” (Figure 1). The masses measured up to 4.5 cm at the right hilum and were not present on prior 2021 CXR. CT Chest showed “Multiple round soft tissue masses in the bilateral lungs, with the largest in the right upper lung measuring 4.0 x 4.0 cm.” Findings were new compared to prior 2021 chest CT and concerning for malignancy, as well as a new “Large right pneumothorax” (Figure 2).

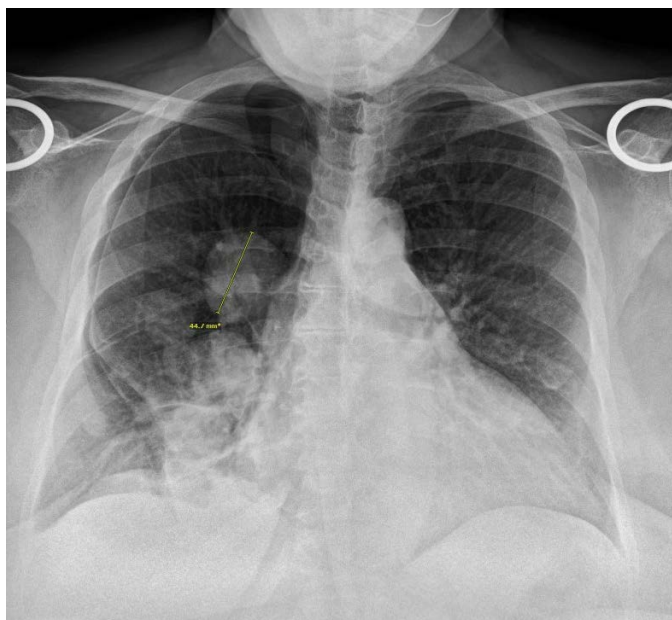


Figure 1 – Chest X-ray

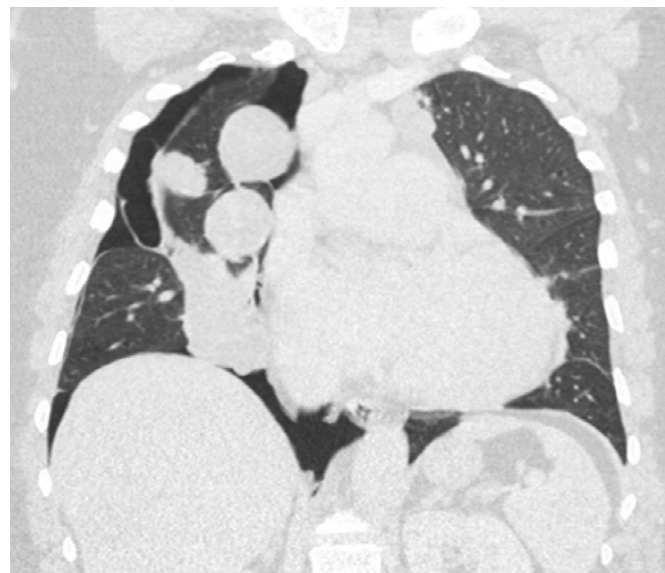


Figure 2 – CT Chest

She underwent chest tube placement and chemical pleurodesis for the pneumothorax. CT guided biopsy of the lung masses revealed “Low-grade smooth muscle cell neoplasm, consistent with benign metastasizing leiomyoma”. On further history, she reported history of uterine fibroids with total abdominal hysterectomy and bilateral salpingo-oophorectomy two years prior. She was also started on anastrozole for benign metastasizing leiomyoma per Gynecologic Oncology with close outpatient follow-up.

Discussion

Uterine leiomyomas (also known as fibroids or myomas) are non-cancerous, monoclonal smooth muscle tumors commonly found in reproductive age females.¹ Benign metastasizing leiomyoma (BML) is a rare condition, first described in 1939,² with tumors present at extra-uterine locations.³ BML may spread to the lungs, and other metastatic sites have been reported including the abdominal/pelvic cavity, skeletal muscle, mediastinum, bone, breast, and heart.⁴

While the pathogenesis of BML is not certain, these extra-uterine tumors are strongly positive for estrogen and progesterone receptors, supporting their derivation from the female genital tract. They have X-chromosome inactivation patterns identical to uterine leiomyomas, support clonal

relationship to uterine leiomyomas. This suggests that metastasizing leiomyomas result from uterine leiomyomas.⁵

The majority of BML patients had uterine surgeries prior to their diagnosis. One case series included 65 women (under 45 years). Nearly 80% had undergone prior gynecologic surgery for uterine leiomyomas including myomectomy and hysterectomy. The mean time from prior gynecologic surgery to diagnosis of BML was 6.3 years with a range 0.08-14 years.⁴

BML is an indolent, slow growing disease and many are asymptomatic at the time of diagnosis. A study of 65 young women (under 45 years) reported 54.8% were symptomatic at the time of diagnosis. Another study of 10 older women included postmenopausal with mean age of diagnosis of 54.1 years. They found only 30% presented with symptoms. The majority in both studies presented with multiple pulmonary lesions. The study of young women reported 60.9% had bilateral pulmonary lesions, whereas the study of older women found 30% of patients only had bilateral nodules. These findings may be related to hormone dependence of these tumors with lower levels of estrogen and progesterone in older, perimenopausal and post-menopausal women.^{4,6}

While tumors can be identified on x-ray and CT imaging of the chest, diagnosis typically requires tissue sampling to rule out other metastatic diseases, such as leiomyosarcomas, and pulmonary lymphangioliomyomatosis (LAM).

Management of BML ranges from observation to medical or surgical intervention. Patients who require treatment, typically have the goal to reduce hormone levels with medications. These include selective estrogen receptor modulators (SERM) such as raloxifene, aromatase inhibitors (AI) such as anastrozole, gonadotropin hormone-releasing hormone (GnRH) agonists such as leuprolide, and progestins. Other treatments including sirolimus and VEGF inhibitors have also been reported. Surgical interventions include excision of lung lesions and/or surgical castration.^{4,7,8}

Patient Follow-up

Due to insurance issues, our patient was unable to follow-up or continue her anastrozole. She presented to the emergency room six months later with acute hypoxic and hypercarbic respiratory failure requiring intubation. Imaging noted increased size of multiple pulmonary nodules and a large right pleural effusion. She was restarted on her anastrozole with close outpatient follow-up.

Summary

Our 64-year-old, post-menopausal female with prior total hysterectomy for uterine fibroids, presented with months of chronic cough. She was found to have bilateral pulmonary masses and subsequently diagnosed with benign metastasizing leiomyoma. Benign metastasizing leiomyoma is a rare condition where smooth muscle tumors are found outside of the

uterus, most commonly in the lungs. While they can be asymptomatic, our patient had significant symptoms and complications including a large pneumothorax on initial presentation. She later developed respiratory failure requiring intubation and received pleural effusions. While these tumors are hormone sensitive, they have reported in post-menopausal patients. It is possible that estrogen production in adipose tissue could have been a factor in our obese patient. Main therapy is anti-hormonal treatment, with variable response. She was started on anastrozole with close follow-up.

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