JPSIM Journal of Pakistan Society of Internal Medicine

Case Report

Benign Metastasizing Leiomyoma (BML) - A Rare Phenomenon with Even Rarer Radiology and Disease Course

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How to cite this:

Usman M, Bakhtawar N, Rajni Sudhir R. Benign Metastasizing Leiomyoma (BML) - A Rare Phenomenon with even Rarer Radiology and Disease Course. J Pak Soc Intern Med. 2022;3(2): 157-159

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Introduction

Benign metastasizing leiomyoma (BML) is an extremely rare condition that occurs in women of all ages. However, it most commonly occurs in women aged 30-75, and particularly among women of late childbearing age.[1] First reported by Steiner in 1939, BML has been reported in the literature only a handful of times.[2] BML occurs in women with a history of uterine leiomyoma (fibroids) followed by hysterectomy or myomectomy. It usually presents within 15 years of the initial diagnosis of uterine leiomyoma.[3] Distant metastasis is common with lungs and lymph nodes being the most common sites of spread. Other sites of metastasis reported in the literature include the central nervous system, heart, skin, esophagus, skeletal muscles, bones, breasts, and trachea.[1] Once diagnosed, BML follows a steady disease course with a mean prognosis of 96 months.[4]

We present an extremely rare case of BML with atypical organ involvement that has not been previously reported in the literature. In addition, our case was unique because of its unusual radiological appearance and rapid disease course, which has also been only scarcely reported in the literature.

Case description

A 60-years old lady presented to the emergency department of a tertiary care hospital with the chief complaints of shortness of breath. Initial chest x-ray showed bilateral lung shadows and bilateral pleural effusions (left more than the right). After a basic blood workup, a decision was made to put a chest drain in the left pleura to relieve the patient's symptoms. Blood-tinged pleural fluid was obtained that was sent for analysis (including cytology, biochemistry, and bacterial culture and sensitivity).

The only significant past medical history of the patient included Per Vaginum (PV) bleeding during her early 40s. She had a transvaginal ultrasound at that time which showed multiple uterine fibroids. Due to excessive bleeding, a decision was made to do a total hysterectomy. Post-surgical tissue analysis of the uterine masses confirmed a diagnosis of fibroids (leiomyoma).

During her stay at the hospital, a CT scan (Computed Tomography) for her chest, abdomen, and pelvis was arranged. The CT scan showed well-demarcated innumerable bilateral lung nodules, bilateral pleural effusions, and hilar and mediastinal lymphadenopathy (Figure 1). The initial set of differential diagnoses was metastatic primary lung cancer, metastatic cancer with unknown primary, and benign metastasizing leiomyoma (BML) considering her previous history of uterine leiomyoma.

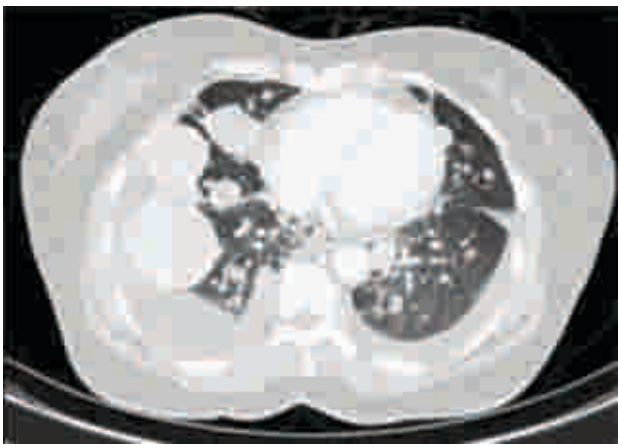

Figure 1: *Cross-Sectional Image of CT Chest Showing Innumerable Well-Demarcated Lung Masses with Bilateral Pleural Effusion, and Lymphadenopathy*

The patient was referred for a CT-guided biopsy of one of the lung nodules and an urgent referral to oncology

was made. The CT-guided biopsy and tissue analysis showed spindle-shaped cells with limited atypia and mitosis, consistent with Benign Metastasizing Leiomyoma (BML). The patient's case was discussed in lung cancer MDT and a decision was made to a whole-body positron emission tomography (PET) scan.

The whole-body PET scan showed increased fluorodeoxyglucose (FDG) uptake in the innumerable bilateral lung masses, and hilar and mediastinal lymphadenopathy. In addition, it showed increased FDG uptake in both the lung pleura (Figure 2).

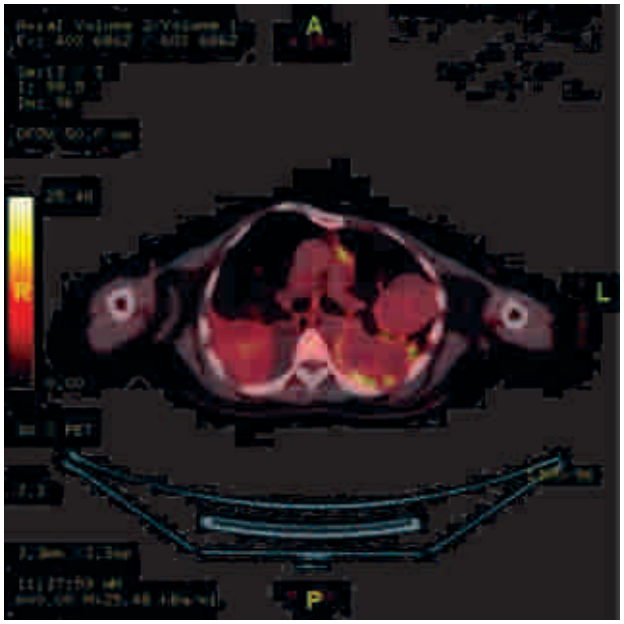

Figure 2: *NM PET Scan Validating the Findings of the CT Scan and Additionally showing Increased Uptake in the Pleura Indicating Pleural Involvement as Well*

During her appointment with an oncologist, based on the biopsy and imaging results, a confirmed diagnosis of Benign Metastasizing Leiomyoma (BML) was made. The patient was started on high-dose anastrozole. However, due to the excessive disease burden, the patient's condition continued to deteriorate rapidly and she passed away within 4 months of her initial presentation.

Discussion

BML has been reported in the literature before but our case is unique due to the atypical organ involvement and disease course.

In our case, the initial diagnosis of uterine leiomyoma was made in the early 40s, which is consistent with the literature where the initial diagnosis was made at 38.5 years. Our lady was in her late 60s and the mean age for diagnosis for BML in literature is 54.1 years.[5]

BML most commonly spreads to the lungs but it is not uncommon for it to involve other organs such as the central nervous system, heart, skin, esophagus, skeletal muscles, bones, breasts, and trachea.[1] However, the involvement of pleura is not common and has not been reported in the literature.

CT scan is the key imaging modality for diagnosing BML. The main findings on a CT scan are nodules with smooth margins and well-defined borders.[7] However, the use of a PET scan has not been commonly reported in the literature. Sawai et al. noted that the uptake of FDG (fluorodeoxyglucose) was associated with aggressive cell proliferation and rapid disease course.[8] In our case, there was increased uptake of FDG that correlated with the aggressive disease course and the fact that the patient passed away within months of diagnosis. An additional benefit rendered by PET scan in our scan was to highlight pleural involvement that was previously not picked on the CT scan.

BML follows a steady disease course and the median survival is 94 months. A more rapid deterioration has been reported in the literature but only scarcely.[1,4] Our case showed marked deterioration within months of initial presentation and passed away within 4 months.

Conclusion

Benign Metastasizing Leiomyoma (BML) is a rare condition but should be considered in females with a prior history of uterine leiomyomas. PET scan is an additional imaging modality that can give key information about disease course. Although most patients have a stable disease course, rapid deterioration can occur.

Lessons Learned

Our case has profound learning lessons with implications for clinical practice. First, despite being rare, BML should be considered in women of late childbearing age with a history of uterine leiomyoma who present with multiple pulmonary or extrapulmonary nodules. Second, although BML most commonly spreads to lungs and lymph nodes, it can spread to atypical places as well (such as pleura in our case). Third, BML follows a steady disease course with a good prognosis but rapid deterioration can occur so patient/family counselling and treatment planning should be done accordingly. Finally, a PET scan can be used as an additional imaging modality that can give additional clues by the possible aggressive nature of the disease.

Conflict of Interest: None

Funding Source: None

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