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Case Report

Metastatic uterine fibroid in postmenopausal woman suspected of leiomyosarcoma: A case report and literature review

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الملخص

الورم العضلي الأملس النقيلي الحميد هو حالة فريدة تظهر بشكل شائع عند النساء في فترة ما قبل انقطاع الطمث، خاصة أولئك اللائي لديهن تاريخ سابق في جراحة أمراض النساء مثل علاج الأورام الليفية في الرحم. تقرير الحالة هذا يحقق في حالة نادرة من الورم العضلي الأملس النقيلي الحميد في أنثى تبلغ من العمر 54 عاما بعد سن اليأس تعاني من كتلة حوض ضخمة كانت تنمو بسرعة جنبا إلى جنب مع العقيدات الرئوية. يشتبه في أن المريض مصاب بساركوما عضلية ملساء والتي تم تأكيدها في النهاية من الناحية المرضية على أنها ورم عضلي أملس نقيلي حميد. على الرغم من ندرته، فإن وصف تحديات التشخيص والمعالجة لمثل هذا الكيان أمر ضروري واعتباره أحد التشخيصات التفريقية للمرضى الذين لديهم تاريخ مشابه.

Abstract

Benign metastatic leiomyoma is a unique event presenting most commonly in premenopausal women especially those with a previous history of gynecological surgery as management of uterine fibroids. In this case report, we investigate a rare case of benign metastatic leiomyoma in a 54-year-old postmenopausal female presented with a huge pelvic mass that was rapidly growing along with pulmonary nodules. The patient is suspected to have leiomyosarcoma which is eventually confirmed pathologically as benign metastatic leiomyoma. Although rare, describing the challenging diagnostic and management approach of such entity is essential and to consider it one of the differential diagnoses of patients who present with similar history.

Keywords: Benign metastasizing leiomyoma; Fibroid; Leiomyosarcoma; Metastasis; Postmenopausal

Introduction

The most prevalent pelvic tumors in females are uterine fibroids (also known as leiomyomas).[1] They are non-malignant monoclonal tumors arising from the smooth muscle cells called the fibroblasts.[2]

While The Uterus is the most common site of origin of leiomyomas, the literature review revealed they could rarely present in almost any site beyond the Uterus, such as the skin, heart and blood vessels, and spine, the lungs, and even the brain.[3,4] These rare metastatic patterns are referred to in the literature by many titles such as benign metastasizing leiomyoma, disseminated peritoneal leiomyomatosis, intravenous leiomyomatosis, parasitic leiomyoma, and retroperitoneal leiomyomatosis.[3]

Although premenopausal women(reproductive age group) are the most commonly affected age group, they can be diagnosed in postmenopausal women but on much fewer occasions.[5]

Furthermore, the mechanism of BML is not well established currently. However, many theories have been suggested, including hormonal stimulation of local fibroblasts of the smooth muscles, coelomic metaplasia, which explains the metastasis of fibroids to sites such as the pericardium, pleura, and peritoneum, and the theory of the lymphatic and hematogenous spread of the tumor cells.[6,7] In this report, we present a unique case of a benign metastasizing leiomyoma to the lungs and pelvis in a postmenopausal woman initially suspected to be leiomyosarcoma necessitating a

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thoracotomy and a bowel resection as part of her challenging diagnostic and treatment approach.

Case presentation

54- year-old Philipino lady who is post-menopause for one year, p1+1. She is diagnosed to have uterine fibroids in 1997. Since then, she underwent extensive inpatient and outpatient medical and surgical management of her fibroids. These managements included two myomectomies in 2005 and 2009, in addition to two uterine artery embolization. She was evaluated by many doctors of many specialities throughout the course of her disease.

Since her diagnosis of uterine fibroids, her symptoms ranged from being asymptomatic to having irregular cycles and pressure symptoms. Her symptoms also included pelvic heaviness and stress urinary incontinence, besides struggling with infertility in her early reproductive years.

Her family history was positive of breast cancer in her mother, who died from complications of the disease; however, no family history of gynecological or gastrointestinal system malignancies.

Moreover, all her physical examinations were normal apart from a firm, irregular pelvic mass reaching above the level of the umbilicus corresponding to a 22-week size uterus. A concerning finding that pelvic CT confirms.

Given this patient's postmenopausal status, worrisome radiological findings, and rapidly growing mass, she was referred to a gynecological oncology center for further evaluation and a definitive management plan.

At the oncology center, CT chest, abdomen, and pelvis were done, and pelvic MRI.CT showed the Uterus enlarged by innumerable masses of variable size; some masses show calcifications and cystic degeneration. The uterus along with the masses measuring 22.0 × 14.1 × 27.3 cm (in ML x AP x CC). There is also a large left adnexal multiloculated complex cystic lesion with internal septation and soft tissue enhancement, measuring about 13 × 10 × 14 cm (in ML x AP x CC), mostly from the left ovary. The pelvic mass is causing a pressure effect on the urinary bladder, sigmoid colon, displacing the small bowel, and invasion to these organs could not be ruled out. There are also peritoneal nodules largest is measuring 3 × 3 cm as well as multiple sub-centimetric retroperitoneal and pelvic lymph nodes (Figures 1,2).

The CT chest revealed small few bilateral pulmonary nodules; the largest is in the lower right lobe measuring 1.1 cm (Figure 3).

Following these CT findings, MRI was obtained to differentiate the tissue of origin, which revealed a large left adnexal multiloculated complex cystic lesion with internal separations and soft tissue enhancement, measuring about 11.5 × 13.7 × 24 cm in ML x AP x CC respectively, mainly arising from the left ovary. The uterine masses and left adnexal mass are adherents with suspicious extra serosal extensions. For example, it extends to the left pelvic sidewall. There are also irregularities along the right lower abdomen and right iliac fossa suggestive of peritoneal disease (Figure 4).

In addition, all her laboratory investigations were normal, including tumor markers CA125, CA19-9, and CEA.

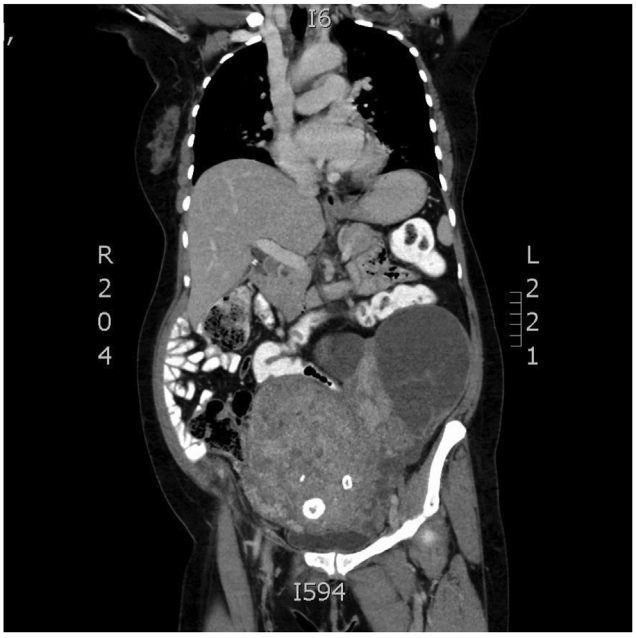

Figure 1: CT abdomen showing an enlarged uterus and a left adnexal cystic lesion.

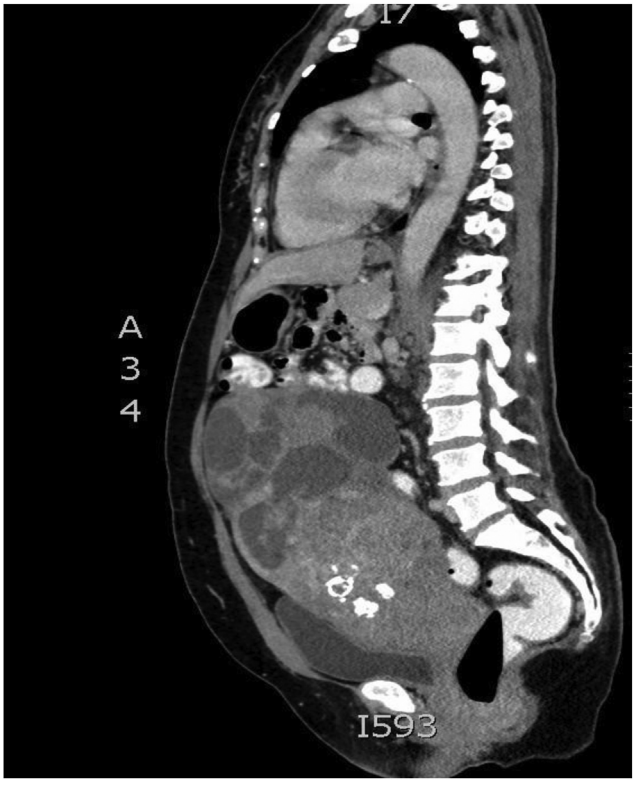

Figure 2: CT abdomen showing a huge uterine mass extending to the umbilicus.

Following the worrisome results of the radiological evaluations, to get more insight into the diagnosis, the patient underwent a pelvic mass biopsy and a lung nodule

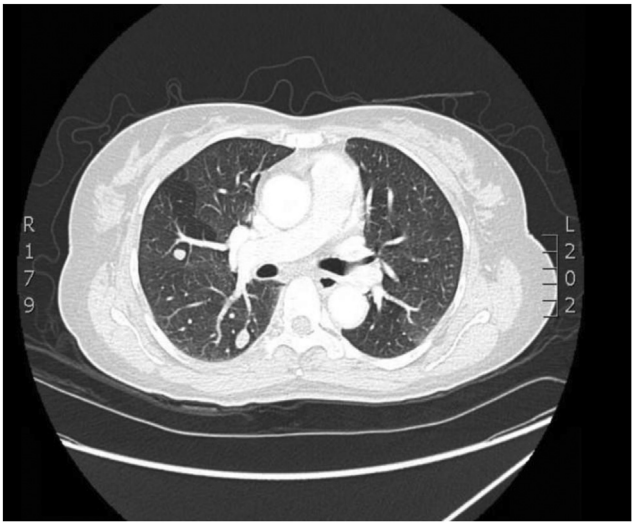

Figure 3: CT chest showing pulmonary nodules.

biopsy. The pelvic biopsy was obtained by interventional radiology, and the lung biopsy was obtained using a right video-assisted thoracoscope (VATS). VATS was attempted by thoracic surgery and was converted to mini-thoracotomy wedge resection of the right lower lobe nodule due to difficult accessibility. Furthermore, the pathology result of pelvic mass and lung nodule biopsy revealed spindle cell neoplasm without evidence of atypia, necrosis, or increased mitotic figures in keeping with leiomyoma. In addition, immunohistochemistry also resulted positive for Desmin and smooth muscle actin (SMA), confirming their smooth muscle origin. Also, estrogen receptor was (ER) positive and Ki67:2%.

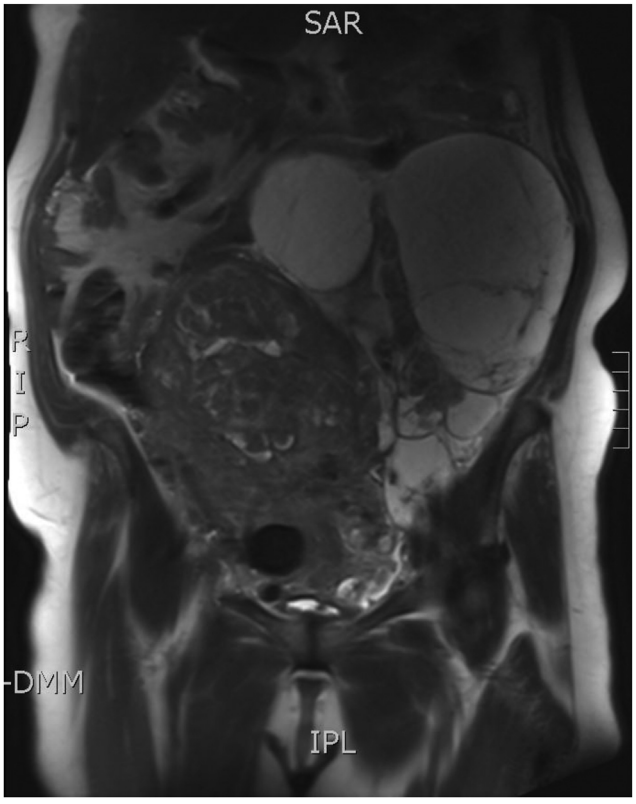

Figure 4: Pelvic MRI showing a huge uterine mass and a left adnexal complex cyst.

Consequently, the patient underwent laparotomy. She is found to have an enlarged uterus with multiple masses, enlarged left adnexa, and the whole Uterus and the left adnexa forming one huge mass reaching above the umbilicus. The mass was severely adherent to the cecum, sigmoid colon, and a loop of the distal ileum. There were also multiple scattered pelvic masses, mainly cystic along the left and right pelvic sidewall. In addition, there were massive adhesions that needed extensive adhesiolysis. However, the procedure was successful and ended in a total abdominal hysterectomy with bilateral salpingo-oophorectomy, tumor debulking, and opportunistic omentectomy and appendectomy. In addition, bowel resection and anastomosis of a loop of small bowel that was severely adherent to the pelvic mass was performed. Also, repair of some serosal injuries of the cecum and sigmoid colon. The resulted final pathology for all the following: Pelvic masses, Uterus, adnexa, and Adnexal mass were all consistent with the biopsy results confirming a benign leiomyoma—however, some with cystic degeneration and no evidence of malignancy. The omentum, appendix, and small bowel were also negative for malignancy.

Eventually, the patient was planned to continue her follow-up in the outpatient setting with a chest CT as a surveillance method for her asymptomatic pulmonary nodules. She was evaluated 4 months later in stable condition with a repeat CT scan showing neither disease progression nor recurrence.

Discussion

The first case of BML ever reported in the literature was described by Steiner in 1939.[8] It was a case of a patient who unfortunately passed away from severe lung metastasis.[8] A metastasis proved later to have the same histology as those concurrently present in her Uterus.[8] It is an uncommon event defined by the presence of histopathologically benign fibroids in sites beyond the original site, which is the Uterus.[9] The most common extra-uterine site is the lung, and the most affected patients are those in their late reproductive years.[9] Our case presents an unusual late presentation as she is postmenopausal.

Interestingly, this disorder commonly presents in patients with a positive history of therapeutic hysterectomy for uterine fibroid or a myomectomy.[9] This raises the suspicion that previous uterine surgery may play a role in such a condition, probably by seeding of the primary tumor.[9,10] Almost all reported cases of metastatic leiomyoma in the literature demonstrated a positive history of uterine surgery many years preceding the diagnosis of BML.[9]

A most recent literature review revealed the mean age of the initial uterine surgery is 38.5 and the mean age at which BML is diagnosed is 47.3.[11] Furthermore, the mean time interval from primary surgery to BML diagnosis is about nine years.[9] This time interval can extend up to 31 years following initial surgery[12] and surprisingly early at the time

of primary surgery.[13–15] This data is proved compatible with our case, which is diagnosed 16 years following her initial surgery.

Although Patients with lung metastasis can present with the usual upper and lower respiratory symptoms such as chest pain, dyspnea, and cough,[16] most cases are symptomless and are discovered on routine evaluations.[17] This is consistent with our case, diagnosed incidentally on CT chest as part of her radiological evaluation in suspicion of leiomyosarcoma. Furthermore, these lung nodules can be identified on conventional CT as round well-circumscribed non-calcified masses of variable sizes and locations within the lung; they can also appear on X-ray as opacifications, either single or multiple.[18] Furthermore, whole-body PET CT with 18F-FDG (fluorodeoxyglucose) is being used more widely to help in distinguishing a benign from a cancerous lesion by testing the avidity of (FDG) which is expected to be increased in malignant lesions.[19]

However, the confirmative diagnosis depends on both histopathologic and immunohistochemical examination of the tissue sample.[18] There are essential for both identifying the features of BML and excluding other malignant conditions, mostly sarcomas.[18] BML exhibits the same features of benign uterine fibroid histopathologically regarding well-differentiated spindle-shaped smooth muscle cells with no evidence of necrosis, significant atypia, or high mitotic figures[20] In addition, immunohistochemically in terms of positive tissue markers (smooth muscle actin SMA, Ki-67 index (<5%), Desmin, hormone receptor-positive for estrogen and progesterone).[17,21]

Up to date, there are no established guidelines on the treatment of BML due to its rarity.[22] Nevertheless, the literature review revealed several proposed modalities of treatment ranging from close observation to oophorectomy and complete surgical resection, as well as hormonal therapy such as GnRH agonists, progesterone, and aromatase inhibitors,[23–25] but these medications' efficacy is yet to be well studied.[26] Furthermore, some authors suggested the mainstay of the treatment of BML is total hysterectomy with bilateral salpingo-oophorectomy and complete resection of the disease as it is hormonally dependent.[27] However, it should be individualized depending on many factors, including the location of the metastasis, their sizes and number, symptoms of the patient, hormone receptor status, disease progress with time,[18] and the desire for future fertility.[28]

Our case presents a unique diagnostic challenge given her menopausal status and rapidly growing metastasis in few months, raising the suspicion of leiomyosarcoma. It also presents a tremendous surgical challenge as her pelvic metastasis was extensive, requiring a midline incision and bowel resection with end-to-end anastomosis. Furthermore, the complexity of this case required incorporating a multidisciplinary team for obtaining a biopsy from a hardly accessible location within the lung and pelvis.

It is essential to keep in mind when choosing surgery to select an approach that will permit complete resection of the disease to minimize its chances of recurrence.[18] This recurrence risk is thought to be higher in patients with BML due to their disease history.[18]

Even though the favorable prognosis of this entity,[16] postmenopausal women might experience less favorable outcomes as the case demonstrated by Barber et al.[11] who showed disease progression despite hormonal therapy. Thus; a long-term follow-up is necessary by radiological imaging either with MRI or CT.[22]

Conclusion

BML presents a challenging diagnostic and management approach as it tends to emulate cancer in distant metastasis. Although most data regarding the treatment of such entity are based on case reports and case series, physicians should have insight into BML and incorporate it in the differential diagnosis. Most importantly, the need to rule out malignancy to establish an accurate diagnosis and, therefore, to provide appropriate treatment and avoid over-management. Long-term follow-up is advised.

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Conflict of interest

The author has no conflict of interest to declare.

Ethical approval

This study didn't require ethical approval by the ethical committee and there is no ethical issues to disclose.

Consent

The patient provided their consent for publication.

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