Benign pelvic metastatic leiomyoma: case report

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Summary

Benign metastasizing leiomyoma is a rare condition characterized by benign soft tissue tumors most frequently involving the lung, and is usually associated with a benign leiomyoma or intravenous leiomyomatosis of the uterus. We present a case of a 58-year-old female patient with abdominal pain and symptoms of urinary tract infection four years after hysterectomy due to uterine fibroid. The results of CT revealed a pelvic mass. Pathological examination confirmed that it was a metastatic pelvic benign metastasizing leiomyoma (BML). BML only involving the pelvis is extremely rare. The patient underwent surgical resection and recovered well.

Key words: Benign metastasizing leiomyoma; Pelvic cavity; Intravenous leiomyomatosis.

Introduction

Benign metastasizing leiomyoma (BML) is an uncommon condition characterized by the presence of multiple soft-tissue tumors most frequently affecting the lung [1]. Rare cases involving other anatomical locations including the heart, lymph nodes, omentum and mesentery, bone, peritoneum, pelvic cavity, soft tissue and skeletal muscles of the back and chest wall, breast, or brachial plexus have also been reported [2]. Although cases of BML affecting both lung and pelvis have been documented, to the best of our knowledge, a case involving only the pelvis has yet to be reported. A case of metastatic pelvic leiomyoma is presented.

Case Report

The patient was a 58-year old female, admitted to the Department of Urology at our hospital on August 4, 2011 with the complaint of two-weeks of right abdominal pain exacerbated in the previous four days, and urinary frequency/urgency, and urodynia for three days. The patient started experiencing lower abdominal distending pain without obvious cause for half a month. The pain gradually exacerbated with increasing frequency four days prior to the admission. Urinary frequency, urgency, and urodynia also started three days before the admission. Computed tomography (CT) scan at the outpatient unit revealed a mass in the right ureter. The patient was admitted to the department of urinary surgery in our hospital with the diagnosis of a right ureter unknown mass.

Physical examination showed right side lateral abdominal tenderness (+), right ureter tenderness (+), and bladder area percussion (+). The patient had undergone hysterectomy and oophorectomy due to uterine fibroids confirmed by pathology in 2007 at the Beijing Maternity Hospital. The diagnosis upon admission to our hospital was right ureter unknown mass/urinary tract infection. Doppler ultrasound (US) exam of the urinary tract system showed right kidney hydronephrosis, and extension of the upper section of the right ureter. Pelvic US indicated 3.7 × 3.3 cm hypoechoic mass with unclear border at the right pelvis. Pelvic CT showed a 3.76 × 3.39 cm solid mass (tumor) in the right adnexal area compressing the distal end of the ureter and causing hydrocele above a section of the ureter, pelvis and kidney (Figure 1). Magnetic resonance imaging (MRI) showed a 4.11 × 4.27 × 4.32 cm mass in the right adnexal area, full dilation of the renal pelvis and ureter caused by obstruction of the right lower ureter near the entrance to the bladder (Figure 2). On August 11, 2011 cystoscopy under local anesthesia was performed. No apparent signs of tumor mass in the bladder and ureteral orifice were identified. On August 17, 2011 the patient was transferred to the Department of Gynecology with the diagnosis of metastatic leiomyoma. On August 23, 2011 exploratory laparotomy was performed under epidural anesthesia. An adhesion between the ileum, colon, bladder and pelvic tumor were identified. A firm tumor without clear border about 4 × 4 cm in size was found surrounding the ureter at the entrance to the bladder with strong adhesion to both the bladder and vagina. A sample of tumor tissue was taken for rapid frozen pathology showing benign tumor with focal calcification. The patient then underwent resection of the pelvic tumor, partial ureter, ureteral bladder anastomosis, vesicostomy, and enterolysis. Postoperative pathological exam showed pelvic leiomyoma, partially actively growing, with hyaline degeneration and focal calcification (Figure 3). The patient was discharged 20 days after the surgery and has continued to be followed. She is in good condition and has returned to normal work. The report was approved by the hospital ethics committee and consented to by the patient.

Discussion

BML, intravenous leiomyomatosis (IVL) and leiomyomatosis peritonealis disseminata (LPD) are three unique growth patterns of uterine smooth muscle tumors. BML is a rare lesion affecting females with a history of uterine leiomyomata. In 1937, Steiner reported the first patient who died from multiple uterine leiomyoma. At the autopsy, well-differentiated smooth muscle nodules were found in the lung, therefore the concept of benign metastatic smooth muscle tumor was introduced [3]. There were several hypotheses regarding the origin of the lesion: a benign uterine tumor spreading through the hematogenous route to the lung or other organs as the majority of BML patients have had prior myomectomy or hysterectomy; it could be a low-grade leiomyosarcoma metastasiz-

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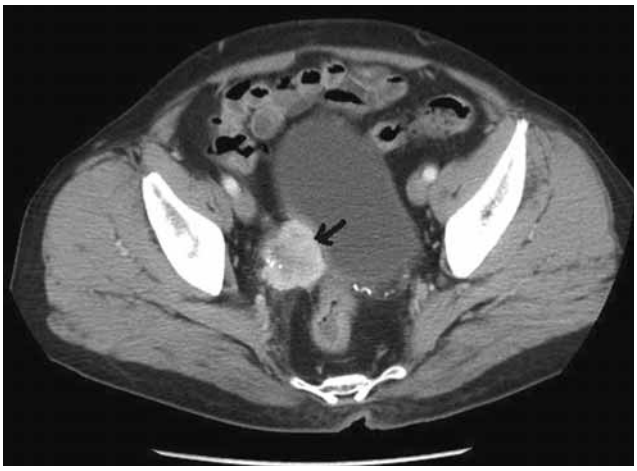

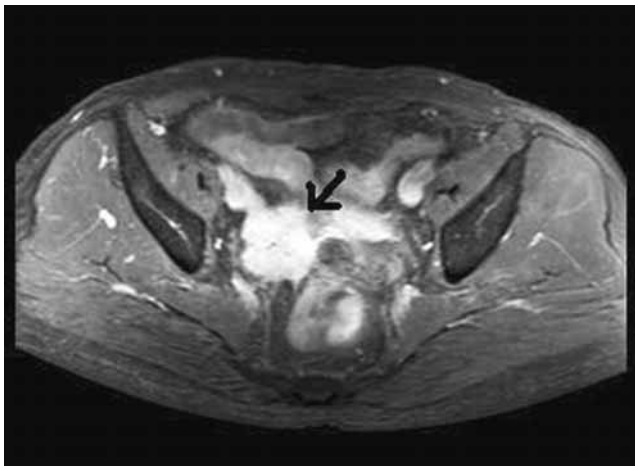

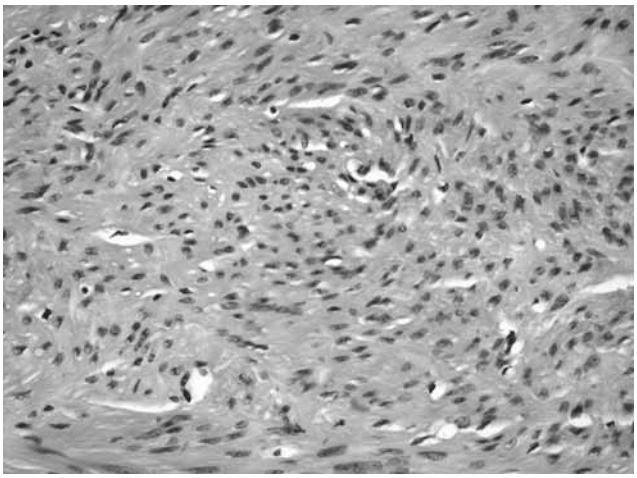

Figure 1. — CT indicating a solid mass of the right pelvis.
Note: arrow (↙) points to the solid mass. CT: Computed tomography.

Figure 2. — Pelvic MRI
Note: arrow (↙) points to the solid mass. MRI: Magnetic resonance imaging.

Figure 3. — Pathological exam indicating pelvic leiomyoma.
Note: Hematoxylin and eosin stain × 400.

ing to the lung; or primary pulmonary leiomyomatosis unrelated to but accidently coexisting with uterine leiomyomata [4].

Most BML patients are either asymptomatic or show only mild symptoms, such as slight cough, chest pain, dyspnea, and fatigue, if there is metastasis to the lung. In our case, the tumor surrounded and compressed the patient's ureter, which resulted in hydronephrosis and secondary infection, and the patient was admitted to the Department of Urology with the symptoms of abdominal pain, urinary frequency urgency, dysuria.

The characteristics of BML remain controversial. Most researchers tend to agree that it is a benign lesion. Disease progression is slow. Most patients have either no clinical symptoms or only mild symptoms, and have been identified accidentally during physical examination. Patients can fully recover after appropriate endocrine treatment or surgical resection of the tumor. For postmenopausal women, the tumor either grew slow or self-resolved. However, some researchers questioned that the pathological examination of uterine leiomyoma reported so far lacked details, such as insufficient sample collection, no record of karyokinesis; or the degree of malignancy could not be determined due to the limitations of currently available pathological tests. Therefore, they speculated that BML might originate from the uterine smooth muscle tumors with unknown malignancy, possibly between benign and low grade of malignancy [5].

For histological origin, most of the literature reports that the lesion is metastasis of a benign uterine leiomyoma to the lung or other organs. The evidence supporting this viewpoint include: 1) from the pathomorphological perspective, metastatic tumors and primary uterine tumors share similar histomorphological and immunohistochemical features, and all are hormone-dependent [6]; 2) Kayser's research [7] of agglutinin histochemistry in 10 BML cases showed that the majority (80%) of BML expressed galectin-1, galectin-3 and their binding sites; only the expression of galectin-8 demonstrated individual variation, which suggested that these tumors had a common origin; 3) Compared with uterine leiomyoma, pulmonary BML also have similar patterns of androgen inactivation allele [8]; 4) Primary uterine leiomyoma and lung BML also have the same molecular genetic changes – the same X-chromosome inactivation pattern, proving that lung BML is metastasis of uterine leiomyoma, and they are the same clone [8-10]; 5) The results of molecular biology research, including CGA repeat polymorphism detection and analysis of X chromosome activity, showed both tumors have chromosome

19q and 22q terminal deletion, which also supported the homology of metastatic leiomyoma and uterine leiomyoma [11]. Since most BML cases also had the history of uterine curettage, myomectomy or hysterectomy, it was suggested that the tumor might be disseminated by surgery [12]. For the patient in our report, the tumor was located at the right ureter, close to the entrance of the bladder. It was a nodular mass 5 × 6 × 6 cm in size, grayish ash in color with interlacing structure on the section. Histopathological findings were leiomyoma with adiponecrosis, degeneration, small cysts, and focal calcification. There was no significant nuclear atypia, mitotic activity or necrosis. Immunohistochemistry showed positivity on vimentin, EMA, SMA, actin, and desmin, which proved its origin of smooth muscle, and the tumors were also estrogen and progesterone receptor positive. The patient had had surgery for uterine leiomyoma four years before. Upon reviewing the pathology slide, there was no significant difference in histology between a pelvic metastatic tumor and primary uterine leiomyoma. Given the pathological findings and location of the tumor, it is likely that the metastasis was the result of surgery.

The diagnosis of BML relies mainly on pathology. For women of childbearing age who have a history of myomectomy or hysterectomy, the findings of nodular and diffuse lesions of the lung or other organs may suggest this disease. The first consideration is whether the primary tumor is malignant. Sometimes, the malignant area is limited; incomplete sample collection may lead to misdiagnosis. Sometimes well-differentiated leiomyosarcoma has been diagnosed as a tumor alive with cells. Therefore, only when the possibility of malignancy has been ruled out, can the spread and metastasis of benign tumors be confirmed. Some researchers have suggested positron emission tomography-computed tomography examination to identify hidden metastatic lesions [13, 14].

Since the report of BML is limited in the literature, currently there is no standard treatment. The main treatment strategy is the surgical removal of the tumor as completely as possible. Because tumor cells express estrogen, hormone therapy has also been attempted [15] using antiestrogen drugs, such as Raloxifene, LHRH or GnRH-a, but efficacy varied, and for some cases, despite strong expressions of estrogen, progesterone, there was no significant treatment effect [16]. During the postoperative follow-up, our patient did not receive any other treatment, and she has now returned to normal work, without any discomfort. We will continue to follow her.

Conclusion

BML is a rare, slow progressing disease, occurring mostly in women of childbearing age who have had history of surgery due to uterine leiomyoma. Lung and lymph nodes are the most common sites of metastases. BML only involving the pelvis is extremely rare. We reported a case of pelvic BML who underwent surgical resection and recovered well. She will continue to be followed.

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