Leiomyomatosis with multiple extrauterine pulmonary sites: an unusual case report

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Summary

The objective of the present study is to present a case of a 48-year-old woman with leiomyomatosis and multiple pulmonary metastases. The classification, pathophysiology, clinical signs, treatment and prognosis of this rare case are discussed. Leiomyomatosis is potentially life-threatening while patients with pulmonary metastases are usually asymptomatic and the condition is incidentally discovered. The treatment of leiomyomatosis is not standardized and many possible variations are under investigation. Still the prognosis is usually excellent.

Key words: Leiomyomatosis; Pulmonary masses.

Introduction

Leiomyoma or fibroid [1] is a growth of the muscular wall of the uterus which is benign most of the time (> 99%). Uterine leiomyomas are the most common gynecological tumors especially in the fourth and fifth decades of life. They are composed of smooth muscle cells with varying amounts of fibrous connective tissue. Leiomyomas occur in 20% to 30% of women of reproductive age. They are thought to arise from a somatic mutation of a monoclonal myometrial cell line. Myomas are sensitive to estrogen levels as they contain both progesterone and estrogen receptors. They usually grow during a woman's reproductive period and frequently shrink after menopause.

Metastatic leiomyomatosis is a rare but potentially life-threatening situation. It presents post-hysterectomy with multiple extrauterine metastatic sites, most commonly pulmonary [2-5], but also the inferior vena cava and right heart can be affected [6]. Other possible metastatic sites are the lymph nodes, peritoneum and retroperitoneum [7].

Case Report

We present a case of a 48-year-old woman (gravida 2, para 2) who presented at our gynecological clinic with the suspicion of leiomyomatosis with multiple pulmonary sites. The patient had a non-productive cough. Her previous medical history included the removal of a leiomyoma of the uterus (5 cm) two years prior to presentation. Vital signs were within normal limits. Gynecological examination revealed a pelvic tumor 6 cm in diameter. Chest X-ray showed multiple bilateral pulmonary nodules. Ultrasonographic examination revealed a pelvic tumor 7 cm in diameter. The chest and abdominal computed tomography confirmed the aforementioned findings.

The patient underwent an exploratory laparotomy. Total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed. The postoperative period was uneventful.

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Pathologic examination revealed that the tumors were composed of smooth muscle cells without mitotic activity. Histology confirmed metastatic leiomyomatosis (Figure 1). A decrease in hormone levels led to stable pulmonary lesions with an excellent prognosis after three years of follow-up.

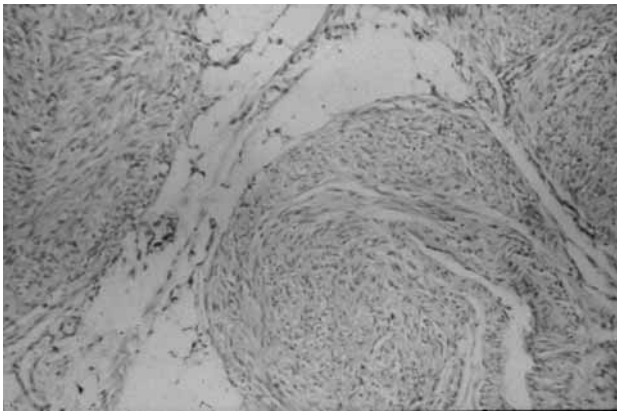

Figure 1. — Leiomyomatosis of the uterus.

Discussion

Our patient had leiomyomatosis in combination with multiple extrauterine pulmonary metastatic sites. According to Martin's classification system three types of smooth muscle tumors exist [8]: leiomyomatosis, metastatic leiomyoma, and multiple fibroleiomyomatous hamartomas. Leiomyomatosis occurs exclusively in women with uterine leiomyomas and is further subdivided into benign metastasizing leiomyoma, lymphangiomyomatosis (lymphangioleiomyomatosis), disseminated peritoneal leiomyomatosis, and intravenous leiomyomatosis.

There is a proposed classification system for multiple lung smooth muscle lesions:

a) benign metastasizing leiomyoma: uterine source in mature women;

[5] Kadry M., Sievers C., Engelmann C.: "Pulmonary leiomyomatosis in women after hysterectomy for uterine myoma. Benign metastasizing leiomyoma?". *Acta Chir. Hung.,* 1999, *38,* 57.
[6] Thukkani N., Ravichandran P.S., Das A., Slater M.S.: "Leiomyomatosis metastatic to the tricuspid valve complicated by pelvic hemorrhage". *Ann. Thorac. Surg*., 2005, *79*, 707.
[7] Dursun P., Salman M.C., Taskiran C., Yuce K., Ayhan A.: "Retroperitoneal leiomyomatosis: a case report". *Int. J. Gynecol. Cancer,* 2005, *15,* 1222.
[8] Martin E.: "Leiomyomatous lung lesions: a proposed classification". *Am. J. Roentgenol*., 1983, *141*, 269.
[9] Birsh-Hirschfeld F.V.: "Lehrbuch der Pathologischen Anatomie". Vol 1. 5th edn. Leipzig, Germany, Vogel, 1896, 226.
[10] Steiner G., Warren J.W., Judd A.S.: "Intravenous leiomyomatosis. A case report". *Am. J. Obstet. Gynecol.,* 1963, *87,* 166.
[11] Canzonieri V., D'Amore E.S., Bartoloni G., Piazza M., Blandamura S., Carbone A.: "Leiomyomatosis with vascular invasion. A unified pathogenesis regarding leiomyoma with vascular microinvasion, benign metastasizing leiomyoma and intravenous leiomyomatosis". *Virchows Arch*., 1994, *425*, 541.
[12] Nardo L.G., Iyer L., Reginald P.W.: "Benign pulmonary metastasizing leiomyomatosis in pregnancy: a rare complication after cesarean section". *Acta Obstet. Gynecol. Scand.,* 2003, *82,* 770.
[13] Abu-Rustum N.R., Curtin J.P., Burt M., Jones W.B.: "Regression of uterine low-grade smooth-muscle tumors metastatic to the lung after oophorectomy". *Obstet. Gynecol.,* 1997, *89,* 850.
[14] Abramson S., Gilkeson R.C., Goldstein J.D., Woodard P.K., Eisenberg R., Abramson N.: "Benign metastasizing leiomyoma: clinical, imaging, and pathologic correlation". *Am. J. Roentgenol*., 2001, *176*, 1409.
[15] Canzonieri V., D'Amore E.S., Bartoloni G., Piazza M., Blandamura S., Carbone A.: "Leiomyomatosis with vascular invasion. A unified pathogenesis regarding leiomyoma with vascular microinvasion, benign metastasizing leiomyoma and intravascular leiomyomatosis". *Virchows Arch.,* 1994, *425*, 541.
[16] Lipton J.H., Fong T.C., Burgess K.R.: "Miliary pattern as presentation of leiomyomatosis of the lung". *Chest,* 1987, *91*, 781.
[17] Shin M.S., Fulmer J.D., Ho K.J.: "Unusual computed tomographic manifestations of benign metastasizing leiomyomas as cavitary nodular lesions or interstitial lung disease". *Clin. Imaging*, 1996, *20,* 45.
[18] Abramson S., Gilkeson R.C., Goldstein J.D., Woodark P.K., Eisenberg R., Abramson N.: "Benign metastasizing leiomyoma: clinical, imaging and pathologic correlation". *Am. J. Roentgen., 2001, 176,* 1409.
[19] Maredia R., Snyder B.J., Harrey L.A., Schwartz A.M.: "Benign metastasizing leiomyoma in the lung". *Radiographics,* 1998, *18,* 779.
[20] Matsumoto K., Yamamoto T., Hisayoshi T., Asano G.: "Intravenous leiomyomatosis of the uterus with multiple pulmonary metastases associated with large bullae-like cyst formation". *Pathol. Int*., 2001, *51,* 396.
[21] Kullo I.J., Oh J.K., Keeney G.L., Khandheria B.K., Seward J.B.: "Intracardiac leiomyomatosis: echocardiographic features". *Chest.,* 1999, *115,* 587.

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