Case Report

Management of Intravenous Leiomyomatosis of Uterus with Extension to Heart

Kambiz Alizade MD[1], Godratollah Maddah MD[2], Amir Hosein Jafarian MD[3], Arash Khamene Bagheri MD[4], Reza Jafarzadeh Esfehani MD[5], Sara Mirzaeian MD·[6]

Abstract

Leiomyoma is a benign smooth muscle tumor. Intra-venous extensions of these tumors occur due to tumor growth within uterine vein or lymphatic vessels. In rare cases, intracaval and intracardiac extension can also be seen. Clinical suspicion of this disease should become certain by use of imaging techniques. While the treatment is complete resection of the tumor, one or two-stage surgery can be planned for patient depending on tumor extension and patient's condition. In this report, a 52-year-old woman with a rare presentation of uterine leiomyoma will be discussed. While the tumor was extended toward right atrium, the patient had nonspecific symptoms. By use of two-stage surgery, separated laparotomy and cardiopulmonary bypass, the tumor was completely removed.

Keywords: Cardiopulmonary bypass, leiomyomatosis, laparotomy

Cite this article as: Alizade K, Maddah G, Hosein Jafarian A, Khamene Bagheri A, Jafarzadeh Esfehani R, Mirzaeian S. Management of Intravenous Leiomyomatosis of Uterus with Extension to Heart. *Arch Iran Med*. 2016; 19(2): 147 – 149.

Introduction

Intravenous leiomyomatosis (IVL) is a benign uncommon tumor which can extend to cardiac cavities and cause various complications.[1] Surgical resection of the tumor is the choice treatment and can prevent venous thromboembolism or sudden death.[2,3] In the present paper, we will discuss a case of IVL extending to the right atrium of a 52-year-old woman presenting with abdominal pain.

Case report

A 52-year-old woman referred to our gynecologic clinic because of a tender abdominal mass, growing gradually from 4 years ago. The mass was located in the hypogastric area, extending to her periumbilical area. The patient also complained of an exertional pain around the hypogastric area and occasional vaginal bleeding since 3 months ago. Vital signs were stable without any abnormal laboratory finding. Ultrasonography study revealed a heterogeneous mass with a diameter of 113 × 213 millimeters in her uterus. The mass extended over her umbilicus. An echogenous mass with diameter of 37 millimeters was seen in inferior vena cava (IVC), resembling venous thrombosis. The mass caused partial obstruction of IVC around hepatic vein origin while supra hepatic veins were open. Also, a solid mass was seen in IVC above the renal vein, extending toward right atrium and causing partial obstruction. Transthoracic echocardiography (TTE) showed a lobulated mass within abdominal IVC, extended above diaphragm toward her right atrium. The mass had dilated and occupied approximately 90% of IVC. Myocardial perfusion scan (Dipyridamol-technetium-99 m-MIBI) showed ischemia of inferior wall with left ventricle ejection fraction of 60%. Computed tomography (CT) also showed a huge 191 × 124 × 207 millimeter nonhomogeneous multilobulated mass within the uterus. The CT scan showed a heterogeneous mass with necrotic tissue which could not be differentiated from uterus and resembled uterine fibromatosis (Figure 1). There was a thrombotic mass consistent with IVC region extending proximally toward cardiac chamber (Figure 2). The slight enhancement of the thrombosis resembled tumoral thrombosis of IVC. According to our diagnostic work-up and with possible diagnosis of uterine leiomyoma extending to IVC, the patient underwent laparotomy by abdominal midline incision above and below umbilicus. A giant tumor originated from the uterus. The tumor firmly adhered to the surrounding tissue. The tumor had vascular extension, extending from ovarian vein toward IVC. After bilateral ureterolysis, ovarian vessels were ligated and total hysterectomy was performed. The tumor was resected and the specimens were sent for pathology study. After omentectomy, and performing adequate hemostasis, IVC was exposed by Cattell-Braasch maneuver and the tumoral portion was removed approximate to thoracic inlet by hand. Then, the IVC was repaired carefully. Due to the possibility of heart chamber involvement as well as the prolongation of the surgery, the surgery team decided to consult with a heart surgeon. So, the operation was terminated and a heart surgery was scheduled for later. Two months later, a TTE was performed for the patient and a huge homogenous 3 × 2.7 centimeter mass was seen in right atrium extending toward abdominal and hepatic course of IVC. The mass had obliterated more than 90% of IVC canal. There was no mass in right ventricle. So, the patient was scheduled for an open heart surgery. Under general anesthesia with mild hypothermia (34°C) and heparinization (3 mg/Kg), me-

Authors' affiliations: [1]Assistant professor, Department of open heart surgery, School of medicine, Mashhad University of medical sciences, Mashhad, Iran. [2]Associated professor, Department of surgery, School of medicine, Mashhad University of medical sciences, Mashhad, Iran. [3]Assistant professor, Department of Pathology, School of medicine, Mashhad University of medical sciences, Mashhad, Iran. [4]Assistant professor, Sabzevar University of Medical science, Sabzevar, Iran. [5]Medical Student, Sabzevar University of medical science, Sabzevar, Iran. [6]Assistant professor, Department of obstetrics and gynecology, School of medicine, Mashhad University of medical sciences, Mashhad, Iran.

·Corresponding author and reprints: Sara Mirzaeian, Assistant professor, Department of obstetrics and gynecology, School of medicine, Mashhad University of medical sciences, Mashhad, Iran. Tel: +985138012477, E-mail: Mirzaeians@mums.ac.ir

Accepted for publication: 17 October 2015

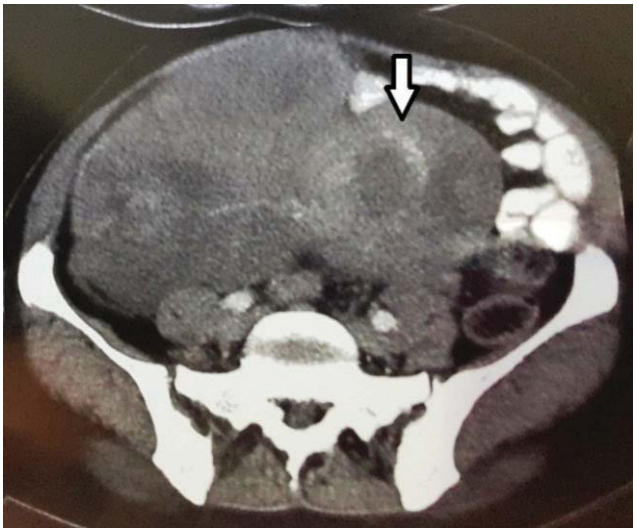

Figure 1. Abdominal Computed Tomography scan with contrast. The arrow shows the uterine mass.

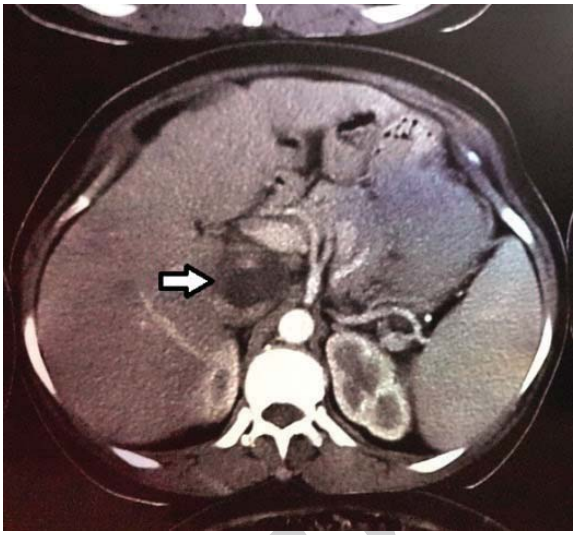

Figure 2. Abdominal Computed Tomography with contrast. The arrow shows the tumoral mass within inferior vena cava.

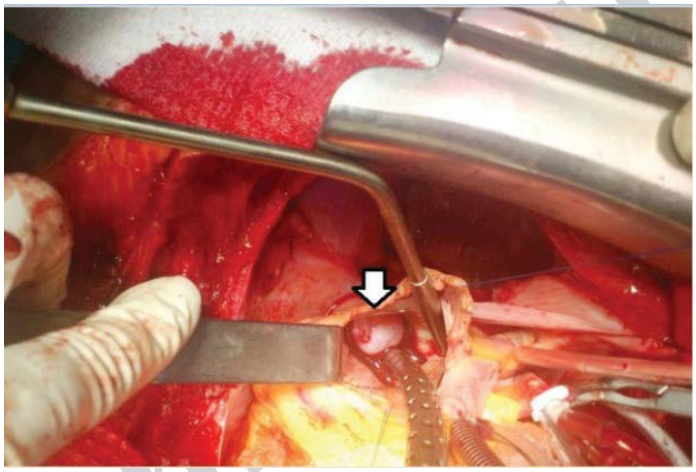

Figure 3. The arrow shows the intra cardiac mass at the time of surgery.

dian sternotomy was performed. The circulation was arrested by hyperkalemic crystalloid cardioplegia. A cardiopulmonary bypass was established between aorta and forearm, superior vena cava (SVC) and right femoral vein. Then right atriotomy and longitudinal venotomy of IVC was performed. Unlike the intra-caval lesion which was firmly attached to vessel wall, the intracardiac mass was not attached and removed easily. By aid of a foley catheter balloon and use of finger, the white solid intra-caval tumor stretching from suprarenal vein to the atrium (1.5 × 15 centimeters) was successfully resected (Figure 3). Aortic cross clamp, cardiopulmonary bypass and circulation arrest times were 60, 65, and 60 minutes, respectively. Three units of blood were transfused to the patient and both atrium and vena cava were repaired. After rewarming and closing sternotomy, the patient was sent to the intensive care unit and was extubated the next day. Six days after surgery, the patient was discharged.

Histological findings showed fascicle of spindled cells with eosinophilic cytoplasm and bland elongated nuclei with blunt end appearance without mitotic activity. On the immunohistochemical study, the tumoral cells were positive for actin, desmin, vimentin and were negative for s100 protein, which had confirmed the diagnosis of leiomyoma. Also, the histological finding of cardiac lesion was in accordance with myxoma.

Discussion

While the most frequent neoplasm in female genital tract is leiomyoma, IVL is a histopathologicaly benign and rare variant.[4,1] Despite its benign histopathology, it is not essentially benign.[5,1] Diagnosis of IVL requires higher levels of suspicion because of its rarity and nonspecific presentations.[6] Clinical presentation of IVL varies in different patients.[7] Approximately 13% of intracardiac leiomyoma patients are asymptomatic.[7] Depending on the extension of tumor, cardiac insufficiency can occur in a previously healthy individual.[7] Other common symptoms are syncope, dyspnea, fatigue, ascites, hepatomegaly or even chest pain.[7] In our case, despite the far extension of tumor, there were not any cardiac symptoms, only an abdominal mass and occasional vaginal bleeding. Caval thrombosis and tumoral thrombosis are two main differential diagnoses of this disease.[6] Distinguishing leiomyosarcoma is not possible by use of diagnostic tools prior to surgery.[1] In middle aged women with uterine leiomyoma or history of myo-

mectomy/hysterectomy, right atrial masses warrant further evaluation and presence of intracardiac leiomyoma should always be kept in mind.[7] Performing an abdominal computed tomography (CT) can be helpful in determining the origin of tumor.[7] Magnetic resonance imaging (MRI) and magnetic resonance venogram (MRV) are appropriate for assessing the lesion extension in order to make better surgical plans.[5,8] As IVL does not usually invade blood vessels, it can be simply removed by downward traction from involved vein.[9] In cases with intracardiac extension, performing a thoracoabdominal surgery with removal of tumor from heart chamber and inferior vena cava is preferred.[9] One-stage surgery can provide complete resection without any residue and in patients with poor conditions due to cardiopulmonary comorbidities, the optimal choice would be two-stage surgery.[10] Intravenous and intracardiac leiomyomatectomy can be the first stage, and the second stage could be pelvic resection and hysterectomy.[3] It is not recommended to remove the entire tumor from a sternotomy and leave the caval attachment sites uncontrolled.[11] In our patient, we first decided to remove the abdominal tumor but because of prolongation of the surgery and instability of patient's conditions, we decided to end the procedure and leave the intra-thoracic removal for future. While one-stage operation can reduce thrombotic event and completely remove tumor, prolonged operation, more bleeding and post operation complications are some of its disadvantages.[12] Since IVL is loosely attached to vessel walls, removal of intracardiac and intracaval lesions could be achieved easily.[12] Complete removal of tumor can guarantee no recurrence and excellent outcome; however, recurrence is expected in one third of patients with incomplete removal.[7] Some other studies recommend long term follow up for IVL because of its possible recurrence.[13]

References

1. Nam MS, Jeon MJ, Kim YT, Kim JW, Park KH, Hong YS. Pelvic leiomyomatosis with intracaval and intracardiac extension: a case report and review of the literature. *Gynecol Oncol*. 2003; 89(1): 175 – 180.
2. Worley MJ Jr., Aelion A, Caputo TA, Kent KC, Salemi A, Krieger KH, et al. Intravenous leiomyomatosis with intracardiac extension: a single-institution experience. *Am J Obstet Gynecol*. 2009; 201(6): 574.e1 – 574.e5.
3. Gan HL, Zhang JQ, Bo P. The classification and surgical strategy of intracardiac leiomyomatosis. *Asian J Surg*. 2009; 32(3): 129 – 136.
4. Clay TD, Dimitriou J, McNally OM, Russell PA, Newcomb AE, Wilson AM. Intravenous leiomyomatosis with intracardiac extension - a review of diagnosis and management with an illustrative case. *Surg Oncol*. 2013; 22(3): e44 – e52.
5. Lai TK, Huang HY, Chan RY, Chin AC, Wong WC, Sit CY, et al. Magnetic resonance venogram of intravenous leiomyomatosis. *Hong Kong Med J*. 2005; 11(6): 524 – 526.
6. Sun C, Wang XM, Liu C, Xv ZD, Wang DP, Sun XL, et al. Intravenous leiomyomatosis: diagnosis and follow-up with multislice computed tomography. *Am J Surg*. 2010; 200(3): e41 – e43.
7. Li B, Chen X, Chu YD, Li RY, Li WD, Ni YM. Intracardiac leiomyomatosis: a comprehensive analysis of 194 cases. *Interact Cardiovasc Thorac Surg*. 2013; 17(1): 132 – 138.
8. Kahveci V, Ogur T, Cipe G, Ozdemir S, Hazinedaroglu S. Symplastic leiomyoma in the suprarenal inferior vena cava. Iran J Radiol. 2012; 10(1): 33 – 36.
9. Lou YF, Shi XP, Song ZZ. Intravenous leiomyomatosis of the uterus with extension to the right heart. *Cardiovasc Ultrasound*. 2011; 9: 25.
10. Sogabe M, Kawahito K, Aizawa K, Sato H, Misawa Y. Uterine Intravenous Leiomyomatosis with Right Ventricular Extension. *Ann Thorac Cardiovasc Surg*. 2014; 20(Suppl): 933 – 936.
11. Harris LM, Karakousis CP. Intravenous leiomyomatosis with cardiac extension: tumor thrombectomy through an abdominal approach. *J Vasc Surg*. 2000; 31(5): 1046 – 1051.
12. Liu B, Liu C, Guan H, Li Y, Song X, Shen K, et al. Intravenous leiomyomatosis with inferior vena cava and heart extension. *J Vasc Surg*. 2009; 50(4): 897 – 902.
13. Mariyappa N, Manikyam UK, Krishnamurthy D, Preeti K, Agarwal Y, Prakar U. Intravenous leiomyomatosis. *Niger J Surg*. 2012; 18(2): 105 – 106.