Management of Intravenous Leiomyomatosis of Uterus with Extension to Heart

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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

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 a diameter of 113 × 213 millimeters in her uterus.

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.\(^2\) 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.

Keywords: Cardiopulmonary bypass, leiomyomatosis, laparotomy

A transdiastolic 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 mass 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.

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

Clinical presentation of IVL varies in different patients. Approximately 13% of intracardiac leiomyoma patients are asymptomatic. Depending on the extension of tumor, cardiac insufficiency can occur in a previously healthy individual. Other common symptoms are syncope, dyspnea, fatigue, ascites, hepatomegaly or even chest pain. 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 tumor thrombosis are two main differential diagnoses of this disease. Distinguishing leiomyosarcoma is not possible by use of diagnostic tools prior to surgery. In middle aged women with uterine leiomyoma or history of myo-

Discussion

While the most frequent neoplasm in female genital tract is leiomyoma, IVL is a histopathologically benign and rare variant. Despite its benign histopathology, it is not essentially benign. Diagnosis of IVL requires higher levels of suspicion because of its rarity and nonspecific presentations. Clinical presentation of IVL varies in different patients. Approximately 13% of intracardiac leiomyoma patients are asymptomatic. Depending on the extension of tumor, cardiac insufficiency can occur in a previously healthy individual. Other common symptoms are syncope, dyspnea, fatigue, ascites, hepatomegaly or even chest pain. 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 tumor thrombosis are two main differential diagnoses of this disease. Distinguishing leiomyosarcoma is not possible by use of diagnostic tools prior to surgery. 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. As IVL does not usually invade blood vessels, it can be simply removed by downward traction from involved vein. In cases with intracardiac extension, performing a thoracoabdominal surgery with removal of tumor from heart chamber and inferior vena cava is preferred. 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. Intravenous and intracardiac leiomyomectomy can be the first stage, and the second stage could be pelvic resection and hysterectomy. It is not recommended to remove the entire tumor from a sternotomy and leave the caval attachment sites uncontrolled. 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. Since IVL is loosely attached to vessel walls, removal of intracardiac and intracaval lesions could be achieved easily. Complete removal of tumor can guarantee no recurrence and excellent outcome; however, recurrence is expected in one third of patients with incomplete removal. Some other studies recommend long term follow up for IVL because of its possible recurrence.

References