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Surgical Treatment of Intracardiac-Extending Intravenous Leiomyomatosis: A Single Center Experience

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ABSTRACT

Background: Few data were known on surgical management of intracardiac-extending in patients with intravenous leiomyomatosis (IVL).

Methods: From June 2007 to December 2014, six women (mean age, 39.3 ± 7.5 years; range, 24-55 years) with intracardiac-extending IVL were treated surgically at our hospital. Data were obtained from medical and pathological records, including characteristics of patients, surgical management, and follow-up.

Results: Surgery was performed successfully in all patients. Of 6 patients, 4 underwent one-stage operation and 2 underwent two-stage procedures. Circulatory arrest with hypothermia was used for a cardiotomy combined with venotomy in 5 patients. Complete resection was done in 5 patients. There were no perioperative deaths or complications in any of the patients. Hospital stay was 11.2 ± 2.9 days (range 7-15 days). All patients were followed-up for a mean of 41.0 ± 19.1 months (range, 17-69 months) after surgery. A recurrence of pelvic mass was found in 1 patient, but no symptoms or intravenous mass were reported. No obstruction occurred in any patient with a venotomy.

Conclusion: Surgery is a better therapy for IVL and complete removal has favorable outcomes.

INTRODUCTION

Intravenous leiomyomatosis (IVL) is a rare benign smooth muscle tumor that may extend along the venous system into the right heart cavity and pulmonary artery. Early diagnosis is difficult, because patients may be asymptomatic despite extensive intravenous extension. When symptoms are manifested, they are often secondary to direct cardiac involvement [Zhang 2010; Guo 2011; Yu 2011; Nam 2003]. To avoid complications of IVL, such as cardiac failure, pulmonary embolization, or sudden death, the appropriate therapy is surgical excision [Larzon 2006; Morice 2001]. However, surgical experience of this disease is limited. The purpose of this study is to summarize our experience of surgical management for IVL with intracardiac extension.

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MATERIALS AND METHODS

Patients

From June 2007 to December 2014, 6 women with IVL extending into the right heart were admitted to our institute. The characteristics of these patients are listed in Table 1. A history of uterine leiomyoma was observed in each case. The age ranged from 24 to 55 years (mean, 39.3 ± 7.5 years). The cardiac symptoms included dyspnea on exertion (n = 2, 33.3%), chest pain (n = 2, 33.3%), recurrent palpitation (n = 1, 16.7%), and syncope episodes (n = 1, 16.7%). Physical examination revealed grade 2-3/6 systolic murmur at the right sternal border in 3 cases and the edema of bilateral lower legs in 2 cases. Gynecological examination revealed the uterus to be enlarged to 14-week gestational size in 2 cases with pelvic mass. Tumor markers, such as CEA, CA-125, β-hCG, AFP, and LDH were all within normal limits. Preoperative imaging findings showed an intracardiac tumor arising from the iliac veins in 2 cases and inferior vena cava (IVC) in 4, and extending into the main pulmonary artery in 1. Imaging evaluation showed no existence of extensive attachment to cardiac and vascular structures. Prior subtotal hysterectomy was noted in 2 patients. The interval between the hysterectomy and the diagnosis of IVL was 2 and 4 years, respectively. Written informed consent was obtained from the patients and/or their family.

Surgical Procedure

All patients were placed in the supine position and general anesthesia was performed successfully. A median sternotomy and laparotomy were performed by the cardiac and vascular surgeons, respectively. Cardiopulmonary bypass (CPB) was established with ascending aortic arterial return and venous drainage through the right atrial appendage. The systemic temperature was reduced to 20-22°C, and total circulatory arrest was started in 5 patients. An atriotomy was created and the tumor was retracted from the right atrium/ventricle or outflow tract. During operation, we found no extensive attachment to cardiac structures, consistent with preoperative imaging evaluations. Limited attachment was addressed easily in the heart cavity. The tumor extending to the main pulmonary artery was also retracted from the right ventricular outflow tract to avoid a pulmonary artery incision. Intraoperative transesophageal echocardiography (TEE) showed no residual tumor in the

Table 1. Characteristics of Patients

Patient	Age, y	Sex	Clinical Manifestation	Diagnostic Method	Extent of Involvement	Prior Hysterectomy
1	55	F	Chest discomfort+palpitation	Venography +ECG	IVC to RA	No
2	45	F	Syncope+murmur+palpitation	ECG+CT	IVC to RV	No
3	37	F	Dyspnea+pelvic mass+murmur	ECG+MRI+CT	IV to PA	Yes
4	24	F	Syncope+murmur	ECG+MRI+CT	IVC to RV	No
5	33	F	Syncope+leg edema+pelvic mass	ECG+MRI+CT	IV to RA	No
6	42	F	Syncope+leg edema	ECG+CT	IVC to RV	Yes

ECG indicates echocardiography; CT, computed tomography; MRI, magnetic resonance imaging; IVC, inferior vena cava; RA, right atrium; RV, right ventricle; IV, iliac vein; PA, pulmonary artery.

Table 2. Operative and Follow-up Data of Patients

Patient	Surgical Approach	CPB	Bleeding, mL	Length of Tumor, cm	Maximal Diameter of Tumor, cm	Complete Resection	Hospital Stay, d	Follow-up, months	Recurrence
1	One-stage	Yes	4210	29	5	Yes	15	69	No
2	Two-stage	Yes	3240	32	10	Yes	13	58	No
3	One-stage	Yes	4290	36	8	Yes	9	42	No
4	Two-stage	Yes	3460	30	6	Yes	11	36	No
5	One-stage	Yes	2900	22	9	No	7	29	Yes
6	One-stage	No	3950	31	7	Yes	12	17	No

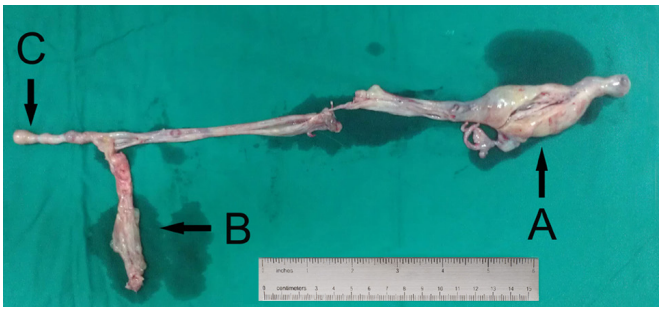
CPB indicates cardiopulmonary bypass.

heart cavity and the main pulmonary artery. And then visceral rotation and hepatic mobilization was conducted by vascular surgeons to obtain visualization of the IVC. A longitudinal venotomy was made and the tumor was removed as far caudally as possible. When IVC was exposed, we found that the left renal vein was completely obstructed in 2 patients (Figure). Vascular surgeons performed angiotomy and removed the tumor from the left renal vein. The remaining tumors were isolated by ligating the proximal internal iliac vein with silk suture. Gynecologists resected the pelvic mass together with the uterus and bilateral adnexa. A frozen section of the biopsy specimen taken intraoperatively was evaluated as benign neoplasm, consistent with a smooth muscle tumor. The patients were separated from CPB, and hemostasis was confirmed before ending the procedure. Without iliac venous involvement, surgical procedure in 2 patients was switched to a second-stage operation to enable a hysterectomy 6 months after first-stage operation. In addition, a laparotomy without CPB was conducted in 1 patient because the tumor was easily extracted from the right atrium through an IVC incision. Intraoperative TEE showed no residual tumor.

RESULTS

Surgery was performed successfully in all patients (Table 2). Of 6 patients, 5 underwent one-stage operation and 1 underwent two-stage operation. Circulatory arrest with hypothermia was used for a cardiotomy combined with venotomy in 5 patients. The mean amount of bleeding was 367 ± 507 mL (range, 2900-4210 mL). Postoperative echocardiography and computed tomography studies confirmed the absence of residual tumoral tissue. The removed tumor was smooth, white, and worm-like neoplasm. The mean length of tumor was 30.0 ± 4.6 cm (range, 22-36 cm) and mean diameter was 7.5 ± 1.9 cm (range, 5-10 cm). The postoperative course was uncomplicated. There were no perioperative deaths or complications in any of the patients. Two patients were moved to the cardiology department on the seventh day to have a vena caval filter inserted percutaneously through the femoral route and sited below the renal vein to prevent pulmonary embolization. Postoperative immunohistochemistry showed smooth muscle actin positivity and Ki67 positivity less than 5% in 2 patients. In the view of high detection of nuclear estradiol and progesterone receptors in the tumor, tamoxifen treatment was used in 2 patients to control the disease. Hospital stay was 11.2 ± 2.9 days (range, 7-15 days).

All patients were followed-up for a mean of 41.0 ± 19.1 months (range, 17-69 months) after surgery. A recurrence of pelvic mass was found in 1 patient at the 5-month follow-up after a two-stage surgery, but no symptoms or intravenous mass were reported, even though further treatment



Gross surgical specimen of the tumor after complete removal. The right enlarged part was in the heart (A). The distal tumor was removed from the left renal vein (B) and the right common iliac vein (C).

was rejected by the patient and her family. No obstruction occurred in patients with a venotomy.

DISCUSSION

IVL is a rare benign neoplasm, which is characterized by the easily identifiable growth of smooth muscle within the lumen of veins. Various surgical approaches have been used for the removal of the tumor [Mandelbaum 1974]. However, there are different opinions on the use of one-stage or two-stage procedures for patients with IVL. Previous studies suggested that staged surgery can reduce the one-stage operative time, and consequently can decrease the risk of bleeding associated with systemic heparinization required for CBP. In recent years, one-stage approaches have been introduced because of the increased knowledge of disease [Nam 2003; Larzon 2006; Gehr 1999; Schindler 2012]. On the basis of our experience with these cases, we recommend a one-stage approach with abdominal venous exposure at the time of removing the tumor through an atriotomy. One-stage operation avoids the risk for tumor embolism, tumor progression, or hemodynamic complications in the interval between the surgical stages. In our patients, 4 cases of one-stage surgery were successful, but the blood loss was relatively more. There were no severe complications or death. This suggested that one-stage operation is a feasible therapy for IVL with intracardiac extension. The two-stage procedure should be performed in patients with critical condition (e.g., severe heart failure) to reduce the risk of sudden death and concomitant venous thromboemblism. In 2 of our patients, a tumor from the IVC to the right heart was removed in the first stage, and limited uterine leiomyoma was resected in the second stage to reduce the amount of bleeding loss and operative time.

The routes and extent of IVL extension have a pronounced effect on the operative procedures available for these patients. Filsoufi et al reported avulsion injury of the renal vein resulting in a large hematoma during extraction of a right atrial tumor without abdominal exposure [Filsoufi 2002]. As a result, traction injury of the abdominal vein necessitated an emergency laparotomy, which increased the risk for operative mortality. In our opinion, although the extraction of IVL can reduce bleeding and operative time, extraction through the right atrium is not indicated for the IVL arising from the pelvic veins (e.g., ovarian vein) because of the existence of extensive attachment to the vessels at the junction of pelvic veins into the iliac vein or the renal vein. Clinical findings suggest that warranted extraction of IVL include tumors with a small diameter, nonsignificant obstruction of blood flow in the IVC, and no markers of right heart failure. Moreover, the staged operation is commonly recommended for tumors adherent to the cardiac and vascular structures, or for a prodigiously long tumor. Thus, a cardiotomy with laparotomy is affected more by the degree of adherence of the tumor than its length.

Preoperative evaluations are helpful for predicting whether IVL is adherent to the cardiac and vascular structures. In our patients, IVL can be extracted easily and safely because of limited attachment to the cardiac and vascular structures on imaging studies. Therefore, successful surgical treatment needs an accurate preoperative assessment.

In conclusion, surgery is a better therapy for IVL and complete removal has favorable outcomes. Accurate preoperative assessment of the tumor size and different routes of extension is essential to enable successful complete tumor excision.

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