Bilateral Salpingo-Oophorectomy for Intracardiac Leiomyomatosis: A Case Report

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Patient: Female, 41-year-old
Final Diagnosis: Intracardiac leiomyomatosis
Symptoms: Abdominal discomfort • menometrorrhagia
Medication: —
Clinical Procedure: —
Specialty: Obstetrics and Gynecology

Objective: Rare disease
Background: Intracardiac leiomyomatosis (ICLM) is an extremely rare tumor which is benign but presents with aggressive behavior. To date, there is still no standard of care for ICLM therapy, and treatment for complicated ICLM has obtained even less attention. Radical surgery was usually recommended to remove the patients’ tumors completely. Since initial complete surgical resection cannot be performed in all cases, bilateral salpingo-oophorectomy (BSO), via its effects of estrogen deprivation, may be a feasible primary step in the treatment of premenopausal women with unresectable ICLM.

Case Report: We describe a case of a residual mass in the inferior vena cava and right atrium that shrank dramatically after BSO. The patient was a 41-year-old woman with initially unresectable ICLM. Total hysterectomy with BSO and excision of the retroperitoneal mass was performed, but the intracaval tumor above L5 was not removed. Pathology revealed a benign leiomyoma which was strongly positive for both estrogen receptor and progesterone receptor. Two weeks after the BSO, the patient’s serum estradiol level had decreased to a postmenopausal level. At the same time, the proximal end of the intracaval tumor shrank dramatically from the level of the right atrium to the level of L3 only 2 weeks after the surgery. Therefore, this may provide a therapeutic window for a second reduction surgery.

Conclusions: BSO, via its estrogen deprivation effect, may provide a simple but effective initial treatment choice for premenopausal women who suffer from primary unresectable ICLM.

Keywords: Leiomyomatosis • Premenopause • Salpingo-oophorectomy

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Background

Intracardiac leiomyomatosis (ICLM) is an extremely rare tumor. It is a special condition of intravenous leiomyomatosis (IVLM) spreading beyond the patient’s inferior vena cava (IVC) and reaching the right atrium [1]. Even though ICLM is thought to be histologically benign, it is clinically aggressive and sometimes life-threatening [1-4]. To date, there are no standard treatment guidelines for patients with ICLM. Furthermore, the current treatment for those women with a complicated condition has received even less attention. Complete tumor resection is the most efficient treatment for patients with ICLM [5-8], but it cannot be achieved in many patients presenting with firm/extensive adhesion or pulmonary involvement [9]. Unsurprisingly, incomplete tumor resection increases the risk of tumor reappearance in one-third of cases [10]. In some cases, a normal extraction of the intravascular mass could result in IVC laceration, causing severe bleeding and even death during surgery [11]. Since initial complete surgical resection cannot be performed in all cases, BSO, via its effect of estrogen deprivation, may be a feasible primary step in the treatment of premenopausal women with unresectable ICLM. In addition, it could also simplify a second-step complete surgical resection.

We describe a clinical case of a premenopausal woman with initially unresectable ICLM. Our findings demonstrate that with a bilateral salpingo-oophorectomy (BSO), an incomplete surgical resection, the residual mass in the IVC and right atrium shrank dramatically. This shrinkage can serve to provide a therapeutic time window to perform a second-step complete surgical resection.

Case Report

A 41-year-old (gravida 4 para 2022) woman presented to her primary physician with a chief complaint of abdominal discomfort and menometrorrhagia approximately 2 months before the diagnosis of multiple uterine leiomyomas. Two myomectomies were performed, 5 and 16 months after the diagnosis, before she was referred to the gynecological oncology department of our hospital in April 2016. The reasons for referral were her persistent symptoms and recurrent uterine fibroids. The patient’s present symptoms were chronic lower abdominal pain and menometrorrhagia, but without anemia or cardiopulmonary symptoms. She had no other relevant medical history. On physical examination, she was noted to have a faint diastolic heart murmur and a mobile and slightly enlarged uterus, with a nodular pelvic mass on its right side. Contrast-enhanced computed tomography (CECT) showed an intravenous tumor arising from a right-side retroperitoneal pelvic mass (6.3×6.1 cm), spreading from the right iliac vein and common iliac vein to the IVC and left hepatic vein, finally extending to the right atrium at the T8 level. A 3.3×1.7×1.6-cm mass that occupied approximately half of the atrial cavity was detected. Through the tricuspid valve, the tumor intruded into the right ventricle during diastole (Figure 1A).

The findings of the laparotomy were in line with the preoperative imaging. A retroperitoneal ball-shaped mass extended from the slightly enlarged uterus into the right broad ligament. The lumina of the dilated right uterine vein and iliac veins were filled with worm-like tumors that extended through the IVC to the right atrium. Total hysterectomy with BSO and excision of the retroperitoneal mass were performed. After gently detaching the mass from the internal iliac vein by venotomy, the remaining proximal tumor in the cavity of the common iliac vein, the IVC, and the right atrium could not be successfully pulled out. Because of the high risk of fatal bleeding, complete removal of the mass was not recommended by cardiovascular surgeons at this stage. Eventually, the right internal iliac vein was ligated and the distal end of the tumor was located at the level of L5.

Pathology revealed benign uterine leiomyoma and intravenous leiomyoma. On immunohistochemistry, the tumor cells were strongly positive for both estrogen receptor (ER) and progesterone receptor (PR) (Figure 1F, 1G). On postoperative evaluation, 2 weeks after BSO, CECT showed that the residual tumor had decreased from the level of the right atrium to the level of L3 (Figure 1B). Furthermore, the tumor was now deemed to be resectable after consultation with a radiologist and cardiovascular surgeon. However, the patient and her next of kin refused secondary surgery due to their low socioeconomic status and fears of potential surgical complications. No evidence of pulmonary embolism was found perioperatively. She has had no symptoms, and no further medical treatment was given after hospital discharge. As shown in Figure 1D, 1E, the proximal end of the ICLM spread slowly to the level of T10 2 years after the surgery, but has remained stable and caused no symptoms so far (over 63 months after surgery). Simultaneously, her serum estradiol level decreased from 122 pg/mL preoperatively to less than 5 pg/mL 2 weeks postoperatively and has stayed at normal postmenopausal levels since then (Figure 2).

Discussion

ICLM is rare and usually affects premenopausal women who have a medical history of uterine leiomyoma. There are no established guidelines on the treatment of ICLM. Since it is believed to be estrogen-dependent [12,13], antiestrogen therapy may be useful in patients with ICLM, but the efficacy is controversial [14,15]. Barksdale et al believed that both aggressive surgical resection and removal of ovaries were necessary for the treatment of ICLM [16]. Unfortunately, complete tumor resection outside the primary step may thus be impracticable due to the disease’s rarity. Our findings demonstrated that a second-step complete surgical resection could also simplify a second-step complete surgical resection. In addition, it may be a feasible primary step in the treatment of premenopausal women with unresectable ICLM. In addition, it could also simplify a second-step complete surgical resection. BSO for ICLM...
Resection is impossible for some women, including patients with either a dense adherence to the venous wall or extensive involvement of the tumor [3,17]. Fatal outcomes related to aggressive surgery have been reported [11]. Therefore, it is critical to tailor a better treatment strategy for women in whom complete excision is difficult or impossible during the preoperative imaging or intraoperative evaluation stages.

In our case, the young woman with an initially unresectable ICLM was given a simple BSO-based surgery as a primary treatment option. The preoperative and postoperative imaging results are shown in Figure 1. The proximal end of the ICLM was located inside the right atrium at the level of T8. Two weeks after BSO, the leiomyoma had shrunk dramatically, and the proximal end was located inside the IVC at the level of L3. At 70 weeks after BSO, the leiomyoma had spread slowly, its proximal end being located inside the IVC at the level of L1. At 119 weeks after BSO, the proximal end of the leiomyoma was located inside the IVC at the level of T10. At 190 weeks after BSO, the proximal end was still located inside the IVC at the level of T10. The expression of estrogen (ER) and progesterone (PR) were tested by immunohistochemistry. Immunohistochemical staining showed that the nuclei of smooth muscle cells were 85% ER+ and 70% PR+.

Figure 1. (A-E) Coronal slices of CECT images of the ICLM, taken preoperatively and postoperatively. (A) Preoperatively, the proximal end of the ICLM was located inside the right atrium at the level of T8. (B) Two weeks after BSO, the leiomyoma had shrunk dramatically, and the proximal end was located inside the IVC at the level of L3. (C) At 70 weeks after BSO, the leiomyoma had spread slowly, its proximal end being located inside the IVC at the level of L1. (D) At 119 weeks after BSO, the proximal end of the leiomyoma was located inside the IVC at the level of T10. (E) At 190 weeks after BSO, the proximal end was still located inside the IVC at the level of T10. (F,G) The expression of estrogen (ER) and progesterone (PR) were tested by immunohistochemistry. Immunohistochemical staining showed that the nuclei of smooth muscle cells were (F) 85% ER+ and (G) 70% PR+. 1 – thoracic vertebrae; 2 – lumbar vertebrae. BSO – bilateral salpingo-oophorectomy; CECT – contrast-enhanced computed tomography; ICLM – intracardiac leiomyomatosis; IVC – inferior vena cava.
treatment. The secondary tumor resection was not performed despite being deemed feasible and less risky 2 weeks after BSO, when the proximal end of the ICLM was seen to have shrunk dramatically from the level of the right atrium to the level of L3. It was still located inside the IVC at the level of L1 70 weeks later (Figure 1B, 1C). The mechanism of significant decrease of tumor burden in our premenopausal case could be related to the estrogen deprivation effect caused by the BSO. Furthermore, as Maneyama et al reported, the tumor pedicle cutting may also have affected the blood supply to the residual tumor, leading to tumor shrinkage [18]. According to our experience with this case, approximately 2 weeks after BSO could be a suitable time to perform a radical resection of such a tumor. Therefore, it is believed that a simple BSO, as a primary treatment, provides a new and efficient treatment option for those premenopausal women suffering from complicated ICLM. This treatment strategy may also be beneficial for IVLM because this disease might in fact be the early stage of ICLM [7].

In addition, rather than undergoing a BSO, estrogen ablation, with gonadotrophin releasing hormone agonists, for example, could also be undertaken before a radical surgical resection [15]. Whereas followup without further surgery seems to have been satisfactory in our case, we do not recommend this as a standard approach because there is still a residual tumor, which has a risk of regrowth.

Conclusions

Via its estrogen deprivation effect, BSO may be considered a first-step treatment option for premenopausal women with primary unresectable ICLM. This surgical procedure may shrink and suppress tumor progression and may facilitate a second-step surgery for eventual surgical resection to lower or prevent persistent or recurrent disease.

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