Case Report

Surgical Management of Intracardiac Extension Leiomyoma: A Case Report and Review Literature

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A 49-year-old woman presented with complete heart block and recurrent syncope, which had caused a left fibula fracture. Transthoracic echocardiography showed a large right atrial mass. She received an emergency operation due to signs of tumor obstruction. A tumor was found originating from the inferior vena cava (IVC). Partial tumor removal was performed. A postoperative computed tomography scan revealed a uterine mass and tumor extension along the IVC. The pathological report confirmed a diagnosis of leiomyoma with intracardiac extension. She underwent residual IVC tumor removal under cardiopulmonary bypass using the deep hypothermic circulatory arrest technique. The patient was scheduled for a hysterectomy and bilateral salpingo-oophorectomy two months later and recovered uneventfully.

Keywords: Intravenous leiomyoma, Intracardiac extension leiomyoma, Cardiopulmonary bypass, Deep hypothermic circulatory arrest

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Intravenous leiomyoma (IVL) is a rare condition. Although leiomyoma is a benign smooth muscle cell tumor originating from the uterus, it can spread to intrauterine venules as biological nature of aggressive tumor. Standard treatment for IVL with intracardiac extension (ICE) is complete resection, which can be accomplished using either a one- or two-stage approach.

Since the first report of ICE leiomyoma in 1907 by Durck until now, nearly 200 cases of ICE leiomyoma have been reported. In the present case report, the authors presented a case of ICE leiomyoma with uncommon presenting symptom. A third degree atrioventricular (AV) block was treated successfully with operation under cardiopulmonary bypass (CPB) and deep hypothermic circulatory arrest (DHCA).

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Case Report

A 49-year-old woman presented to the emergency room (ER) with chest pain and recurrent syncope. Initial electrocardiogram (EKG) showed third-degree AV block. She was referred to our department after echocardiography revealed a large right atrial mass that obstructed the tricuspid valve (no revealed echo). The provisional diagnosis was right atrial mass, suspected of myxoma. Since the patient had signs and symptoms of tumor obstruction, she received an emergency surgery for tumor removal and found that the tumor originated from the inferior vena cava (IVC). The tumor had adhered firmly to the vessel wall, so a partial tumor removal was performed, and the residual part was left in the IVC. A postoperative computed tomography (CT) found the large lobulated heterogeneous enhancing lesion with some of central necrosis attached to anterior aspect of uterus, suspected of uterine sarcoma. It was also noted that patient had intraluminal filling defects in IVC, right common iliac vein, and right internal iliac vein (Figure 1).

Although the CT scan suggested for uterine sarcoma, the pathologic report of specimen has been confirmed for leiomyoma with myxoid change. The spindle cells stained positive with desmin,

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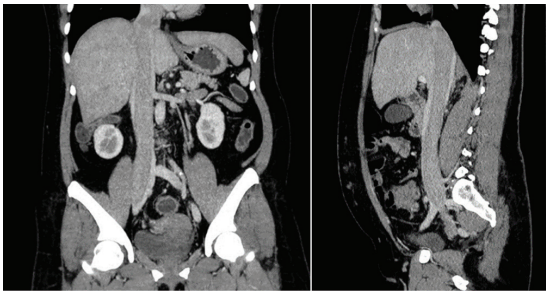

Figure 1. CT scan with contrast revealed a uterine mass, enhancing intraluminal filling defect in the right internal iliac vein, right common iliac vein, IVC, and proximal right hepatic vein.

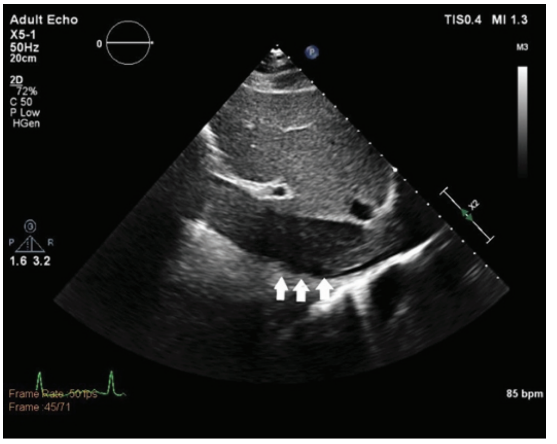

Figure 2. Preoperative echocardiography for the second operation revealed that tumor was along the IVC (arrow) and extended to the right atrium-IVC junction.

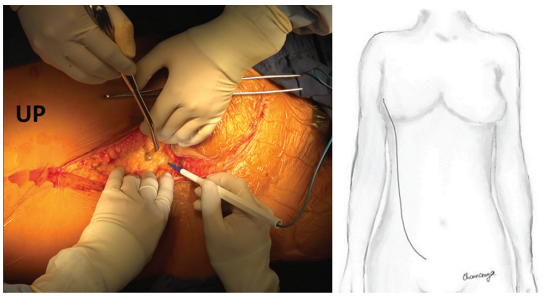

Figure 3. The photograph and illustration show the right thoracoabdominal incision. The incision started from the lower abdomen and extended to the right chest.

focal positive with caldesmon. The stage approach was decided upon after discussion among the multidisciplinary team. The plan was to remove the tumor in the IVC, followed by hysterectomy and bilateral salpingo-oophorectomy (Figure 2).

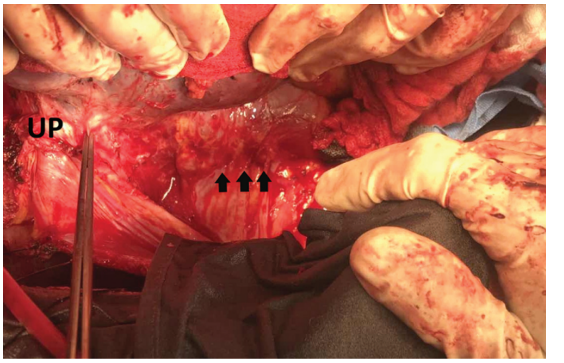

Figure 4. The right lobe of the liver was dissected and mobilized and the retro hepatic IVC was exposed as indicated by the arrow.

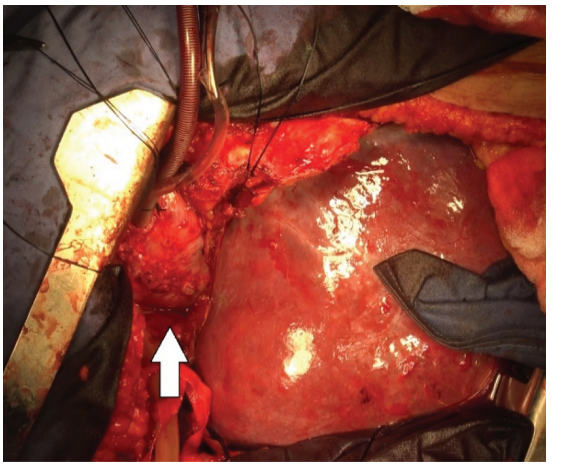

Figure 5. The incision was partially made at the diaphragm to approach the right atrium and supra-diaphragmatic IVC. An additional venous cannula was placed at the right atrium (arrow).

The operation was performed via right thoraco-abdominal incision, as shown in Figure 3. The incision extended to the right chest.

After entering the abdominal cavity, the entire infrahepatic IVC was approached. The triangular ligament was resected and the right lobe liver was mobilized. The retrohepatic IVC was dissected (Figure 4). The diaphragm was partially resected to approach the heart and supradiapragmatic IVC.

CPB was established via the left femoral artery and vein because the right common iliac vein was occupied with the tumor. Additional venous cannula was placed at the right atrium (Figure 5). The temperature was cooled down. When the temperature reached 20 degrees Celsius, circulatory arrest was initiated. A right atriotomy and tumor removal from

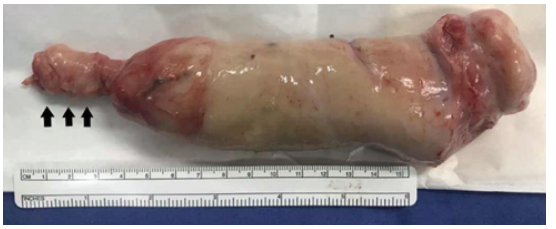

Figure 6. The tumor occupied the IVC and extended part in the right common iliac vein (arrow).

the heart and proximal IVC were performed. Skip venotomy was created from the retro hepatic IVC through bifurcation. The IVC tumor was totally removed, then the right internal iliac vein was ligated. The venotomy site was closed. CPB was resumed and the patient was rewarmed. Postoperatively, her EKG showed sinus rhythm. The patient was admitted to the intensive care unit for three days and discharged seven days after surgery with no adverse events.

The tumor was firm and rubbery with a smooth surface and had partially extended into the right common iliac vein (Figure 6). The tumor size was 22×5 cm. Microscopic imaging revealed uniform spindle cells with abundant cytoplasm. The pathologic report confirmed the diagnosis of leiomyoma. The patient was scheduled for an elective hysterectomy and bilateral salpingo-oophorectomy two months later. The patient recovered uneventfully and no evidence of tumor recurrence during three months follow-up.

Discussion

Leiomyoma is a common low-grade benign tumor of the uterus. However, it can spread to the venous system, known as IVL. IVL was first described by Hirschfield in 1896. In approximately 45% of cases, IVL can have ICE[9]. There are two theories that attempt to explain the mechanism by which these tumors spread. The first is that the tumor arises from the smooth muscle cells in the vessel wall. The other theory is that the uterine myoma, itself, invades the venous vessels. Since this condition is rare and the majority of patients are asymptomatic, early diagnosis is difficult[9,23].

The presenting symptoms of ICE patients can be varied, from no symptoms to sudden death[25]. The common symptoms are chest tightness, syncope, lower limb swelling, and abdominal discomfort[15]. Some patients may present with arrhythmia, most common is atrial fibrillation[7], followed by supraventricular tachycardia[15]. However, our patient presented with a complete heart block, an uncommon symptom that caused significant recurrent syncope.

The diagnosis of ICE leiomyoma requires a differential diagnosis with other right atrial masses, such as right atrial myxoma, right atrial thrombus, and metastatic tumors. Transthoracic echocardiography (TTE) is a good, non-invasive technique for diagnosis of ICE leiomyoma. An important point of distinction between ICE leiomyoma from right atrial myxoma, which is more common, is that the former tumor originates from the IVC. However, TTE may result in a limited view and may not be able to detect tumors originating from the IVC. Misdiagnosis can occur when TTE alone is used[6]. The fact that the first diagnosis was suspected for right atrial myxoma in the present study supports this finding.

CT scanning and magnetic resonance imaging (MRI) are useful in making a definitive diagnosis and allow us to examine the intrabdominal pathology. CT scans are commonly used to detect the primary tumor at the uterus, as well as the tumor extension. Another benefit of CT and MRI is that they are able to detect distant metastasis. Zhang et al[4] reported pulmonary metastasis in uterine leiomyoma, which was treated successfully using a one-stage approach. Otherwise, MRI is considered to be a great tool for investigating soft tissue lesions. In cases of IVL and ICE leiomyoma, MRI may help to differentiate tumors from thrombus[15].

The standard treatment for ICE is complete resection. Currently, there are one- and two-stage approaches for surgical management. There is no difference in terms of clinical outcomes between the two approaches. Multidisciplinary team involvement is needed to formulate a plan for treatment and surgery. The most challenging part of surgery is removing the tumor from the IVC and heart because there are risks of fatal hemorrhage and tumor emboli[17]. CPB with deep hypothermic circulatory arrest is the most commonly performed technique. Recently, there have been reports about new techniques to remove IVC and ICE tumors, such as the use of an endovascular technique to occlude the IVC and remove the tumor without CPB[20], tumor removal under beating heart[22], and an isolated total abdominal approach without sternotomy[21]. No matter which technique is used, the main goal is complete resection, as leiomyoma has a high recurrence rate (around 16% to 30%)[9]. Successful complete resection will allow for the best disease remission[10]. However, if complete resection is not possible, postoperative administration of gonadotropin-releasing hormone agonists and antiestrogen is recommended[10] to decrease the

recurrence rate.

What is already known on this topic?

IVL with cardiac extension is a rare condition that required to differentiate with other intracardiac mass. The multidisciplinary approach is necessary and complete surgical resection is the standard treatment.

What this study adds?

This case report presented with complete heart block, which is unusual. Misdiagnosis may occur; thus, ICE leiomyoma should be in the differential diagnosis in women presented with intracardiac mass found originating from the IVC. The multi-staged approach is amenable to perform, depending on a condition of the patient and the experience of the surgeons.

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Conflicts of interest

The authors declare no conflict of interest.

References

1. Rispoli P, Santovito D, Tallia C, Varetto G, Conforti M, Rinaldi M. A one-stage approach to the treatment of intravenous leiomyomatosis extending to the right heart. J Vasc Surg 2010;52:212-5.
2. Zhang G, Yu X, Shi H, Fan Q, Lang J, Liu B. Clinical characteristics and prognostic features of intravenous leiomyomatosis with inferior vena cava or intracardiac extension. J Vasc Surg Venous Lymphat Disord 2017; 5:485-92.
3. Guo X, Zhang C, Fang L, Guo L, Zhu W, Fang Q, et al. Echocardiographic characteristics of intravenous leiomyomatosis with intracardiac extension: a single-institution experience. Echocardiography 2011;28:934-40.
4. Zhang H, Miao Q, Liu J, Wang C, Zhang C, Zhai H. Giant intravenous leiomyomatosis with intracardiac extension. Ann Thorac Surg 2012;94:1013.
5. Maylar NM, Piotrowski AJ, Erbel R, Schmitz KJ. Images in cardiothoracic surgery. Giant right atrial mass: intracardiac extension of intravenous leiomyoma 18 years after hysterectomy. Ann Thorac Surg 2009; 88:684.
6. Price JD, Anagnostopoulos C, Benvenisty A, Kothuru RK, Balaram SK. Intracardiac extension of intravenous leiomyomatosis. Ann Thorac Surg 2017; 103:e145-e147.
7. Fogarty SJ, Hart G, Nicklin J, Venkatesh B, Boyne N, Wong DC, et al. Intracardiac leiomyomatosis - an unusual cause of syncope in a middle-aged woman. Heart Lung Circ 2017;26:e22-5.
8. Demirkiran F, Sal V, Kaya U, Alhan C, Tokgozoglu N. Intravenous leiomyoma with extension to the heart: a case report and review of the literature. Case Rep Obstet Gynecol 2013;2013:602407.
9. Du J, Zhao X, Guo D, Li H, Sun B. Intravenous leiomyomatosis of the uterus: a clinicopathologic study of 18 cases, with emphasis on early diagnosis and appropriate treatment strategies. Hum Pathol 2011;42:1240-6.
10. Low HY, Zhao Y, Huang KS, Shen HP, Wu PJ, Tseng CJ. Intravenous leiomyomatosis of the uterus: A clinicopathological analysis of nine cases and literature review. Taiwan J Obstet Gynecol 2017;56:362-5.
11. Harris LM, Karakousis CP. Intravenous leiomyomatosis with cardiac extension: tumor thrombectomy through an abdominal approach. J Vasc Surg 2000;31:1046-51.
12. Araki E, Koide Y, Fujimoto K, Okazaki K. Intravenous leiomyomatosis with extension into the right atrium: differential diagnosis from right atrial myxoma by intraoperative transesophageal echocardiography. Masui 2008;57:1008-12.
13. 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;50:897-902.
14. Zhang G, Yu X, Lang J. Intravenous leiomyomatosis with inferior vena cava or intracardiac extension and concurrent bilateral multiple pulmonary nodules: A report of 2 cases. Medicine (Baltimore) 2016;95:e4722.
15. 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:e44-52.
16. Li Y, Mei F, Yang C, Lv P, Ouyang C, Jin B. Intravenous leiomyomatosis with right heart involvement-a report of 4 cases and literature review. J Huazhong Univ Sci Technolog Med Sci 2011;31:586.
17. Marcus SG, Krauss T, Freedberg RS, Culliford AT, Weinreich DJ, Kronzon I. Pulmonary embolectomy for intravenous uterine leiomyomatosis. Am Heart J 1994;127:1642-5.
18. Zhang C, Liu X, Ma G, Zhang H, Wang C, Liu J, et al. Pulmonary embolization as the primary clinical manifestation of intravenous leiomyomatosis with intracardiac extension. Ann Thorac Surg 2012;94:1012.
19. Jiang WL, Zhang TH, Zhang YN, Hu SJ, Yamakawa T. Recurrent pelvic intravenous leiomyomatosis with extension up the inferior vena cava. EJVES Extra 2009;17:4-6.
20. Derubertis BG, Clair D, Faries P, Kapur S, Park K, Kent KC. Resection of an intravenous leiomyoma with intracardiac extension with use of endovascular techniques. J Vasc Surg 2004;40:554-8.
21. Matsuo K, Fleischman F, Ghattas CS, Gabrielyan AS, Ballard CA, Roman LD, et al. Successful extraction of cardiac-extending intravenous leiomyomatosis through

gonadal vein. Fertil Steril 2012;98:1341-5.
22. Atalay A, Poyrazoglu HH, Gocen U, Yuksel M, Eray IC, Guzel AB, et al. Successful one stage surgical removal of intravenous leiomyomatosis with on pump beating heart technique. Heart Lung Circ 2016;25:e72-4.
23. Valdés Devesa V, Conley CR, Stone WM, Collins JM, Magrina JF. Update on intravenous leiomyomatosis: report of five patients and literature review. Eur J Obstet Gynecol Reprod Biol 2013;171:209-13.
24. Murphy MA, Kothari A, Westaby S, Kehoe S, Handa A. Uterine leiomyoma with intravenous leiomyomatosis. EJVES Extra 2005;9:4-6.
25. Steinmetz OK, Bedard P, Prefontaine ME, Bourke M, Barber GG. Uterine tumor in the heart: Intravenous leiomyomatosis. Surgery 1996;119:226-9.