Intracardiac Extension of Intravenous Leiomyomatosis in a Patient with Vascular Pelvic Tumor and Prior Hysterectomy: A Case Report

Palanisamy Nithiyanandhan, Puthuvasserry R. Suneel, Aspari M. Azeez, Vivek V. Pillai, Shivanesan Pitchai
Department of Cardiothoracic and Vascular Anesthesia, Cardiovascular and Thoracic Surgery, Sree Chitra Thirunal Institute for Medical Sciences and Technology (SCTIMST), Trivandrum, Kerala, India

ABSTRACT
Uterine leiomyoma (UL) is the most common benign smooth muscle tumour of the premenopausal women. Rarely it shows malignant behaviour by metastasizing through the pelvic veins into systemic veins, inferior vena cava (IVC), there it is termed as intravenous leiomyomatosis (IVL). IVL may restrict itself within the IVC or it may extend into right heart chambers reaching up to pulmonary arteries. Here we report a case of single staged excision of intracardiac (IC) extension of IVL of a 45-year-old premenopausal women, who have undergone abdominal hysterectomy five years ago, with the complaints of shortness of breath aggravated on bending forward for the past two years.

Keywords: Intracardiac, intravenous leiomyomatosis, premenopause, uterine leiomyoma

INTRODUCTION
Intravenous leiomyomatosis (IVL) is a rare benign smooth muscle tumor taking its origin predominantly from the mesenchymal cells of the uterus. Uterine leiomyoma has preponderance for intravascular invasion with venules being the most commonly invaded structure, followed by lymphatics and arteries. Intracardiac (IC) extension of the IVL makes the diagnosis of this disease difficult as it could be misdiagnosed as thrombus or tumors arising in the heart chambers. Although complete surgical excision is the only available curative option, hormonal therapy has gained significance in recent years in view of the possibility of incomplete resection and for preventing recurrence. The surgery can be done either as a staged procedure or as a single-stage procedure depending on the extent of the disease and clinical condition of the patient.

CASE DESCRIPTION
The patient, a 45-year-old female, who had undergone hysterectomy 5 years ago, presented with complaints of shortness of breath for the past 2 years, which worsens on bending forward. On evaluation, her contrast-enhanced computed tomography (CECT) revealed a large mass of 9 × 8 cm with multiple dilated vessels posterior to the urinary bladder with arteriovenous fistulous communication and early draining dilated veins within the mass in the pelvis which are communicating with the dilated right common
Nithiyanandhan, et al.: Intracaval leiomyomatosis extending into right atrium

Ilial vein (CIV) and inferior vena cava (IVC). Tumor was seen extending from the dilated veins in the mass to the CIV, IVC, right atrium (RA), and right ventricle (RV). Transthoracic echocardiography (TTE) showed a mass in RA of about 7 x 3 cm moving in and out of RV through the tricuspid valve with good biventricular function. Other laboratory parameters were within the normal range. She was planned for a single staged tumor excision along with venacavotomy and IVC tumor removal.

Following the pre-anesthesia evaluation, the patient was premedicated with Tab Pantoprazole 40 mg and Tab Diazepam 5 mg the night before and on the day of surgery. In the operating room, after confirming the patient identity, all the standard monitors were attached. A 16G intravenous (IV) cannula and a 20G arterial catheter were secured in the right arm and the left radial artery respectively after local anesthetic infiltration. General anesthesia was induced with fentanyl, propofol, and pancuronium followed by endotracheal intubation with 7.5 mm cuffed endotracheal tube. The triple lumen central venous catheter and a 7 Fr venous sheath were placed in the right internal jugular vein (IJV) under ultrasound guidance. A temperature probe was inserted in the nasopharynx. Transesophageal echocardiography (TEE) probe was inserted into the esophagus. Anesthesia was maintained with sevoflurane, air-oxygen, and fentanyl.

TEE examination in the mid esophageal (ME) four-chamber view revealed an IC mass of about 8 x 3 cm protruding from the IVC into RA and RV, which was not attached to any part of the heart and moved freely with the cardiac cycle [Figure 1a, Video 1]. An atrial septal defect of about 8 mm was also noted in the ME modified bicaval view, which was not detected preoperatively [Figure 1b]. The IVC seen in the ME bicaval view revealed tumor mass within the lumen entering into RA but not adherent to the walls [Figure 1c]. The TEE was used to assess the hepatic and IVC blood flow and it was found out that there was good antegrade flow from both. Surgically, the abdominal team of surgeons dissected and took control of the suprarenal and infrarenal IVC, and thereafter the team of cardiothoracic surgeons did the sternotomy and pericardial separation.

After heparin administration and achievement of activated clotting time of above 480 seconds, ascending aorta (AA) and RA were cannulated for cardiopulmonary bypass (CPB). The superior venacava (SVC) was cannulated prior to CPB. Once CPB was established and adequate systemic perfusion was assured, cardioplegia was administered. The RA venous cannula was removed, the CPB was maintained with SVC-aortic cannulation and under TEE guidance the mass was removed from the RA and IVC. Once the tumor was removed from the visible portion of the IVC using TEE, the IVC was cannulated and the CPB was re-established using the bi caval-aortic cannulation. The ASD was closed, the heart was de-aired and the patient was weaned off CPB without difficulty.

Post CPB, the TEE examination confirmed that the IVC and the cardiac chambers were free of the tumor mass [Figure 2a]. TEE also confirmed that there was forward flow from IVC and hepatic veins without interruption and there was no damage to the tricuspid valve [Video 2]. Thereafter the heparin was reversed with protamine. Following this venacavotomy was done after looping suprarenal and infrarenal IVC with total removal of luminal mass from the IVC and iliac veins followed by pelvic tumor excision [Figure 2b]. Pelvic tumor dissection was associated with excessive blood loss on account of dilated arteriovenous malformation. Mean arterial pressure was maintained above 50 mm Hg by transfusion of crystalloids, blood products, and infusion of norepinephrine. After complete repair of IVC and complete tumor excision, the patient was shifted to the cardiac intensive care unit (ICU). The patient was extubated on the 1st postoperative day (POD) and sent to ward on the 2nd POD. Histopathology of the excised mass showed the tumor to be IVL [Figure 2c].

DISCUSSION

Uterine leiomyoma is the most common benign tumor in women. Although most of the time it confines its

![Figure 1: (a): Mid Esophageal (ME) 4-chamber view with the probe turned to right to focus more on the right atrium (RA) showing mass in the RA. (b): ME modified bicaval view showing atrial septal defect which was not diagnosed preoperatively. (c): ME bicaval view showing mass entering from inferior venacava into RA](image-url)
therefore there is no recommended technique of CPB or IVL.

Cardiac extension, uncommon sequelae, happens due to the smooth muscle proliferation extending cranially up the caval system to RA with the potential to enter RV and PA. Intracaval leiomyomatosis patients remain asymptomatic until the growth reaches RA. Echocardiography helps in the early diagnosis of IVL extending into cardiac chambers and also helps in the assessment of right heart structures and valves. Our patient had undergone hysterectomy for leiomyoma five years prior to the current presentation. She developed a vascular pelvic mass with arteriovenous malformations and dilated veins with the tumor mass extending into CIV, IVC, and RA.

The intraoperative TEE is a must and it helps in confirming the diagnosis, to know the extent of the spread of the tumor mass, and helps in guiding the surgeon for the effective removal of the tumor. It also helps in detecting tumor embolization even before hemodynamic instability sets in. Furthermore, it enables us to assess the IVC blood flow after tumor removal along with the knowledge of the intravascular volume status and ventricular function. In our case, additionally, it helped in the identification of an atrial septal defect (ASD) which was not detected preoperatively by transthoracic echocardiography. The presence of ASD has implications in a patient with IVC tumor because of the potential destination of a tumor embolus will include both the right side and left side of the circulation.

Intraoperative TEE assessment of IVC and hepatic veins showed adequate forward flow to RA despite the tumor mass. Therefore, we were confident of achieving adequate systemic perfusion on aorto-RA CPB. There is the risk of tumor embolization during RA cannulation and this should be prevented by careful handling of the cardiac chambers and the use of TEE guidance for the placement of the RA cannula. In our patient, we also cannulated the (SVC) prior to establishing CPB.

For a brief duration, the CPB was maintained on SVC-aortic cannulation, so that the RA cannula could be removed to facilitate tumor removal from the RA and IVC. The tumor removal from the IVC was closely monitored by TEE. The practice of clearing the tumor from the IVC before the institution of CPB to remove the IC portion was risky in our case as the tumor had a bulbous distal end which could break off and embolize if we attempted to pull the tumor out through the IVC. Since our patient also had an ASD, there was the risk of systemic embolization of the tumor and therefore we felt that the manipulation of the tumor had to be avoided prior to CPB and cardioplegia. The IVC was exposed and control obtained at the suprahepatic, suprarenal, and infrarenal levels prior to CPB. Once the tumor was removed from the heart and distal IVC, the proximal portion of the intravascular tumor was removed through an infrarenal transverse incision on the IVC.

Pringle maneuver, which is practised in abdominal surgeries by clamping hepato-duodenal ligament to reduce the bleeding, was not required in our case because the tumor was easily extracted from the IVC as anticipated from the echo imaging. Hepatic and portal veins were not looped or clamped at any time during the surgery as clamping these structures increases the chance of hepatic ischemia.

Deep hypothermic circulatory arrest (DHCA) has been practised for the removal of IVL. In such cases, the preferred method for achieving CPB is to go on SVC-left femoral vein cannulation. CPB with circulatory arrest on moderate hypothermia with ante-grade cerebral circulation is another option in these patients. CPB without circulatory arrest can be safely achieved as was done in our case. The use of DHCA is associated with the risk of bleeding and is potentially very risky in our patient as she required resection of the pelvic vascular mass which contained arteriovenous

margin within the pelvis, the involvement of the pelvic and systemic veins, described as IVL, occurs in 30% and IC extension of IVL occurs in 10%. First described by Birch-Hirschfeld in 1896, IVL is a rare benign tumor histologically characterized by the proliferation of smooth muscle cells within the venules. Durck first reported the IC extension of the IVL in 1907.

Intracaval leiomyomatosis patients remain asymptomatic until the growth reaches RA. Echocardiography helps in the early diagnosis of IVL extending into cardiac chambers and also helps in the assessment of right heart structures and valves. Our patient had undergone hysterectomy for leiomyoma five years prior to the current presentation. She developed a vascular pelvic mass with arteriovenous malformations and dilated veins with the tumor mass extending into CIV, IVC, and RA.

The intraoperative TEE is a must and it helps in confirming the diagnosis, to know the extent of the spread of the tumor mass, and helps in guiding the surgeon for the effective removal of the tumor. It also helps in detecting tumor embolization even before hemodynamic instability sets in. Furthermore, it enables us to assess the IVC blood flow after tumor removal along with the knowledge of the intravascular volume status and ventricular function. In our case, additionally, it helped in the identification of an atrial septal defect (ASD) which was not detected preoperatively by transthoracic echocardiography. The presence of ASD has implications in a patient with IVC tumor because of the potential destination of a tumor embolus will include both the right side and left side of the circulation.

IVL is a rare tumor and the presentation is varied and therefore there is no recommended technique of CPB or IVL.

Deep hypothermic circulatory arrest (DHCA) has been practised for the removal of IVL. In such cases, the preferred method for achieving CPB is to go on SVC-left femoral vein cannulation. CPB with circulatory arrest on moderate hypothermia with ante-grade cerebral circulation is another option in these patients. CPB without circulatory arrest can be safely achieved as was done in our case. The use of DHCA is associated with the risk of bleeding and is potentially very risky in our patient as she required resection of the pelvic vascular mass which contained arteriovenous

Figure 2: (a): ME 4-chamber view showing complete removal of tumor. (b): Complete tumor excised. (c): Histopathological confirmation of leiomyoma
malformations and dilated venules. The resection of this vascular pelvic mass after CPB was challenging on account of massive blood loss and could have been catastrophic if there had been any attendant coagulopathy on account of DHCA.

The clinical course of the patients with IVL varies depending upon the degree of intravascular obstruction ranging from pelvic pain, menorrhagia to chest pain, dyspnea on exertion, and palpitations.[12] The cardiac extension is associated with high morbidity and mortality due to caval compression and cardiovascular collapse. Unlike other lesions, tumor embolism is very uncommon due to the dense fibromuscular and vascular composition of the IVL.

Complete surgical removal being the only curative option available, the patient must be thoroughly investigated and a proper surgical plan should be made before taking the patient to the OR. IVL was removed using a two-stage approach in earlier days but advancement in surgical and anesthetic techniques have meant that a single-stage repair is now the norm. The literature reports a 33% recurrence rate of IVL with IVC extension in case of incomplete resection.[13] In case of incomplete resection or tumor reaching inaccessible sites, it is advisable to start hormonal therapy in the postoperative period to reduce the chances of recurrence.

CONCLUSION

Although IVL with IVC extension secondary to uterine leiomyoma is challenging, through multidisciplinary teamwork, proper planning, and usage of advanced imaging modalities like CECT and intraoperative TEE, it is possible to offer these patients a single-stage removal of the tumor.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Singh T, Lamont P, Otton G, Thomson DS. Intravenous leiomyomatosis with intracardiac extension: First reported case in Australia. Heart Lung Circ 2010;19:50-2.
2. Lou Y, Shi X, Song Z. Intravenous leiomyomatosis of the uterus with extension to the right heart. Cardiovasc Ultrasound 2011;9:25-9.
3. Ip PPC, Tse KY, Tam KF. Uterine smooth muscle tumours other than the ordinary leiomyomas and leiomyosarcomas: A review of selected variants with emphasis on recent advances and unusual morphology that may cause concern for malignancy. Adv Anat Pathol 2010;17:91-112.
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;22:44-52.
5. Choi IJ, Han MS, Cha MS. A case of huge intravenous leiomyomatosis with intracardiacextension. J Womens Med 2010;3:29-31.
6. Birch-Hirschfeld FV. Textbook of Pathological Anatomy. 5th ed. Leipzig: F.C.W. Vogel; 1896. p. 226-58.
7. Durek H. Ueber ien kontinierlich durch die entere holhlvene in das herz vorwachsendes: Fibromyom des uterus. Munch Med Wochenschr 1907;54:1154.
8. Gu X, He Y, Li Z, Chen J, Liu W, Zhang Y, et al. Intracardiac leiomyomatosis: Clinical findings and detailed echocardiographic features—a Chinese institutional experience. J Am Soc Echocardiogr 2014;27:1011-6.
9. Li R, Shen Y, Sun Y, Zhang G, Yang Y, Yang J, et al. Intravenous leiomyomatosis with intracardiac extension: Echocardiographic study and literature review. Tex Heart Inst J 2014;41:502-6.
10. Piardi T, Lhuaire M, Memeo R, Pessaux P, Kiannmanesh R, Sommacale D. Laparoscopic Pringle maneuver: How do we do it? Hepatobiliary Surg Nutr 2016;5:345-9.
11. Gaudino M, Spatuzza P, Snider F, Luciani N, Cina G, Possati G. Surgical management of a uterine leiomyoma extending through the inferior vena cava into the right heart. Heart Vessels 2002;17:80-2.
12. Zeng H, Xu Z, Zhang L, Luo YI, Chen H, Zhu H, et al. Intravenous leiomyomatosis with intracardiac extension depicted on computed tomography and magnetic resonance imaging scans: A report of two cases and a review of the literature. Oncol Lett 2016;11:4255-63.
13. Li B, Chen X, Chu Y-D, Li R-Y, Li W-D, Ni Y-M. Intracardiac leiomyomatosis: A comprehensive analysis of 194 cases. Interact Cardiovasc Thorac Surg 2013;17:132-8.