Can J Anesth/J Can Anesth (2014) 61:446–451
DOI 10.1007/s12630-014-0130-0

CASE REPORTS / CASE SERIES

Successful one-stage extraction of an intracardiac and intravenous leiomyoma through the right atrium under transesophageal ultrasound monitoring

Réussite d'une extraction en une étape d'un léiomyome intracardiaque et intraveineux par l'oreillette droite sous surveillance échographique transœsophagienne

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Received: 27 November 2013 / Accepted: 17 February 2014 / Published online: 1 March 2014

Abstract

Purpose *Intravenous leiomyomatosis is a rare disorder characterized by benign smooth-muscle tumours, termed leiomyomas, which originate from uterine leiomyomas or pelvic veins. Tumours may extend into the right-sided heart chambers, termed intracardiac leiomyomatosis (ICLM), and may be potentially life-threatening due to mechanical interference with cardiac structures or pulmonary arteries. While surgical excision is the optimal therapy, incomplete retrieval of a tumour or fatal retroperitoneal hemorrhage may occur. We present a case where intraoperative transesophageal ultrasound (TEU) guided complete removal of an intracardiac leiomyoma in a single-stage surgery solely through the right atrium without vein injury.*

Clinical features *A 46-yr-old female patient presented with a two-week history of exertional dyspnea, palpitations, and syncope. Preoperative imaging modalities revealed a continuous solid mass extending from the inferior vena cava (IVC) into the right atrium, and the patient subsequently underwent open heart surgery for tumour removal and definitive diagnosis. A systematic intraoperative TEU examination performed before resection showed that the serpentine tumour was free from any attachment to the IVC and the heart. Furthermore, the diameter of the intracardiac end of the tumour was wider than that of the IVC. Given these findings, the surgeons carefully drew the cord-like tumour out of the right atrium under close TEU monitoring without vein injury. Post-extraction TEU examination showed complete removal of the tumour. Microscopic examination of the specimen confirmed the diagnosis of intravenous leiomyomatosis.*

Conclusions *For cases with ICLM, intraoperative TEU plays a significant role in helping to plan the surgical approach, monitor the movement of the tumour and the IVC during the extraction, and assess the completeness of tumour resection.*

Résumé

Objectif *La léiomyomatose intraveineuse est une affection rare caractérisée par des tumeurs bénignes des muscles lisses (appelées léiomyomes) qui ont pour origine des léiomyomes utérins ou des veines pelviennes. Les tumeurs peuvent s'étendre dans les cavités droites du cœur (constituant la léiomyomatose intracardiaque) et menacer la survie des patients en raison de la gêne mécanique*

Author contributions *Hao-Rui Sun*, *Hai-Bo Song*, *Ke Lin,* and *Jin Liu* contributed to the conception and design of this manuscript. *Hao-Rui Sun* and *Yan-Zi Zhang* participated in data acquisition. All authors made significant contributions to manuscript preparation and final editing.

Electronic supplementary material The online version of this article (doi:10.1007/s12630-014-0130-0) contains supplementary material, which is available to authorized users.

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engendrée dans les structures cardiaques ou les artères pulmonaires. Bien que le traitement optimal soit l'exérèse chirurgicale, une exérèse incomplète de la tumeur ou une hémorragie rétropéritonéale fatale peuvent survenir. Nous présentons un cas dans lequel une échographie transœsophagienne (ETO) peropératoire a guidé l'extraction complète en un seul temps chirurgical d'un léiomyome intracardiaque, uniquement par voie auriculaire droite, sans lésion veineuse.

Caractéristiques cliniques *Une femme âgée de 46 ans a présenté pendant deux semaines une dyspnée d'effort, des palpitations et une syncope. Les modalités d'imagerie préopératoire ont révélé une masse pleine continue s'étendant de la veine cave inférieure (VCI) jusque dans l'oreillette droite et la patiente a subi en conséquence une intervention chirurgicale à cœur ouvert pour l'ablation de la tumeur et un diagnostic définitif. Un examen peropératoire systématique par ETO réalisé avant la résection a montré que la tumeur allongée et flottante était dépourvue de tout lien à la VCI et au cœur. De plus, le diamètre de son extrémité intracardiaque était supérieur à celui de la VCI. Compte tenu de ces constatations, le chirurgien a précautionneusement tiré sur la tumeur semblable à une corde hors de l'oreillette droite sous monitorage ETO continu sans léser la veine. L'examen par ETO post extraction a confirmé l'ablation complète de la tumeur. L'examen microscopique de l'échantillon a confirmé le diagnostic de léiomyomatose intraveineuse.*

Conclusions *Dans les cas de léimyomatose intracardiaque, l'ETO peropératoire apporte une contribution significative à la planification de l'abord chirurgical, à la surveillance des mouvements de la tumeur et de la VCI au cours de l'extraction, et permet de s'assurer que la tumeur a été complètement réséquée.*

Intravenous leiomyomatosis is a rare disorder characterized by benign smooth-muscle tumours which originate from uterine leiomyomas or the venous wall of the pelvic veins. The disorder is usually confined to the pelvic veins, but it may extend via the inferior vena cava (IVC) and reach the heart, termed intracardiac leiomyomatosis (ICLM).[1,2] Intracardiac leiomyomatosis can be life-threatening due to mechanical interference with right cardiac structures or pulmonary arteries.[3,4] Surgical excision is the optimal therapy, but incomplete retrieval of the tumour[5] or fatal retroperitoneal hemorrhage may occur.[6] Intraoperative transesophageal ultrasound (TEU) is a useful tool throughout surgical management of ICLM. The authors present a successful case in which TEU guided complete removal of an intracardiac leiomyoma from the right atrium (RA) in a single-stage surgery. Written informed consent was obtained from the patient for publication of this article.

Case report

A 46-yr-old female patient (gravida 2, para 1) presented to our hospital with a two-week history of exertional dyspnea, palpitations, and syncope. She had no significant medical history. On physical examination, there was no abnormality except for mild bilateral lower limb edema. The laboratory examinations revealed mild elevated liver enzymes (alanine aminotransferase 59 $U \cdot L^{-1}$, aspartate aminotransferase 40 $U \cdot L^{-1}$), a decreased albumin level of 36.1 $g \cdot L^{-1}$, a decreased platelet count of 95×10^9 L^{-1}, and an elevated carbohydrate antigen 125 level of 41.93 $U \cdot mL^{-1}$. Lower extremity venous ultrasound showed no thrombus. Transthoracic echocardiogram showed a serpentine solid mass extending from the IVC into the RA. For a further evaluation of the origin and extent of the mass, a contrast-enhanced abdominal computed tomography (CT) scan and enhanced cardiac magnetic resonance imaging (MRI) were performed. Results showed a continuous filling defect arising from an enlarged left common iliac vein and extending to the RA via the IVC. The intracardiac end of the mass was noticeably wider than the stalk and the IVC. Computed tomography images also showed mild hepatic congestion and multiple uterine nodules. The patient was initially treated with anticoagulants and diuretics and was subsequently scheduled for open heart surgery for mass removal and definitive diagnosis.

After monitors were applied according to American Society of Anesthesiologists standard guidelines, a left radial arterial catheter was placed. Anesthesia was induced with intravenous midazolam 0.05 $mg \cdot kg^{-1}$, sufentanil 1.5 $\mu g \cdot kg^{-1}$, and rocuronium 0.8 $mg \cdot kg^{-1}$ to facilitate tracheal intubation, and cannulation of the right internal jugular vein was then performed. Anesthesia was maintained by inhalation of 1.5-2.5% sevoflurane and infusions of propofol 50 $\mu g \cdot kg^{-1} \cdot min^{-1}$ and sufentanil 0.5 $\mu g \cdot kg^{-1} \cdot hr^{-1}$, with intermittent boluses of vecuronium and sufentanil as indicated. Transesophageal ultrasound was performed to examine the cardiac chambers, the IVC, and the potential course of the mass. In the mid-esophageal four-chamber view, an oval cardiac end of the mass measuring 3.8 cm at the greatest diameter floated in the RA with no intracardiac attachment. The cardiac end protruded slightly into the tricuspid annulus during diastole, and a slight tricuspid regurgitation was detected during systole (Fig. 1). The mid-esophageal right ventricle (RV) inflow-outflow view revealed no mass in the RV or pulmonary arteries (Fig. 1). There were no interseptal defects and all

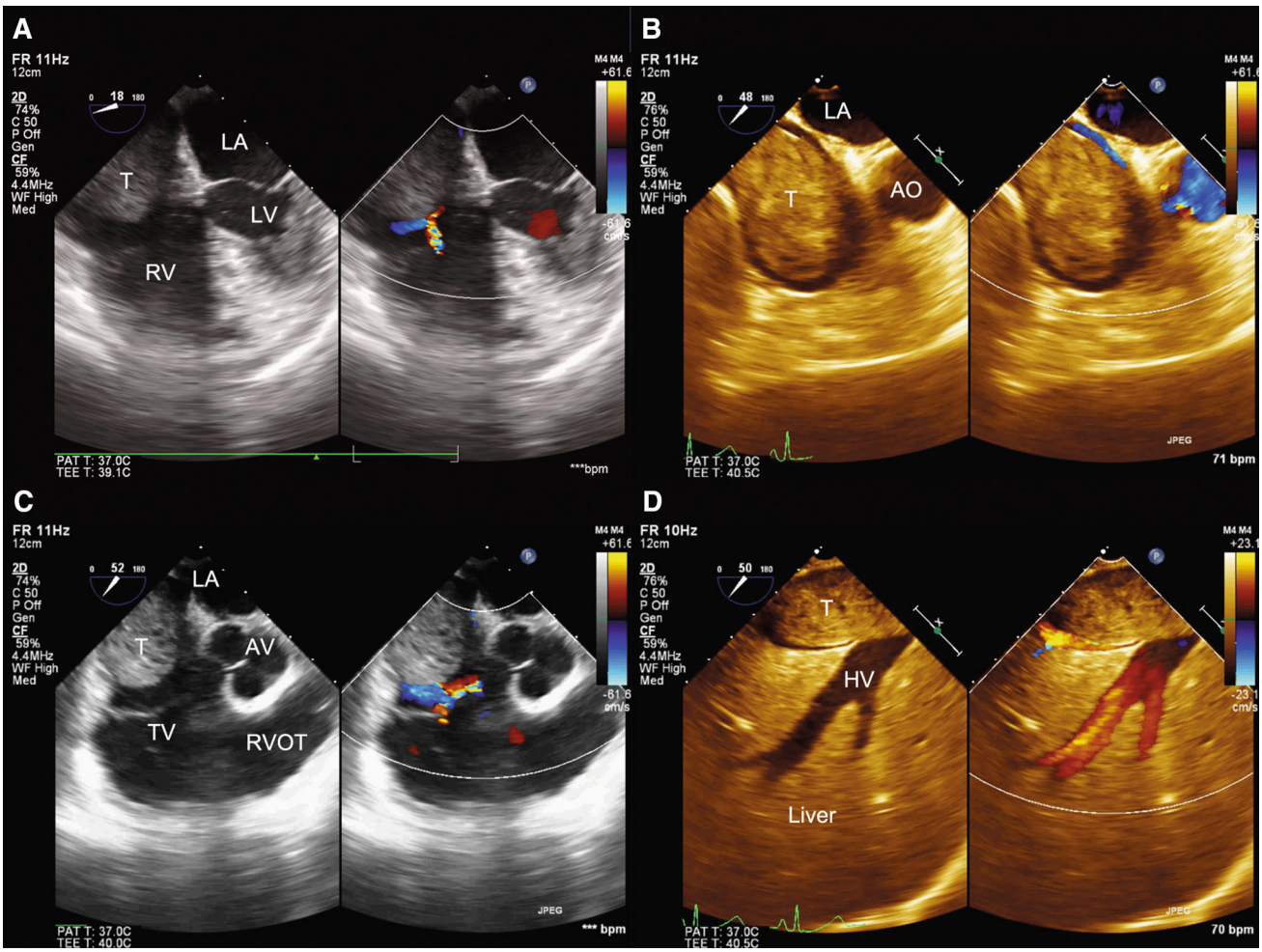

Fig. 1 Intraoperative transesophageal ultrasound examination before surgical manipulation of the tumour. A) A modified mid-esophageal four-chamber view showing an oval cardiac end of the tumour floating in the right atrium and a slight tricuspid regurgitation detected by color flow Doppler imaging during systole. B) A modified mid-esophageal right ventricular inflow view showing the cardiac end of the tumour with no intracardiac attachment. C) A mid-esophageal right ventricular inflow-outflow view showing no tumour in the right ventricle or the main pulmonary artery. D) A long-axis view of the intrahepatic inferior vena cava showing the tumour floating in the lumen without adhesion to the vessel wall or involvement of the hepatic veins. T = tumour; LA = left atrium; LV = left ventricle; RV = right ventricle; AO = aorta; AV = aortic valve; TV = tricuspid valve; RVOT = right ventricular outflow tract; HV = hepatic veins

heart valves were normal. To visualize the IVC, the mid-esophageal bicaval view was first obtained, and then, the ultrasound probe was advanced until the liver could be viewed. The probe was turned clockwise after that to display the junction of the IVC and RA. Rotating the multiplane angle 40-90° facilitated long-axis imaging of the distended IVC. The probe was then advanced progressively to examine the intrahepatic portion of the IVC. In this process, TEU showed a smooth-walled mass that nearly filled the lumen of the IVC without involvement of the hepatic veins and free from any attachment to the venous channel (Fig. 1) (Video 1, available as Electronic Supplementary Material). Nevertheless, it was difficult to obtain an optimal view of the IVC and intracaval mass below the right renal level. Moreover, the cardiac portion of the mass was confirmed to be the widest. Based on these findings, the cardiac surgeons made a decision to extract the mass through the RA with TEU monitoring.

The patient underwent median sternotomy. After heparinization (375 $U \cdot kg^{-1}$) and activated clotting time (ACT) over 480 sec, cardiopulmonary bypass (CPB) was set up with a venous cannula in the superior vena cava and an aortic cannula in the distal ascending aorta. The RA was then opened and a soft suction tube was inserted. After onset of CPB, a flow rate of 2.5 $L \cdot min^{-1} \cdot m^{-2}$ was achieved, and then, the nasopharyngeal temperature was slowly reduced to 32°C. The cardiac end of the tumour was found protruding into the RA from the IVC. The cardiac surgeons gripped the cardiac end of the tumour and attempted to pull it out slowly and gently via live

visualization by TEU (Fig. 2) (Video 2, available as Electronic Supplementary Material). By means of gentle pressure, a long rope of tumour was extracted from its point of attachment to the left common iliac vein without vein injury. The duration of tumour extraction was five to ten minutes. Repeat systematic TEU examination of the heart and the IVC confirmed complete removal of the tumour (Video 3, available as Electronic Supplementary Material). After the RA was closed, the patient was weaned from CPB. The total CPB time was 47 min, and ACT was maintained over 480 sec throughout perfusion.

The cord-like tumour was grey-white, smooth, and rubbery. It measured approximately 19 cm in length and 3.8 cm at the greatest diameter. The cardiac end was rounded, while the caval end had short blunt finger-like projections coated with a complete envelope (Fig. 3). Histological and immunohistochemical analyses of the specimen revealed bundles of benign spindle-shaped cells (Fig. 3) positive for desmin, smooth muscle actin, estrogen receptor, and progesterone receptor, which confirmed the diagnosis of intravenous leiomyomatosis. The patient had an uneventful postoperative course and was discharged on the 11th day. At follow-up two months later, abdominal and pelvic ultrasound revealed tumour-free IVC and bilateral iliac veins. Considering the uterine nodules, the patient went to a gynecologic hospital for further evaluation. The presence of adenomyosis with uterine myomas was suspected, and she was advised to undertake regular checkups. At nine-month follow-up, the patient remained well and asymptomatic.

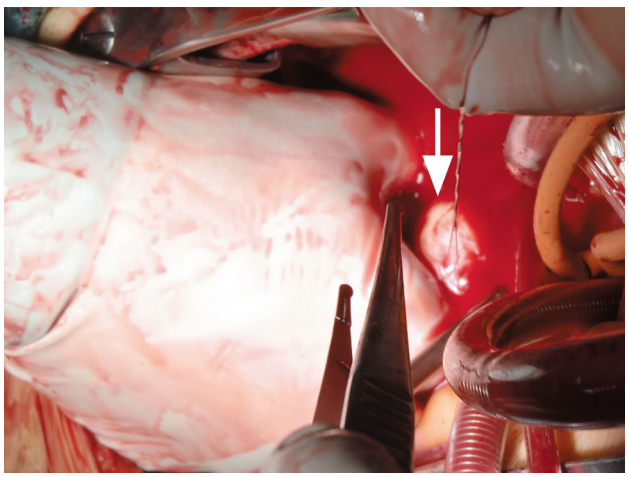

Fig. 2 Surgical extraction of the tumour. The surgeon grips the grey-white tumour at the cardiac end (arrow) in the right atrium and attempts to pull the tumour out

Discussion

Intravenous leiomyomatosis is a rare disorder characterized by benign tumours. It occurs primarily in middle-aged females with prior hysterectomy or coexisting uterine leiomyomas.[7] Its extension of central veins into the right heart chambers, termed intracardiac leiomyomatosis, occurs in more than 10% of cases.[8,9] In most cases, intracardiac leiomyomas grow freely within the venous channels without invading the tissues, but some may have multiple endovascular points of attachment along the route from the pelvis to the heart. Symptoms may be absent until the tumour interferes with cardiac function. The clinical manifestations of ICLM are various and nonspecific depending on the severity of the intracardiac involvement. The most common symptoms include shortness of breath, syncope, swelling in the lower extremities, and palpitations.[7] Sudden death may occur due to total obstruction of the right ventricular outflow tract or pulmonary embolism.

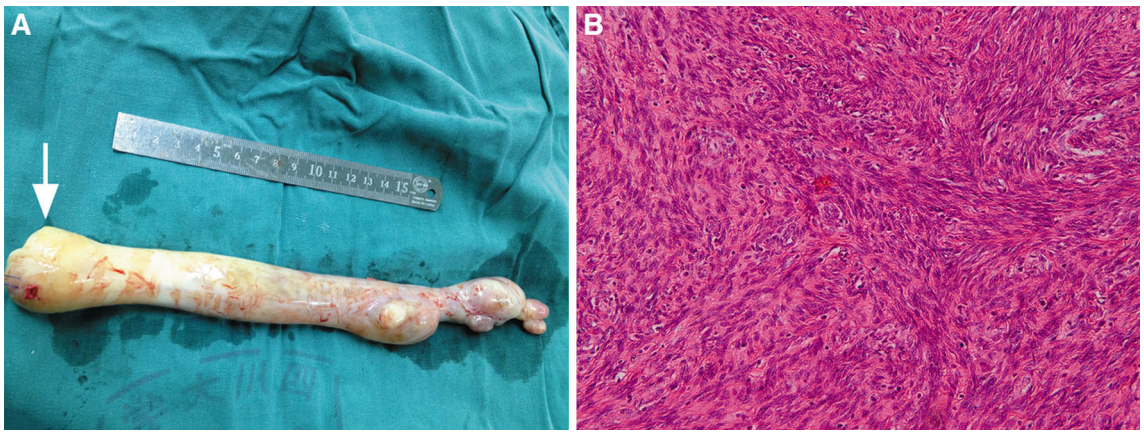

Fig. 3 Gross specimen and microscopic view of the tumour. A) The cord-shaped tumour measures 19 cm in length and 3.8 cm at the broadest point of the cardiac end (arrow), and the tumour is coated with a complete envelope. B) Histological study of the specimen (hematoxylin-eosin stain at 200 × magnification) showing bundles of benign spindle-shaped cells

Table The role of intraoperative transesophageal ultrasound in surgical extraction of an intracardiac leiomyoma

Phases	Phase focus
Pre-extraction	planning the surgical approach: tumour removal through laparotomy or sternolaparotomy*
During extraction	monitoring tumour and IVC movement, detection of embolism
Post-extraction	assessment of residue tumour†
Throughout surgery	monitoring hemodynamic status

*If the diameter of the intracardiac portion of the tumour is smaller than that of the IVC and the tumour has no adherence to the heart or IVC, tumour extraction from laparotomy only may be possible. On the other hand, if the diameter of the intracardiac portion of the tumour is larger than that of the IVC or if the tumour is attached to the heart and IVC, an abdominal and thoracic approach is safer

† Potential residual tumour below the right renal level of IVC could not be defined by intraoperative TEU

TEU = transesophageal ultrasound; IVC = inferior vena cava

Given the potential life-threatening risk, surgical removal for ICLM is the best treatment of choice. A multidisciplinary surgical strategy has previously been applied, including one-stage or two-stage abdominal and thoracic procedures.[10] Nevertheless, there have been reports of incomplete removal or fatal retroperitoneal hemorrhage due to vein injury. Wang *et al.* analyzed a cohort of 20 patients and found incomplete excision resulted in recurrence in one-third of patients.[10] Thus, total resection of the tumour in the safest manner is vital for successful treatment.

Intraoperative TEU has been reported to be significant in guiding surgeons throughout surgery for ICLM (Table). First, TEU plays a significant role in predicting and guiding the surgical approach before tumour extraction. Subramaniam *et al.* presented a case report of a patient with ICLM who underwent exploratory laparotomy with CPB on standby. During mobilization of the vena cava, TEU showed that the cardiac end of the tumour protruded down into the hepatic IVC by 5 cm. This phenomenon suggested that tumour extraction through the abdominal vena cava was feasible and spared the patient from a sternotomy and CPB.[11] Similarly, Matsuo *et al.* presented a novel case of a patient whose tumour was completely removed solely from the gonadal vein after intraoperative TEU showed a "pull-back" sign of the tumour.[12] Both cases showed that tumour extraction through abdominal veins might be possible if the tumour has a narrower diameter than that of the IVC and is free from any attachment to the IVC and heart. This would spare patients the risks associated with open heart surgery and CPB. In the present case, TEU confirmed that the intracardiac portion of the tumour had a wider diameter than the IVC, and the tumour had no attachment to the venous channels. Thus, the decision was made to pull the tumour out through the RA with close TEU monitoring. Second, during the process of tumour extraction, TEU can provide live visualization of the movement of the tumour and the IVC and detect tumour embolism. Third, TEU is critical for surveillance for residual tumour after resection so as to prevent possible recurrence due to incomplete excision. Little *et al.* presented a case report of a patient who underwent surgical extraction of a tumour through the IVC under CPB.[13] After initial resection of a grossly intact tumour, the authors performed a repeat TEU examination during the rewarming phase. Unexpectedly, they detected a thin residual tumour in the RV which led to complete resection of the mass.

The use of comprehensive preoperative imaging (including echocardiography, CT and MRI) is extremely important for successful management of ICLM[14,15] as is its correct interpretation and thorough consideration by the surgeons involved. Transesophageal ultrasound imaging is useful but limited, as clear visualization of the IVC becomes increasingly difficult below the right renal level, and the potential residual tumour below that level cannot be defined intraoperatively. Even so, this problem may be solved by simultaneous use of intraoperative abdominal ultrasound. In our patient, preoperative diagnosis of ICLM was uncertain, and simultaneous laparotomy was not planned. As the tumour extracted from the RA was grossly intact, in our view, complete resection was achieved, and this was confirmed by post-surgical abdominal and pelvic ultrasound. Nevertheless, it must be stressed that exploratory laparotomy could help guarantee total resection of the tumour, and withdrawing the tumour from the RA alone may increase the risk of hemorrhage due to avulsion of the vein at the point of the pelvic attachment.

In conclusion, intravenous leiomyomatosis is a disorder characterized by rare histologically benign tumours that may exhibit aggressive clinical behaviour. Extension of a tumour into the heart is potentially life-threatening and may require multidisciplinary surgical management. Thorough preoperative imaging and evaluation are of utmost importance for surgical planning. Transesophageal ultrasound can be a useful tool throughout the surgery for planning the surgical approach, monitoring the movement of the tumour and IVC during the procedure, and assessing the completeness of tumour resection.

Acknowledgments The authors declare that there was no assistance with the manuscript, no financial support, and no conflicts of interest.

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