

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case report

Point of care ultrasound facilitated diagnosis of right ventricular mass as the etiology of syncope; A case report of intravenous leiomyomatosis ☆☆☆★

Kristine L. Schultz, MD^{a,*}, Shawn M. Quinn, DO^a, Andrew H. Miller, DO^a, Rachel E. Fieman, MD^a, Mark D. Cipolle, MD, PhD^b, Timothy S. Misselbeck, MD^b, Kevin R. Roth, DO, FACOEP^a

^a Department of Emergency and Hospital Medicine, Lehigh Valley Health Network/University of South Florida Morsani College of Medicine, Lehigh Valley Campus, Cedar Crest Boulevard & I-78, Allentown PA 18103, USA

^b Department of Surgery, Lehigh Valley Health Network/University of South Florida Morsani College of Medicine, Lehigh Valley Campus, Cedar Crest Boulevard & I-78, Allentown PA 18103, USA

ARTICLE INFO

Article history:

Received 14 December 2020

Revised 25 February 2021

Accepted 25 February 2021

Keywords:

Intravenous leiomyomatosis

Syncope etiology

Point-of-care ultrasound

Intracardiac tumors

Syncope differential

ABSTRACT

Syncope is a common emergency department (ED) chief complaint. Rarely, syncope can be the result of right ventricular outflow obstruction from an intracardiac tumor, such as an intracardiac extension of intravenous leiomyomatosis (IVL). Typically, this type of tumor is confined to the pelvic veins, but in very rare cases, it can extend through the inferior vena cava into the right atrium. Point-of-care ultrasound (POCUS) can be a crucial tool in the ED for identifying intracardiac tumors presenting as syncope and expediting clinical management.

We present the case of a 39-year-old female with no prior medical history that presented to the ED having experienced dyspnea on exertion and two syncopal episodes prior to ED admission. POCUS use in the ED elucidated the presence of a right atrial mass and further imaging showed a mass on the patient's uterus. After surgical removal of a portion of the atrial mass, a subsequent biopsy revealed it had leiomyoma-like features; as such, the patient was diagnosed with IVL. This case illustrates the importance of using POCUS in the ED to help determine the etiology of syncope. Although intracardiac extensions of IVL are rare, it is important for emergency physicians to keep this diagnosis in the differential in patients with symptoms or risk factors suggestive of IVL with intracardiac extension.

© 2021 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

☆ Acknowledgments: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors. The authors would like to acknowledge Erin Shigo, BA for assisting for assistance in the writing and formatting of this report.

☆☆ Competing Interests: The authors declare that there is no conflict of interest regarding the publication of this article. The authors have no outside support information, conflicts, or financial interest to disclose, and this work has not been published elsewhere.

* Patient Consent: We confirm that written, informed consent for publication of this case was obtained from the patient.

* Corresponding author.

E-mail address: Kristine_L.Schultz@lvhn.org (K.L. Schultz).

<https://doi.org/10.1016/j.radcr.2021.02.062>

1930-0433/© 2021 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

Introduction

Syncope is a common emergency department (ED) chief complaint seen in 1%-1.5% of ED visits nationwide [1]. While intracardiac tumors and other obstructive and structural cardiac etiologies are more rare causes of the presentation of syncope, they remain an important part of the differential. Here we present a case of point-of-care ultrasound (POCUS) facilitating the diagnosis of obstructive syncope due to leiomyomatosis in the ED.

Intravenous leiomyomatosis (IVL) is a rare, smooth muscle tumor arising from uterine mesoderm that is histologically benign, but behaviorally malignant [2,3]. Typically, the tumor is confined to the pelvic veins, but in very rare cases, the tumor can extend through the inferior vena cava (IVC) into the right atria, ventricles, and pulmonary outflow tract [3–6]. Most patients with IVL do not exhibit any detectable symptoms until the tumor infiltrates the cardiac structures [7,8]. Once intracardiac, patients may experience syncope, pulmonary embolism, or heart failure [9,10]. Because intracardiac extension of this tumor is clinically very rare and the clinical presentation of the condition can be similar to heart failure, it is often misdiagnosed as other cardiac tumors or malignant thrombus [11]. Consequently, IVL is frequently treated incorrectly without appropriate imaging or management.

Due to the unpredictable biological behavior of this tumor, proper imaging techniques are crucial for early detection and guiding treatment. The present case discusses a 39-year-old woman that presented to the ED with dyspnea on exertion and several syncopal episodes. This case shows the importance of using POCUS in the ED to identify and diagnose obstructive syncope and to expedite the characterization and treatment of rare cardiac masses like IVL.

Case report

A 39-year-old female with no prior medical history presented to the ED having experienced dyspnea on exertion (DOE) for the past month and two syncopal episodes within the last 10 days. The patient stated that she had intermittent chest pain accompanying the DOE that radiated to her upper extremities and the right side of her back. In addition, the patient noted swelling in both her lower extremities for the past month. Upon physical examination, the patient was hypertensive with a blood pressure of 235/148 mmHg and an electrocardiogram revealed sinus tachycardia but was otherwise unremarkable.

A bedside echocardiogram (ECHO) was performed in the ED and revealed an abnormal soft tissue density in the right atria, ventricle, and IVC (Fig. 1). Based on the abnormal ECHO finding, a follow-up emergent computerized tomography pulmonary angiogram scan was ordered and showed a small subsegmental pulmonary embolism in the left lower lobe of the lung with a hypoattenuating area of approximately 4.2 cm involving the right atrium, suggesting an atrial thrombus (Fig. 2). In addition, the computerized tomography pulmonary angiogram revealed a relative enlargement of the

IVC concerning for an underlying IVC thrombus. Emergent cardiothoracic surgery and cardiology consultations were obtained based upon the findings. A duplex ultrasonography test was negative for deep vein thrombosis. A 2D Doppler ECHO showed a 2.4 × 2.0 cm echogenic mass and cardiac MRI confirmed that there was a very large thrombus originating in the IVC that was prolapsing across the tricuspid valve between the right atrium and ventricle (Fig. 3). Based on the aforementioned findings, the patient was taken urgently to the operating room to remove the right atrial mass due to the possibility of a fatal embolization.

A sternotomy with cardiopulmonary bypass was performed and revealed a large mass filling the entire right atrium and prolapsing into the right ventricle. The mass was approximately 8-9 cm long with multiple pedunculated arms and extended at least 4 cm into the intrahepatic IVC. It did not appear to be attached to an intracardiac chamber. During surgery, the intracardiac portion of the mass was excised, but the portion within the intrahepatic IVC was unable to be removed. A subsequent biopsy of the removed mass showed benign, smooth muscle fiber features consistent with a leiomyoma.

After the surgery, a renal ultrasound image was ordered due to the patient's worsening acute kidney injury and revealed a mid-abdominal mass measuring approximately 15.5 cm of uncertain etiology. A follow-up CT of the abdomen and pelvis (Fig. 4) showed a large cystic, solid mass emanating from the posterior uterine body and extending cranially to the lumbar 3-4 level. The mass measured 10.3 × 17.3 × 10.4 cm and extended into the right common iliac vein. Because the features of the excised right atrial mass were consistent with a leiomyoma and a large uterine mass was discovered, the patient was suspected to have IVL.

The patient was carefully monitored following surgery and ultimately discharged a week later. Approximately 3 weeks after discharge the patient underwent uterine artery embolization to reduce the size of the leiomyoma. At 2 months after the patient's initial presentation - the patient was identified as having concerned radiographic features of acute appendicitis on routine CT imaging for multidisciplinary surgical planning. Although the patient had no recent changes to her intermittent chronic abdominal pain, the appendix was dilated up to 9 mm in diameter and periappendiceal inflammatory changes were noted on CT. The patient was admitted to the hospital and was cared for by a large multidisciplinary team, which included physicians from: cardiothoracic surgery, vascular surgery, gynecologic oncology, general surgery, infectious disease, and hematology oncology. The patient underwent laparoscopic appendectomy, which was somewhat challenging due to the size of the uterus. While the pathology report revealed "appendicitis", it was the feeling of the general surgery team that the juxtaposition of the appendix to the recently embolized uterus, which caused inflammatory changes, was the reason for the appendiceal inflammation. After a short period of time to recover, the patient underwent a modified radical hysterectomy and bilateral oophorectomy salpingectomy to remove the remaining uterine tumor, and extraction of the remaining IVC tumor, which extended distally into the iliac veins. The patient met all postoperative milestones and was discharged on hospital day 15 in stable condition.

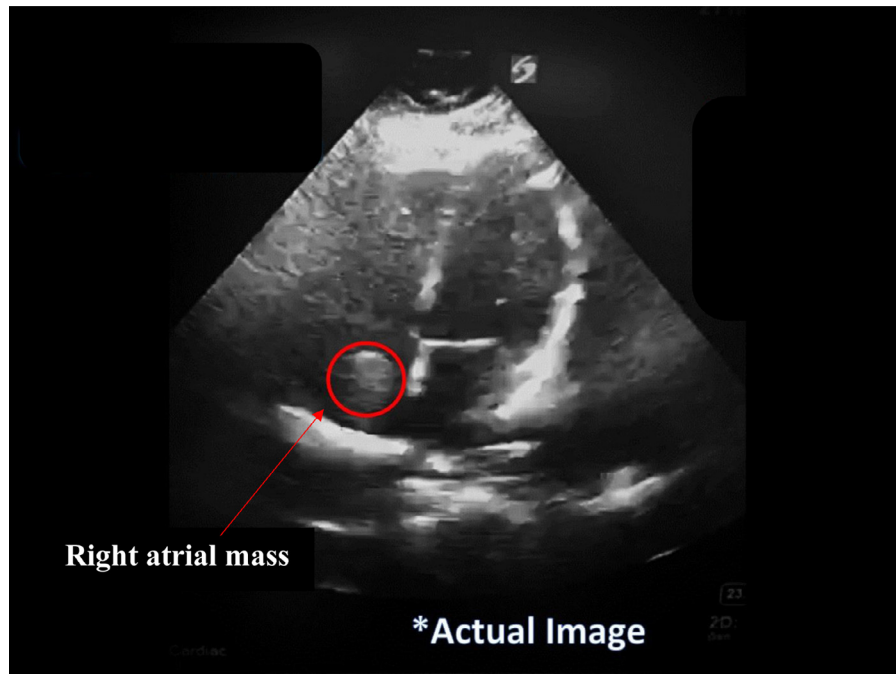


Fig. 1 – Point-of-care ultrasound showing right atrial mass as a result of an intracardiac extension of intravenous leiomyomatosis.

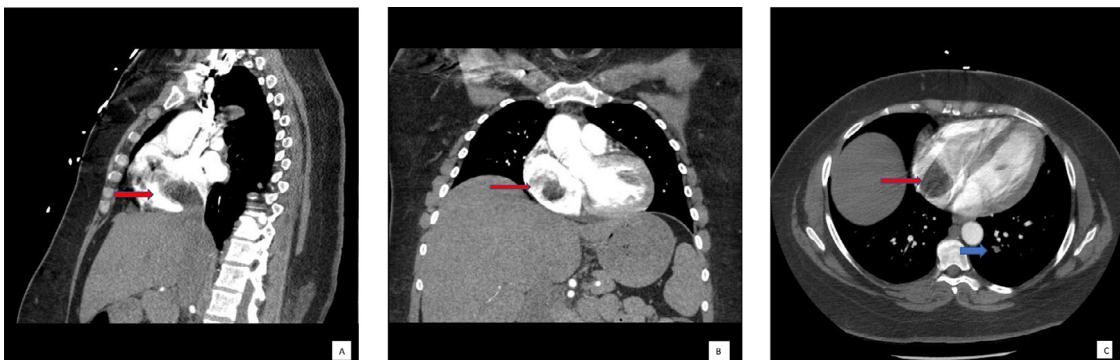


Fig. 2 – CT chest with intravenous contrast pulmonary embolism protocol. (A) Sagittal images. (B) Coronal images. (C) Axial images. Red arrows demonstrate the atrial filling defect/hypoattenuation, concerning for mass or thrombus. Blue arrow demonstrates pulmonary filling defect, subsegmental in the left lower lobe. (Color version of this figure is available online.)

Discussion

POCUS has become a widely used diagnostic tool in the ED, allowing clinicians to readily obtain ultrasound images to supplement and expedite treatment decisions [12]. Use of the tool has been associated with faster and more accurate clinical decisions in emergency settings [13]. The benefits of using POCUS have been demonstrated in a wide variety of clinical conditions, including cardiac arrest, chest pain, syncope, dyspnea, and abdominal pain [14]. In our case, the use of POCUS in the ED helped to narrow the differential diagnosis and ex-

pedite treatment for a patient with obstructive intracardiac syncope from extension of IVL.

IVL is a rare, histologically benign tumor that typically occurs in premenopausal, multiparous women around the age of 45 [15]. In most cases, patients have a history of hysterectomy due to uterine leiomyoma [4,5,16,17]. The tumor is thought to arise either as a proliferation of the vascular walls within the myometrium or pelvic veins, or be the result of an extensive invasion by a leiomyoma within the myometrium [5]. High levels of estrogen have also been thought to influence the development of IVL [18]. Most patients remain asymptomatic in the early stages of IVL and clinical signs and symptoms are related

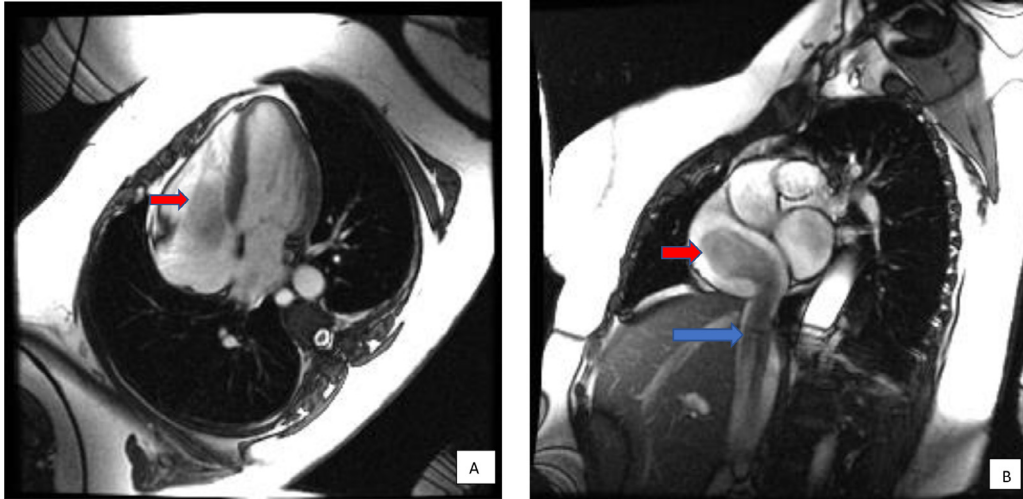


Fig. 3 – MRI heart without contrast. (A) Axial image. Red arrow demonstrates mass prolapsing into the right ventricle. (B) Sagittal image. Red arrow indicates thrombus. Blue arrow indicates extension to the inferior vena cava. (Color version of this figure is available online.)

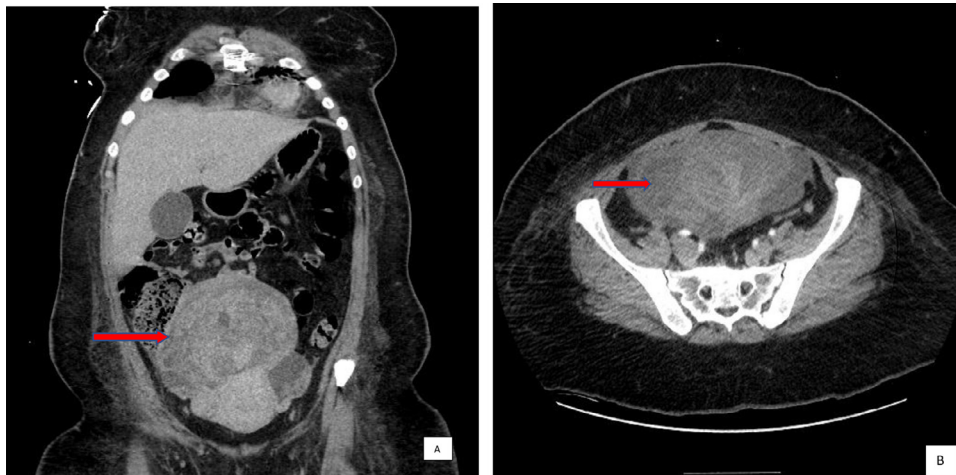


Fig. 4 – CT abdomen and pelvis with and without contrast. (A) Coronal image of the abdomen and pelvis. (B) Axial image of the pelvis. Red Arrows indicate a large, heterogeneous cystic and solid mass emanating from the posterior uterine body extending cranially to the L3-4 level. Size is approximately 10.3 x 17.3 x 10.4 cm. (Color version of this figure is available online.)

to size and localization of the tumor [9,15]. Tumor extension into the systemic veins and right heart chambers can lead to chest tightness, chest pain, swelling of the lower extremities, syncope, sudden death, and rarely heart failure [9,15,17].

Differential diagnosis of right atrial mass includes right atrial myxoma, thrombus, and metastatic tumors [26]. In order to prevent misdiagnosis, it is important to use a combination of imaging techniques to identify this pathology [19]. CT and MRI scans are commonly used to identify this pathology [19]. CT scans of a patient with IVL with intracardiac extension typically show an enlarged right atrium and expansion and thickening of affected veins [19]. When contrast is used, CT scans commonly

show a free filling defect in the right heart cavity that can extend into the pulmonary arteries, IVC, and uterine veins [19]. MRI scans can further confirm the diagnosis by revealing the scope of the tumor clearly [19].

However, while CT and MRI are considered to be the most sensitive imaging techniques in diagnosing IVL, bedside ultrasonography can play an important role in expediting the diagnosis of this pathology in the appropriate patients. Many studies have used transthoracic and transabdominal ultrasound as a supplemental imaging technique to identify intracardiac masses and uterus masses [2,5,11,17,20]. Depending on the degree of extension into the heart, ultrasound imag-

ing can elucidate a free-floating echogenic mass within the right atrium in addition to echoic masses within the internal iliac vein and IVC indicative of IVL with intracardiac extension [21]. Importantly, echocardiography can help distinguish between a right atrial myxoma, which is often included in the differential diagnosis, and IVL with intracardiac extension [8]. Right atrial myxoma is highly mobile and commonly attached to the interatrial septum, while IVL typically extends through the IVC to the right atrium [8,15]. If a right atrial mass is detected, it is important to image the IVC to help determine if the tumor is a result of IVL [8]. Overall, ultrasound provides clinicians with real-time images that can elucidate characteristic presentations of IVL and direct next steps in clinical management.

Total surgical excision of the extra-uterine tumor and myomectomy, or total hysterectomy, if necessary, is the recommended treatment for IVL to prevent recurrences [4,10,22]. Surgical excision is completed in either a single or two-stage procedure depending on the history of the patient and nature of the tumor [17,23,24]. Antiestrogen treatment, whether by medication or bilateral oophorectomy, has also been recommended as the tumor is considered to be estrogen dependent [3,25].

To our knowledge, this is one of the only reports demonstrating the importance of using POCUS in the ED to facilitate the diagnosis and treatment of IVL with intracardiac extension in a patient without a history of hysterectomy due to leiomyoma. In patients that present with syncope, it is important for emergency physicians to use POCUS to evaluate for obstructive and structural causes of syncope. POCUS is a readily available, portable, efficient, and low-cost imaging modality, which can give initial evidence as to the etiology of syncope. In addition, although intracardiac extensions of intravascular leiomyomatosis are clinically very rare, it is important for emergency physicians to be aware of this tumor and add it to their differential in patients with symptoms or risk factors suggestive of IVL with intracardiac extension.

REFERENCES

- [1] Huff JS, Decker WW, Quinn JV, Perron A, Napoli A, Peeters S, et al. Clinical policy: critical issues in the evaluation and management of adult patients presenting to the emergency department with syncope. *Ann Emerg Med* 2007;49(4):431–44. doi:10.1016/j.annemergmed.2007.02.001.
- [2] Price JD, Anagnostopoulos C, Benvenisty A, Kothuru RK, Balaram SK. Intracardiac extension of intravenous leiomyomatosis. *Ann Thorac Surg* 2017;103(2):e145–7. doi:10.1016/j.athoracsur.2016.07.037.
- [3] 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(4):897–902. doi:10.1016/j.jvs.2009.04.037.
- [4] Galajda Z, Copotioiu C, Suciuc H, Tint D, Glasz T, Deac R. The diagnosis, morphological particularities, and surgical technique in a case of intravascular leiomyoma extended to the right heart chambers. *J Vasc Surg* 2010;51(4):1000–2. doi:10.1016/j.jvs.2009.09.061.
- [5] Kutay V, Tuncer M, Harman M, Ekim H, Yakut C. Intracardiac extension of intravenous leiomyoma. *Texas Hear Inst J* 2005;32(2):232–4.
- [6] Ricci MA, Cloutier LM, Leavitt BJ, Mount S, Welander C. Intravenous leiomyomatosis with intracardiac extension. *Vascular* 1995;3(6):693–6. doi:10.1177/096721099500300625.
- [7] Caldenty G, Flores E, San Antonio R, Caixal G, Sánchez P. A rare cause of intracardiac mass. *Int J Cardiol* 2016;223(2016):91–2. doi:10.1016/j.ijcard.2016.08.140.
- [8] Fang BR, Ng YT, Yeh CH. Intravenous leiomyomatosis with extension to the heart: Echocardiographic features: a case report. *Angiology* 2007;58(3):376–9. doi:10.1177/0003319707302504.
- [9] Zhang L, Duan Y, Song F, Xiao J, Yang B. Intravenous leiomyomatosis with right atrium extension in two patients: a case report. *Mol Clin Oncol* 2016;5(5):604–6. doi:10.3892/mco.2016.1023.
- [10] Worley MJ, Aelion A, Caputo TA, Kent K, Salemi A, Krieger K, et al. Intravenous leiomyomatosis with intracardiac extension: a single-institution experience. *Am J Obstet Gynecol* 2009;201(6) pp. 574.e1574.e5. doi:10.1016/j.ajog.2009.06.037.
- [11] Firoj KM, Sasank S. Intravenous leiomyoma with extension to heart: a case report. *J Rare Disord Diagnosis Ther* 2016;2(3):1–3. doi:10.21767/2380-7245.100044.
- [12] Henneberry RJ, Hanson A, Healey A, Herbert G, Urbain I, Mensour M, et al. Use of point of care sonography by emergency physicians. *Can J Emerg Med* 2012;14(2):106–12. doi:10.2310/8000.CAEPES.
- [13] Bobbia X, Zieleskiewicz L, Pradeilles C, Hudson C, Muller L, Claret PG, et al. The clinical impact and prevalence of emergency point-of-care ultrasound: A prospective multicenter study. *Anaesth Crit Care Pain Med* 2017;36(6):383–9. doi:10.1016/j.accpm.2017.02.008.
- [14] Schaub A, Noble VE. Point of care ultrasound : an overview. *Am Coll Cardiol* 2017;1–14. Available from: <https://www.acc.org/latest-in-cardiology/articles/2017/10/31/09/57/point-of-care-ultrasound>.
- [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(3):44–52. doi:10.1016/j.suronc.2013.03.004.
- [16] Zhang C, Miao Q, Liu X, Zhang H, Ma G, Chen G, et al. Intravenous leiomyomatosis with intracardiac extension. *Ann Thorac Surg* 2010;89(5):1641–3. doi:10.1016/j.athoracsur.2009.09.044.
- [17] Morales MM, Anacleto A, Leal JC, Carvalho S, Del'Arco J. Intravascular leiomyoma with heart extension. *Clinics* 2012;67(1):83–7. doi:10.6061/clinics/2012(01)14.
- [18] Mitsuhashi A, Nagai Y, Sugita M, Nakajima N, Sekiya S. GnRH agonist for intravenous leiomyomatosis with cardiac extension: a case report. *J Reprod Med Obstet Gynecol* 1999;44(10):883–6.
- [19] Ouyang A, Su X, Li S, Li K, Wang J. Imaging manifestations of intravascular leiomyoma involving the right atrium: a case report and literature review. *Int J Clin Exp Med* 2015;8(7):11602–6.
- [20] Habibie YA, Hanafy D. Recurrence of intravenous leiomyomatosis: a rare cause of the obstruction of the right heart. 2018;1:221–228. doi:10.1108/978-1-78756-793-1-00004.
- [21] Ge Z, Wang Y, Qi Z, Zhang Q, Jin J, Li J. Ultrasound appearance of intravenous leiomyomatosis: A case report. *Medicine (Baltimore)* 2019;98(35):e16913. doi:10.1097/MD.00000000000016913.
- [22] Wang J, Yang J, Huang H, Li Y, Miao Q, Lu X, et al. Management of intravenous leiomyomatosis with intracaval and intracardiac extension. *Obstet Gynecol* 2012;120(6):1400–6. doi:10.1097/AOG.0b013e31826ebb90.
- [23] 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(1):212–15. doi:[10.1016/j.jvs.2010.02.018](https://doi.org/10.1016/j.jvs.2010.02.018).
- [24] Castelli P, Caronno R, Piffaretti G, Tozzi M. Intravenous uterine leiomyomatosis with right heart extension: Successful two-stage surgical removal. *Ann Vasc Surg* 2006;20(3):405–7. doi:[10.1007/s10016-006-9024-0](https://doi.org/10.1007/s10016-006-9024-0).
- [25] Lam PM, Lo KWK, Yu MY, Wong WS, Lau JY, Arifi AA, et al. Intravenous leiomyomatosis: two cases with different routes of tumor extension. *J Vasc Surg* 2004;39(2):465–9. doi:[10.1016/j.jvs.2003.08.012](https://doi.org/10.1016/j.jvs.2003.08.012).
- [26] Mankad R, Herrmann J. Cardiac tumors: echo assessment. *Echo Res Pract* 2016;3(4):R65–77. doi:[10.1530/erp-16-0035](https://doi.org/10.1530/erp-16-0035).