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Multidisciplinary Management of Uterine Intravenous Leiomyomatosis with Inferior Vena Cava and Right Atrial Extension: A Case Report

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Abstract

Background: Intravenous leiomyomatosis (IVL) is a rare benign smooth-muscle tumor characterized by intraluminal growth within venous channels, with potential extension into major veins and cardiac chambers. Despite its benign histology, IVL may demonstrate aggressive clinical behavior. Intracardiac extension, particularly into the right atrium, is extremely rare and presents significant diagnostic and surgical challenges.

Case Presentation: We report the case of a 53-year-old postmenopausal woman with uterine intravenous leiomyomatosis extending through the right gonadal vein, inferior vena cava, and into the right atrium.

Management and Outcome: The patient was successfully managed using a single-stage multidisciplinary surgical approach under cardiopulmonary bypass.

Conclusion: This case underscores the importance of early recognition, accurate imaging, and coordinated multidisciplinary surgical planning in achieving complete resection and favorable outcomes in IVL with intracardiac extension**.**

Keywords: Intravenous leiomyomatosis; Inferior vena cava; Right atrium; Cardiopulmonary bypass; Multidisciplinary surgery

Learning Points / Highlights

- Intravenous leiomyomatosis is a rare benign tumor with potentially aggressive intravascular and intracardiac extension.
- Cardiac involvement may be clinically silent and should be suspected in patients with venous symptoms or lower extremity edema.
- Multimodal imaging is essential for accurate preoperative assessment and surgical planning.
- Complete tumor excision, often requiring cardiopulmonary bypass and a multidisciplinary surgical team, is the cornerstone of management.
- Single-stage surgical resection can be safely performed and may reduce recurrence risk.

Keywords

Intravenous leiomyomatosis, Inferior vena cava, Right atrium, Cardiopulmonary bypass, Multidisciplinary surgery

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Introduction

While Intravenous Leiomyomatosis (IVL) is a rare condition, the total number of documented cases has increased significantly in recent years due to better imaging and diagnosis. Recent literature reviews indicate that approximately **750+ cases** of IVL have been reported worldwide.[1]

Uterine leiomyomas are the most common benign tumors of the female reproductive system. Intravenous leiomyomatosis represents a rare variant characterized by benign smooth-muscle proliferation within venous channels. Early-stage intravenous leiomyomatosis often presents with nonspecific clinical and imaging findings, resulting in frequent misdiagnosis or delayed detection. Although histologically benign, Progressive intravascular extension into the pelvic veins, inferior vena cava, right atrium, or pulmonary arteries may lead to severe complications, including ascites, dyspnea, heart failure, and sudden death.[1]

Two principal theories explain the pathogenesis of IVL. The first suggests origin from the smooth muscle of the venous wall, while the second—more widely accepted—proposes intravascular extension of a uterine leiomyoma through the myometrial venous plexus [2,3]. Sitzenfry first described this mechanism in 1911, postulating that a uterine leiomyoma gains access to venous channels and propagates intravascularly toward the heart [2].

Because of its rarity and nonspecific clinical presentation, IVL is frequently diagnosed late, often after cardiac involvement has occurred, making surgical management complex and high-risk [3,4].

Case Presentation

A 53-year-old postmenopausal woman presented with acute abdominal pain associated with one episode of vomiting. There was no prior history of cardiac symptoms such as dyspnea, syncope, chest pain, or palpitations.

Initial ultrasonography revealed multiple uterine fibroids and a right adnexal pedunculated mass. During routine preoperative evaluation, transthoracic echocardiography demonstrated a mobile echogenic mass extending from the inferior vena cava into the right atrium, raising suspicion of either thrombus or tumor extension [5].

Contrast-enhanced CT revealed an 11.5 × 6.1 cm inhomogeneously enhancing right adnexal mass with cystic areas and dilatation of the right gonadal vein. A continuous, non-occlusive intraluminal filling defect was noted along the entire length of the inferior vena cava, protruding into the right atrium, consistent with intravenous leiomyomatosis with intracardiac extension [6].

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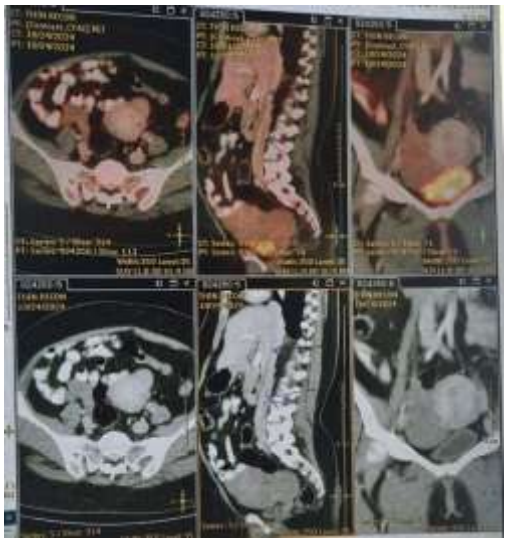
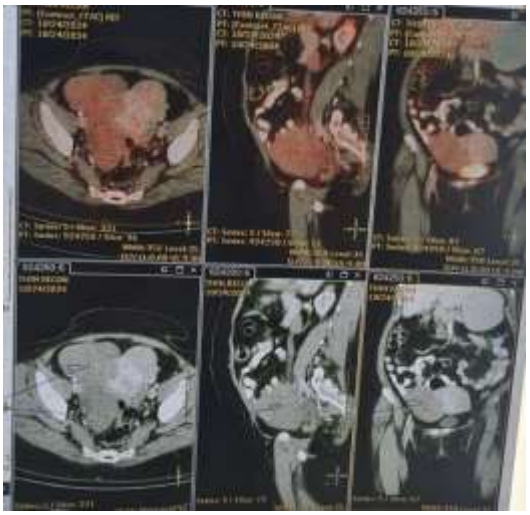


IVC THROMBUS



RIGHT ADNEXAL MASS

Given the imminent risk of embolization and sudden hemodynamic compromise, a multidisciplinary team involving gynecologic oncology, cardiothoracic surgery, anesthesia, and intensive care was assembled. After detailed counseling, the patient was taken up for urgent surgical intervention.

Surgical Management

A multidisciplinary team was assembled that included members from anesthesiology, cardiothoracic surgery (CTS) and surgical oncology department. The team discussed and debated several aspects of the perioperative care and after thorough deliberations and in light of the patient's stable cardiac status, decided to proceed with a single surgical approach for complete resection. Midline laparotomy was performed which revealed a variegated right adnexal mass with tortuous dilated vessels and a grossly dilated gonadal vein extending toward the inferior vena cava and iliac veins, total abdominal hysterectomy with bilateral salpingoopherctomy done. The tumor was transected at the level of gonadal vein to facilitate removal of the uterus after completion of the hysterectomy. Following completion of the hysterectomy, a median sternotomy was performed. Cardiopulmonary bypass was established with ascending aortic arterial cannulation and bicaval venous cannulation. A right atriotomy exposed the intracardiac component, which was divided at the inferior vena caval orifice. An inferior vena cava venotomy extending toward the iliac veins allowed complete en bloc extraction of the remaining intravascular tumor. The atriotomy and venotomy were closed in the standard fashion, and the patient was successfully weaned from cardiopulmonary bypass without difficulty. The reconstructed specimen demonstrated continuous extension from the uterus through the IVC into the right atrium and was sent for histopathological examination.

Histopathology

Histopathology confirmed benign smooth-muscle proliferation without cellular atypia or malignant features, consistent with intravenous leiomyomatosis [3,4].

Discussion

Intravenous leiomyomatosis, first described by Birch-Hirschfeld in 1896, is a rare benign smooth-muscle neoplasm characterized by intravascular growth. Despite its benign histology, it may exhibit clinically aggressive behavior because of progressive extension through the venous system [3,4,] Cardiac involvement typically occurs via extension through the gonadal veins and inferior vena cava into the right atrium and may mimic thrombus or cardiac tumors on imaging [5,7].Echocardiography remains a valuable initial tool for detecting intracardiac masses, while CT and MRI are crucial for defining tumor extent and planning surgical strategy [5,6]. Complete surgical excision is the definitive treatment, as incomplete resection is associated with recurrence [10].Single-stage surgical management under cardiopulmonary bypass has gained favor due to reduced embolic risk and improved completeness of excision [8,9,11]. Multidisciplinary coordination is critical to achieving optimal outcomes in such complex cases. Bilateral salpingo-oophorectomy is recommended, particularly in postmenopausal women, to reduce estrogen-driven recurrence, which has been reported even years after surgery [10].

Given the fact that estrogen and progesterone receptors are present in the leiomyoma cells, bilateral salpingoophorectomy is essential, and exogenous estrogens must be avoided to prevent subsequent growth of microscopic or unresected foci of intravenous leiomyomas. For then on-castration cases, post-surgery hormone therapy should also be considered.

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Gonadotropin-releasing hormone agonists or antiestrogen therapy have also been used to prevent tumor growth and to reduce the tumor mass but no efficacy has been demonstrated to date [12,13]

Conclusion

Intravenous leiomyomatosis with right atrial extension is a rare but potentially life-threatening condition. Early diagnosis, comprehensive imaging, and a single-stage multidisciplinary surgical approach under cardiopulmonary bypass enable safe and complete tumor removal with excellent outcomes.

References

1. Zhou X, Qi X, Zhao X, Yang F. Update on clinical characteristics and molecular insights for uterine intravenous leiomyomatosis. Oncol Lett. 2024;27(1):31.

2. Sitzenfrey A. Über Venenmyome des Uterus mit intravaskulärem Wachstum. Z Geburtshilfe Gynaekol. 1911;68:1-25.

3. Norris HJ, Parmley T. Mesenchymal tumors of the uterus. V. Intravenous leiomyomatosis. Cancer. 1975;36:2164-2178.

4. Clement PB. Intravenous leiomyomatosis of the uterus. Pathol Annu. 1988;23(Pt 2):153-183.

5. Price JD, Anagnostopoulos C, Benvenisty A, et al. Intravenous leiomyomatosis with intracardiac extension: echocardiographic features. J Am Soc Echocardiogr. 2000;13:455-459.

6. Wei J, Li B, Chen X, et al. Intravenous leiomyomatosis with intracardiac extension: CT and MRI findings. AJR Am J Roentgenol. 2011;197:W944-W951.

7. Lam PM, Lo KW, 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):465-469.

8. Castelli P, Caronno R, Piffaretti G, et al. One-stage surgical removal of intravenous uterine leiomyomatosis with cardiac extension. Ann Vasc Surg. 2006;20:278-282.

9. Li R, Shen Y, Sun Y, et al. Single-stage surgical management of intravenous leiomyomatosis with cardiac extension. Ann Thorac Surg. 2012;93:1953-1955.

10. Yu X, Zhang G, Lang J, et al. Factors associated with recurrence after surgical resection of intravenous leiomyomatosis. Obstet Gynecol. 2016;128:1018-1024.

11. Clay TD, Dimitriou J, McNally OM, et al. Intravenous leiomyomatosis with intracardiac extension: diagnosis and management. Ann Thorac Surg. 2013;95:e15-e17.

12. Mitsuhashi A, Nagai Y, Sugita M, Nakajima N, Sekiya S. GnRH agonist for intravenous leiomyomatosis with cardiac extension: a case report. J Reprod Med. 1999;44(10):883-886.

13. Efthimiadis C, Petousis S, Grigoriou M, Ioannidis A, Tzouveleki I, et al. Successful multiple-step management of intravenous leiomyomatosis diagnosed after an episode of acute abdominal pain: case 0report and review of the literature. Int J Surg Case Rep. 2015;14:176-180.