

# Case Report: First Reported Case of a Pelvic Myxoma Extending through the Inferior Vena Cava to the Right Atrium

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## Abstract

This is the first report of an extra-cardiac pelvic myxoma extending through the inferior vena cava to the right atrium, and coinciding with a concurrent large uterine leiomyoma. Complete excision was performed to prevent complications and future recurrence. This rare case highlights the importance of multidisciplinary management and careful surgical planning in these rare conditions.

## Keywords

Extra-Cardiac Pelvic Myxoma, Right Atrium, Surgery

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## 1. Introduction

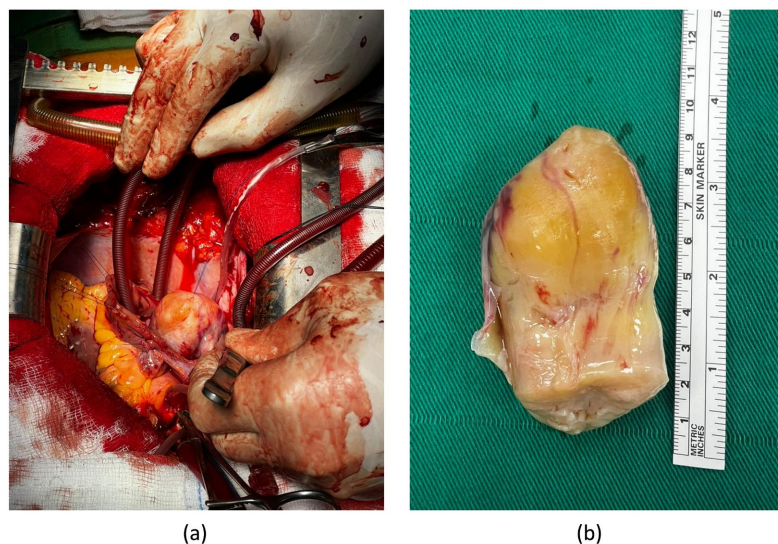
Atrial myxomas are the most common primary cardiac tumors, most frequently encountered in the left atrium [1]. Right atrial myxomas are less common, and those originating from the inferior vena cava (IVC) and extending into the right atrium are even more rare [2]. To our knowledge, an extra-cardiac myxoma arising from the pelvic veins and propagating cranially through the IVC has not been previously reported in the medical literature. In this report, we describe the case of a young woman who was found to have an extra-cardiac pelvic myxoma extending through the IVC and presenting as a right atrial mass.

## 2. Case Presentation

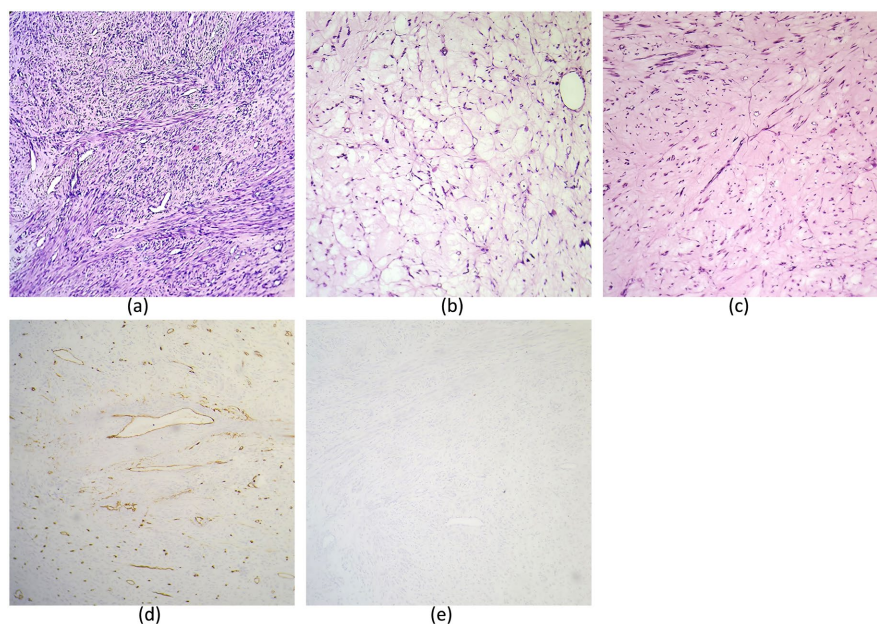
A 39-year-old woman presented with recent progressive dyspnea over a period of

one month, associated with recurrent vomiting for 10 days. In her past medical history, she had undergone surgical excision of a uterine fibroid 18 months earlier. On physical examination, she appeared pale, with mild generalized edema and dyspnea at rest. Laboratory tests showed moderate anemia (hemoglobin 9 g/dL), while other results were normal. Transthoracic echocardiography revealed a dilated right atrium containing a  $52 \times 40$  mm heterogeneous and highly mobile mass prolapsing through the tricuspid valve during diastole, and causing severe tricuspid valve regurgitation. No obvious stalk could be visualized.

Immediate surgery was considered indicated without further imaging as per recent recommendations [3] [4], and was performed through median sternotomy, aortic and bi-caval cannulation, full cardiopulmonary bypass perfusion, and cold blood cardioplegic arrest. An oblique right atriotomy uncovered a large yellow-colored and rubbery mass protruding from the IVC orifice and filling the right atrium, with no attachment to the right atrial wall (**Figure 1(a)**). Deep hypothermic circulatory arrest at  $16^{\circ}\text{C}$  was carried out in order to inspect the caudal extent of the mass, and this confirmed that there were no attachments between the mass and IVC wall, and that the mass extended well below the hepatic veins. Partial excision ( $85 \times 50$  mm) of the mass was performed as deep into the IVC as could be reached (**Figure 1(b)**). Rewarming and weaning off-cardiopulmonary bypass were uneventful. The patient was transferred to ICU in stable condition without inotropic support and was extubated routinely. Postoperative echocardiography confirmed preserved cardiac function (EF 62%) with no residual intracardiac masses. Histological and immunohistochemical study of the excised specimen confirmed the diagnosis of a myxoma composed of proliferation of bland spindle cells in myxoid background in intersecting fascicles, with tumor cells negative for SMA and Desmin, and focally positive for Calretinin immune staining (**Figure 2**).

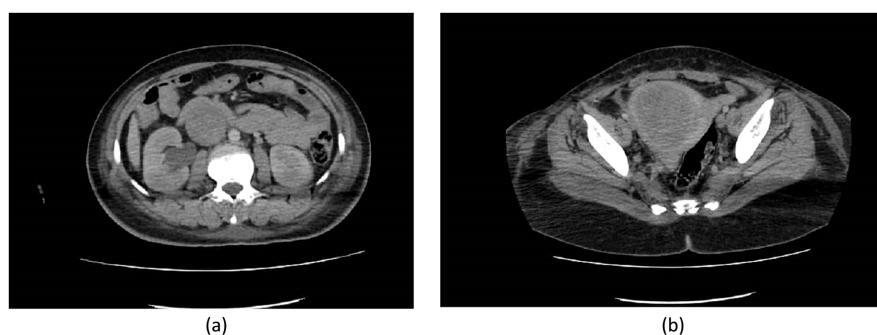


**Figure 1.** (a) Operative view of the large, yellow-colored and rubbery mass protruding from the IVC orifice and filling the right atrium; and (b) operative image of the excised specimen.



**Figure 2.** Histological and immunohistochemical study of the excised specimen showing (a) proliferation of bland spindle cells in myxoid background in intersecting fascicles, H&E  $\times 100$ ; (b and c) H&E staining,  $\times 100$ ; (d) CD34 immune staining,  $\times 100$ ; (e) negative Calretinin immune staining,  $\times 100$ .

One week postoperatively, a multi-slice computed tomography was performed and showed the presence of a filling defect in the lower IVC measuring  $140 \times 60$  mm, along with renal vein congestion. A mixed-density uterine mass ( $110 \times 80$  mm) was also noted, causing uterine enlargement, right ureter compression, and moderate hydronephrosis (**Figure 3(a)** and **Figure 3(b)**).



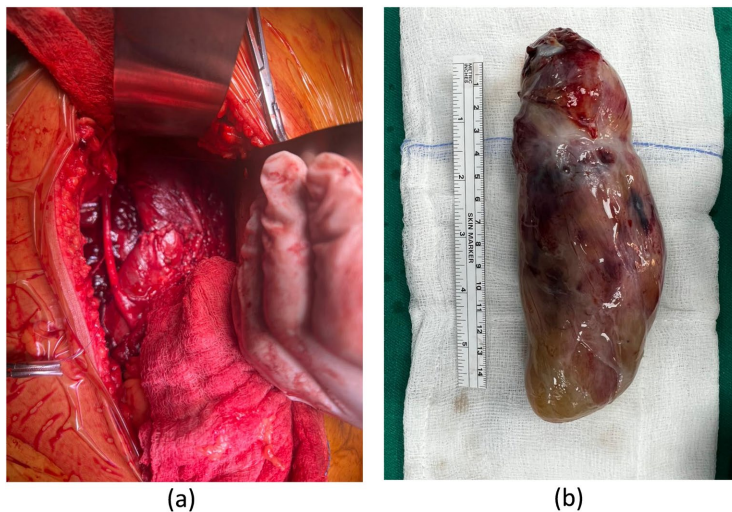
**Figure 3.** (a) Multi-slice computed tomography showing the presence of a filling defect in the lower IVC with moderate hydronephrosis; and (b) a mixed-density large uterine mass.

Two weeks after the first procedure, pelvic exploration was carried out through a Pfannenstiel incision. The uterus was found to be significantly enlarged ( $140 \times 120$  mm) with multiple leiomyomas, and both ovaries were markedly enlarged. Multiple nodules were also noted along the course of the right uterine artery. Total hysterectomy with adnexectomy was performed (**Figure 4**), and abdominal wall layers were closed sequentially. In the same setting, the IVC was explored through

a right paramedian abdominal incision and right retroperitoneal approach, revealing a markedly dilated IVC. Proximal and distal control of the IVC were done, and the vessel was incised transversely, removing a 160 × 60 mm smooth homogeneous mass. The IVC was repaired directly (**Figure 5(a)** and **Figure 5(b)**). No clear attachment point of the myxoma within the pelvic veins could be identified. Histopathological examination of the excised specimens confirmed that the IVC mass was identical to the right atrial myxoma, while the uterine mass was a distinct leiomyoma.



**Figure 4.** Surgical specimen of total hysterectomy with adnexectomy.



**Figure 5.** (a) Operative view of the enlarged IVC following excision of the intra-caval myxoma; and (b) operative image of the excised smooth homogeneous mass.

Postoperative recovery was uneventful, and the patient was discharged home one week after surgery. Follow-up confirmed complete resolution of her symptoms, with no cardiac, renal or gynecologic complaints. At 6 months of follow-up,

repeated transthoracic echocardiography and abdominal ultrasound have not shown any signs of local recurrence or elevated pulmonary artery pressure.

### 3. Discussion

Myxomas are the most common primary cardiac tumors, mostly encountered in the left atrium [1], while right atrial occurrence is less common. Symptoms of right atrial myxomas depend on tumor size and location, and may include exertional or resting dyspnea. Right atrial myxomas in particular have been noted to be particularly prone to fragmentation and distal embolization [1] [2], which can lead to the development of chest symptoms, hypoxemia or even to potentially life-threatening pulmonary embolization, and may require immediate surgical intervention [3]. Surgery is indicated once diagnosed, regardless of severity of symptoms [4].

Inferior vena cava myxomas are very rare, and all reported cases were located above the hepatic veins [5]. To our knowledge, an extra-cardiac myxoma originating from the subhepatic or pelvic venous segments of the IVC has not been previously documented, and this highlights the need to consider the possibility of atypical infra-diaphragmatic venous myxomas in right atrial masses, and the importance of meticulous imaging when planning surgical intervention.

In addition, the unusually low origin of the myxoma in this case carries significant clinical implications. A myxoma arising within the pelvic venous system can extend cranially through the IVC before reaching the right atrium, potentially delaying detection and complicating therapy. There is no single correct method for venous cannulation, and options include direct intra-pericardial SVC/IVC cannulation or femoral venous cannulation. Deep hypothermic circulatory arrest is likely to be needed for complete excision of the tumor, including the stalk, to avoid recurrence. Primary tumors typically have an attachment point, and despite the fact that we were unable to identify a clear attachment point of the myxoma within the pelvic veins during exploration, we believe that performing total hysterectomy with adnexectomy was an adequate approach to achieve complete excision of the tumor.

The specific differential diagnosis of intravenous leiomyomatosis with myxoid degeneration had to be contemplated in this case. Histological and immunohistochemical study of the excised specimen confirmed the diagnosis of a myxoma composed of proliferation of bland spindle cells in myxoid background in intersecting fascicles, with tumor cells negative for SMA and Desmin, and focally positive (few cells) for Calretinin immune staining. This immunohistochemical pattern is consistent with myxoma, particularly cardiac myxoma, which is characteristically negative for smooth muscle markers such as SMA and Desmin. In contrast, smooth muscle tumors including intravenous leiomyomatosis typically demonstrate diffuse positivity for SMA and Desmin and are Calretinin negative.

The coexistence of a uterine leiomyoma in our patient is also noteworthy. Myxomas have been reported to coincide with the presence of other benign or malign-

nant extracardiac tumors, including laryngeal and uterine tumors [6]-[8]. The present coexistence of an unusually low IVC myxoma with an independent uterine neoplasm presents a surgical challenge. There are no established guidelines on which lesion is to be approached first, even though most surgeons would tend to treat the lesion causing the most significant symptoms first, provided cardiac status is stable. Clearly, more studies are needed to develop clear protocols.

#### 4. Conclusion

This is the first report of an extra-cardiac pelvic myxoma extending through the IVC to the right atrium, and coinciding with a concurrent uterine leiomyoma. Preoperative imaging including MSCT was used for planning the surgical intervention. Complete excision was performed to prevent complications and future recurrence. This rare case highlights the importance of multidisciplinary management and careful surgical planning in these rare conditions.

#### Declarations

- **Ethics approval and consent to participate:** Not applicable. However, all procedures performed in this study were in accordance with the ethical standards of the Damascus University Research Ethics Committee and with the 1964 Helsinki declaration and its later amendments.

#### Author Contributions

- Albaraa Bara: Drafting and revising the manuscript critically, and giving final approval for the version to be published.
- Ahmad Walid Izzat: Drafting and revising the manuscript critically, and giving final approval for the version to be published.
- Hisham Hamzeh: Performing surgery, revising the manuscript critically, and giving final approval for the version to be published.
- Issa Ahmad: Performing surgery, revising the manuscript critically, and giving final approval for the version to be published.
- Bashar Kurdi: Performing surgery, revising the manuscript critically, and giving final approval for the version to be published.

Mohammad Bashar Izzat: Performing surgery, revising the manuscript critically, and giving final approval for the version to be published.

#### Conflicts of Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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