Intravascular leiomyoma with intracardiac extension associated with hepatorenal polycystic disease

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Introduction

We present the case of a woman with hepato-renal polycystic disease and chronic hemodialysis admitted for paroxysmal atrial fibrillation. A right heart mass was diagnosed by echocardiography. MRI and angio-CT scans confirmed a calcified tumor with the origin in the right internal iliac vein that extended through inferior vena cava to the right cardiac chambers. The intracardiac tumor was excised by cardiovascular surgery due to perceived pulmonary embolic risk. The tumor mass was a leyomyoma. Despite extensive inferior vena cava extension, tumor dimensions did not increase at one year follow-up and no intracardiac recurrence was noticed.

Case Report

A 55-year-old woman with hepatorenal polycystic disease and end-stage renal failure who was on chronic hemodialysis was admitted to our center because of a recent history of dyspnea and palpitations.

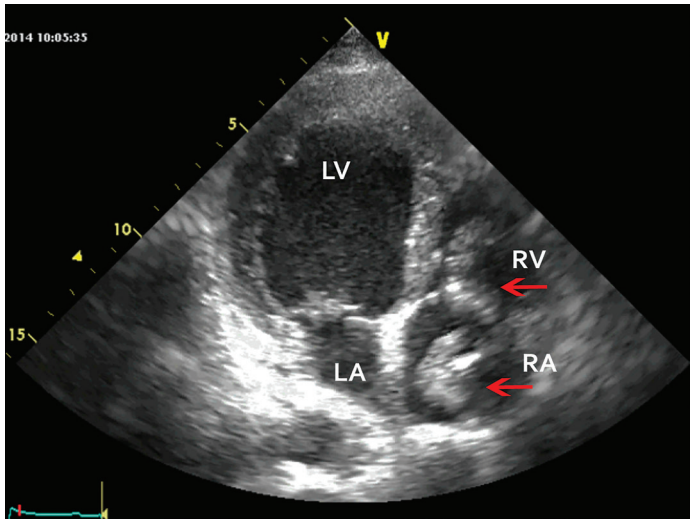

Figure 1. Transthoracic echocardiography, four-chamber view: a large polylobulated echogenic and highly mobile mass occupies most of the right atrium and prolapses through the tricuspid valve and remains trapped in systole (red arrows; RV, RA - right ventricle and right atrium; LV, LA - left ventricle and left atrium)

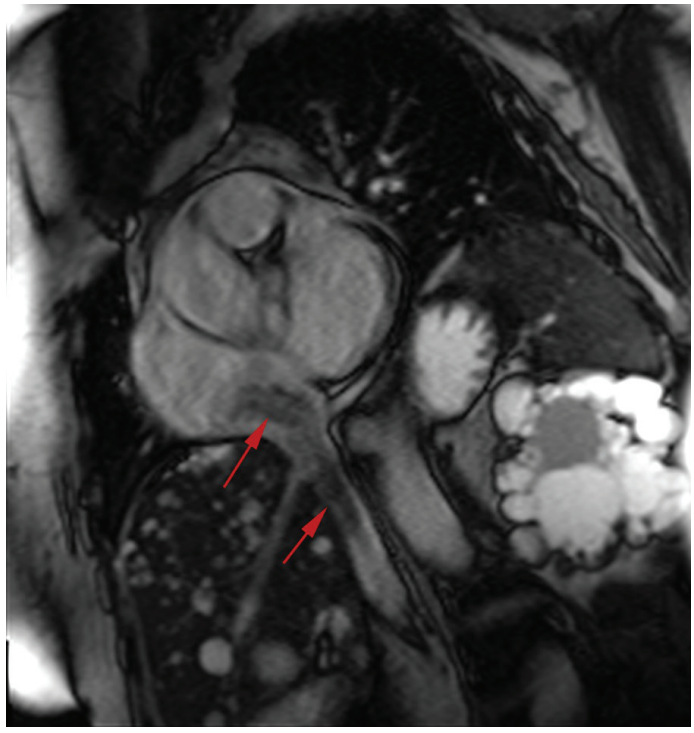

Figure 2. Cardiac MRI cine-gradient short-axis view: a hypointense structure (thrombus or tumor) extends from the inferior vena cava to the right atrium (red arrows)

Physical examination revealed wide splitting of the first heart sound and an enlarged liver and left kidney. Bilateral mild edema of the calves was observed. A 12-lead electrocardiogram (ECG) showed paroxysmal atrial fibrillation. Blood tests revealed high creatinine and blood urea nitrogen levels due to severe chronic kidney failure (serum creatinine=10.4 mg/dL).

Transthoracic echocardiography showed a large, polylobulated, echogenic, highly mobile mass that occupied most of the right atrium and extended from the inferior vena cava (IVC) and prolapsed into the right ventricle through the tricuspid valve during diastole (Fig. 1 and angiographic run in Video 1, 2).

Cardiac magnetic resonance imaging (MRI) failed to reveal if the observed mass represented thrombus or the terminal part of an intravascular tumor (Fig. 2).

CT angiography (angio CT) showed an extensive, serpiginous, highly vascularized mass originating in the right internal iliac vein, which extended to IVC up to the right cardiac chambers. Massive linear calcifications were seen along IVC and above the renal veins (confirmed by angiography run in Video. 2). Cystic transformation of the liver and kidneys was extensive. The right kidney was ectopic and occupied most of the pelvis (Fig. 3). No pulmonary artery embolus was noticed.

Coronary angiography showed calcified coronary atherosclerosis, but no significant obstructive coronary artery disease.

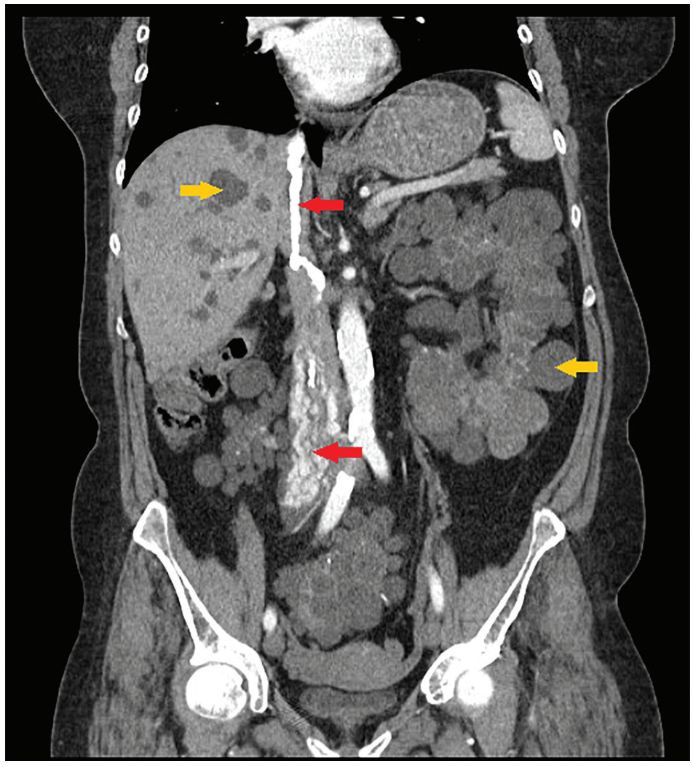

Figure 3. CT angiography frontal view (arterial phase): the tumor mass (red arrows) exceeded the outline of the inferior vena cava; massive linear calcifications were seen along the course of the inferior vena cava; the cystic transformation of the liver and kidneys (orange arrows) can be observed

Management

The terminal mobile segment of the tumor was considered to carry an increased pulmonary embolic risk, although at the time of diagnosis no signs of pulmonary embolism were noted. Therefore, surgical excision of the tumor was considered. The high calcific burden and the out of shape aspect of IVC led to the decision of a two-stage procedure.

To ensure an appropriate access and to facilitate the resection of a large tumor with unknown histology, median sternotomy was performed. A solid, cylindrical, highly adherent, 11-cm long, and 1.5-cm wide mass was extracted from the right atrium and partially from IVC (Fig. 4).

The postoperative course was uneventful, and the patient made a full recovery. She continued to receive hemodialysis thrice per week. Atrial fibrillation did not relapse, and oral anticoagulation was withheld due to high bleeding risk.

Pathological examination by optical microscopy of the extracted mass showed a hyaline central part with small peripheral groups of muscular cells. Minimal thrombotic deposits were described on its surface. Rare elastic and reticulin fibers were identified by orcein and Gomori staining, respectively. Immunohistochemical analysis revealed estrogen and progesterone receptors in 15% and 35% of the muscular cells, respectively. Muscle cells expressed actin alpha and desmin. CD31 staining identified rare, isolated endothelial cells on the tumor surface. The pathology was consistent with leiomyomatosis.

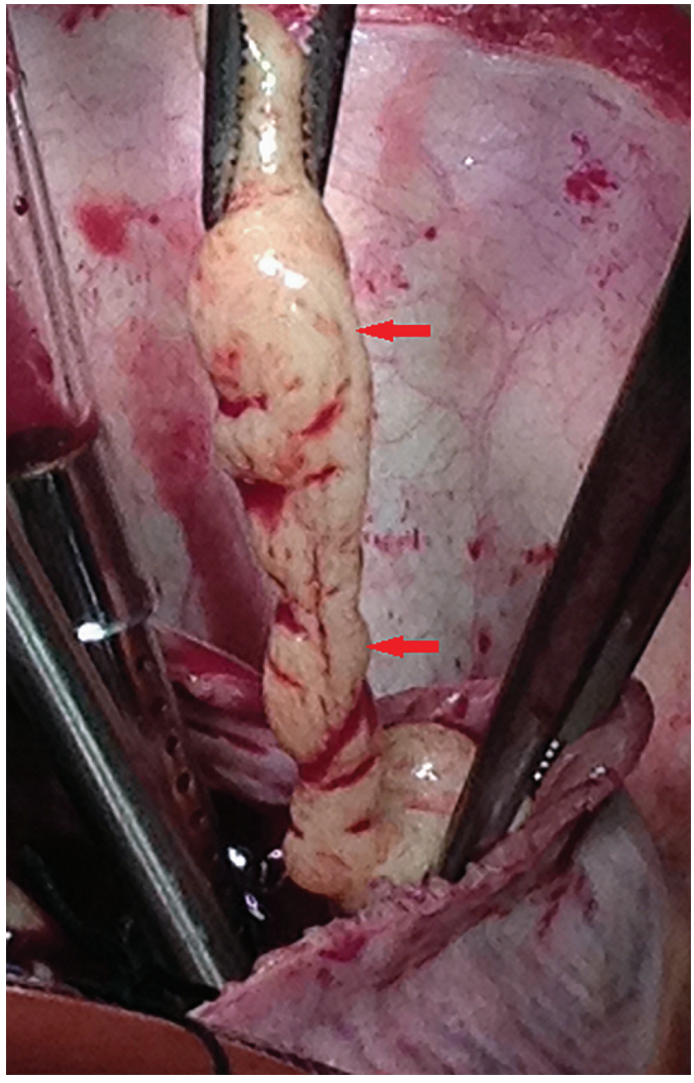

Figure 4. Intraoperative image: a solid polylobulated tumor mass (red arrows) was excised through a right atrial incision from the right heart chambers and partially from inferior vena cava. No macroscopic necrosis or calcified areas were noticed

At one-year follow-up, the patient was asymptomatic, and the tumor remained confined to IVC. The patient refused to undergo a second surgery due to the additional risks involved.

Discussion

Intravascular leiomyomatosis is a rare disorder that consists of smooth muscle cell proliferation within vascular spaces. Although it is a benign tumor, its outcome may be severe due to the extension pattern. It is usually due to a uterine leiomyoma which extends into the uterine venules and from there into IVC and upward (1). Rarely, as in the case of this patient, it originates directly from the smooth muscle cells of the walls of the internal iliac vein or IVC. Although it is cytogenetically similar, a monoclonal origin distinguishes intravascular leiomyomatosis from multiclonal uterine leiomyomas (2). As the smooth muscle

cells of intravascular leiomyomas express estrogen and progesterone receptors, it was presumed that tumor growth might respond to hormonal manipulation. However, literature data is inconclusive, and hormonal therapy needs further evaluation (3). The association between polycystic kidney disease and leiomyomatosis is highly intriguing and was reported in rats as a consequence of the somatic loss of function of the tuberous sclerosis-2 tumor-suppressor gene (4). In humans, the cytogenetic and molecular characteristics of intravascular leiomyomas have yet to be fully described.

Conclusion

We describe a case of intravascular leyomyomatosis with extension from the internal iliac vein to IVC and right heart chambers, which was symptomatic by paroxysmal atrial fibrillation. Significant comorbidities such as polycystic hepatorenal disease and chronic hemodialysis were associated. Successful cardiac mass resection was not followed by recurrence at one-year follow-up.

References

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Video 1. Echocardiographic run demonstrating the prolapsing tumor from the right atrium to the right ventricle.

Video 2. Angiographic run showing the calcified tumor all along from the intracardiac part to inferior vena cava. The distal RV prolapsing part of the tumor is highly mobile.

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