

# Succesfull Two Staged Surgical Treatment of Intravenous Leiomyoma with Intracardiac Extension: Case Report

## İntrakardiyak Uzanım Gösteren İntravenöz Leiomyomun İki Aşamalı Başarılı Cerrahi Tedavisi

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**ABSTRACT** Intravenous leiomyomatosis (IVL) with cardiac extension is a rare uterine tumor. We present an unusual case of angioleiomyoma that progressed along the inferior vena cava into the right atrium. At first, the patient was diagnosed as right atrial myxoma. The diagnosis changed after removal of the right atrial mass and the patient was diagnosed with angioleiomyoma. A second stage operation was performed involving removal of the right adnexial mass, simple hysterectomy and bilateral salpingo-oophorectomy. IVC veinotomy and tumor resection was performed using cardiopulmonary bypass. This is a short report IVL with intracardiac extension which was successfully removed with a two staged procedure.

**Key Words:** Angiomyoma; vena cava, inferior; cardiopulmonary bypass

**ÖZET** Kardiyak uzanımlı intravenöz leiomyomatozis (IVL) nadir bir uterin tümördür. İnförör vena kavadan sağ atriuma uzanım gösteren nadir bir anjiroleiomyom olgusunu sunmayı amaçladık. Başlangıçta hastaya sağ atrial miksoma tanısı konulmuş idi. Sağ atrial kitle çıkarıldıktan sonra tanı anjiroleiomyom olarak değişti. Sağ adneksiyal kitle çıkarılması, basit histerektomi ve bilateral salpingo-oofektomiye içeren ikinci aşama operasyonu planlandı. İnförör vena cava venotomi ve tümör rezeksiyonu kardiyopulmoner bypass kullanılarak gerçekleştirildi. Bu olgu sunumunda iki aşamalı cerrahi ile tedavi edilen intrakardiyak uzanımlı intravenöz leiomyomatozis vakası tartışılmıştır.

**Anahtar Kelimeler:** Anjiyomiyom; vena kava, inferior; kardiyopulmoner baypas

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**I**ntravenous leiomyomatosis (IVL) is a rare benign tumor which is characterised histologically by smooth muscle cells. It is usually confined to the pelvic veins but very rarely can progress to the inferior vena cava, and even to the heart. Intracardiac leiomyomatosis (ICL) can result in life threatening symptoms and appropriate therapy is complete excision of the tumor. The ICL case presented here depicts diagnostic and surgical difficulties that can arise from the rarity and complexity of this condition.

## CASE REPORT

A 48-year-old woman with chest pain and progressive dyspnea admitted to cardiology clinic, and the diagnosis was set as atrial myxoma by echocardiography, and an operation was scheduled. There was no abdominal symptoms at that time. The patient underwent median sternotomy and atriotomy under cardiopulmonary bypass (bicaval cannulation). At operation, the mass

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was found to be an extension of a tumor emanating from the inferior vena cava (IVC). The atrial component and a portion of the lesion within the IVC were excised and the operation was terminated. The patient spent 9 uneventful days in hospital including one day in intensive care unit (ICU). The final histologic analysis of the lesion revealed a benign angioleiomyoma. Computed tomography was performed, and it demonstrated a right adnexial mass with 100x72x65 mm dimensions, and a big mass, reported as thrombus, beginning from right ovarian vein, rising up through the inferior vena cava (Figure 1a, 1b) which was confirmed by venography (Figure 2).

Patient was counsulted to gynecology department 4 weeks after the initial surgery with the suspicion of a pelvic malignency and for possible vascular invasion. Her cervico-vaginal smear was normal. Her tumor markers were not related with any kind of gynecological malignency. A second operation was scheduled. Surgery was performed utilising cardiopulmonary bypass, and involved removal of the right adnexial mass, simple hysterectomy and bilateral salphingooferectomy, IVC veinotomy and tumor resection by median laparotomy (Figures 3a, 3b). Patient was cooled to 32 degrees and no circulatory arrest was used. Tumor within the IVC was rubbery in consistency and adherent to but not infiltrating the vein wall; it was easily removed by gentle traction. An additional embolectomy was performed by using a 4F Fogarty catheter. The veinotomy was primarily closed with 5/0 polypropelene suture. Pathological analysis confirmed benign leiomyomatosis for the second time, the patient was hospitalized for seven days including 2 days spent in ICU.

## DISCUSSION

Intravenous leiomyoma is a rare benign intravascular tumor for which the tissue of origin is not clearly understood. This tumor is found exclusively in women; many have undergone hysterectomy because of uterine leiomyoma. While some believe the tumor results from vascular invasion from a uterine leiomyoma, it has also been proposed that the tumor is vascular in origin and derived from

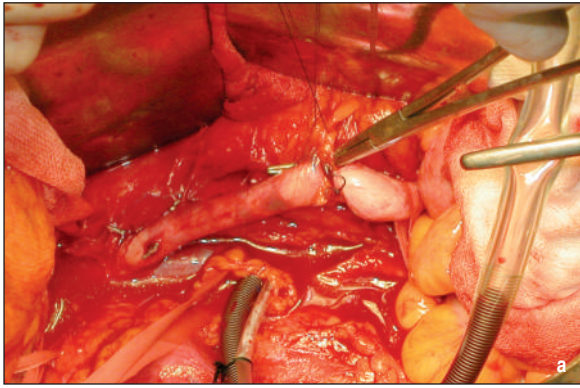


**FIGURE 1a,b:** Computed tomography was performed, and it demonstrated a right adnexial mass with 100x72x65mm dimensions and a big mass reported as thrombus beginning from right ovarian vein, rising up through the vena cava inferior.

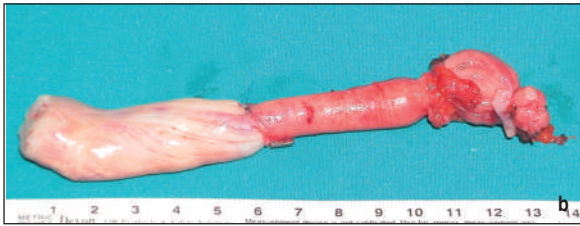


**FIGURE 2:** Venography demonstrated the mass in vena cava inferior.

medial smooth muscle cells of a vessel wall.<sup>1,2</sup> The tumor is most commonly isolated in the pelvic veins, but occasionally extends into the IVC, the right



**FIGURE 3a:** Surgical view of IVC veinotomy and tumour resection.



**FIGURE 3b:** Postoperative view of the resected tumor.

atrium, or even the pulmonary circulation. Attachment of the tumor to the wall of the IVC or atrium has not been described in benign leiomyomas, and the finding of wall attachment at echocardiography should raise suspicion of a leiomyosarcoma. A surprising and unusual characteristic of benign leiomyomas is that they grow freely within the vessel lumen but do not invade the wall. Lam et al. have recently reviewed the literature and identified 200 reported cases of leiomyomatosis, 68 of which had intra-cardiac extension.<sup>3</sup> They identified signs of cardiac failure, venous obstruction or abdominal distension in women with a history of hysterectomy as being the most common mode of presentation in those with involvement of the right atrium. In the early stages of venous extension, the diagnosis has often remained unrecognized during hysterectomy and the patient presents due to further extension of intravenous leiomyomatosis not prevented by resection of the primary tumour.

Surgery can be performed as both single and staged procedures and cardiopulmonary bypass is advocated for resection of tumors with intracardiac extension.<sup>3</sup> Cardiopulmonary bypass with circulatory arrest is used to create a bloodless field and facilitate complete resection of the tumor with minimal blood loss.

If the tumor is too extensive, or adheres to the cardiac and vascular structures requiring resection of the abdominopelvic and intrathoracic components, then a separate operation may be mandatory; otherwise, one-stage resection under total circulatory arrest and hypothermia can be used with success. In this case, we performed a two-staged operation because the tumor was located within the inferior vena cava and right ventricle and it was misdiagnosed as a myxoma. The correct diagnosis was made intraoperatively. Therefore, in the first step, the intracardiac mass was removed en bloc by using cardiopulmonary bypass after gentle traction was applied to the tumor limb extending from the inferior vena cava, and patient was rescheduled for the second operation. For the removal of the intracaval tumor, a literature review showed a preference of supra and infrarenal vena cava venotomy.<sup>4,6</sup> The leiomyoma usually does not adhere to the vessel wall, therefore some authors recommend iliac venotomy for removal of the remaining caval portion.<sup>7,8</sup> This incision may have some advantages over caval and especially suprarenal caval level incisions: such as fast recovery due to less retroperitoneal exploration, easier surgical complications of the iliac region to cope with compared to the caval region, more tolerable venous thrombosis in the iliac region when compared with caval region and better cosmetic results. On the other hand, there is no guarantee that the tumor will be easily removable and it may persist along the vein lumen. In addition it vena cava rupture occurs due to uncontrolled traction, the result may be catastrophic.

In conclusion, surgical resection is the best treatment for intracardiac extension of intravenous leiomyoma and must be performed immediately because of sudden death. We recommend cardiopulmonary bypass and proximal control of the venous system for any possible hazardous complication, and a concomitant venotomy as a safe approach to remove the caval portion of the tumor in intravascular leiomyomatosis in a two-staged operation.

### **Conflict of Interest**

*Authors declared no conflict of interest or financial support.*

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