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Case report of an intracaval leiomyomatosis 10 months after complete hysterectomy



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ABSTRACT

INTRODUCTION: Intravenous leiomyomatosis (IVL) is a rare smooth muscle tumor, usually found in women with tumors of the reproductive organs, such as uterus myomatous. Surgically, this case belies the call for sternotomy and two-stage surgery in caval IVL extending to the right atrium: we suggest one-stage median laparotomy as a minimal procedure with maximal benefit.

PRESENTATION OF CASE: We present the case of a 60-year-old postmenopausal woman with suspected intravenous leiomyomatosis of the right internal iliac vein. The patient had undergone hysterectomy and bilateral adnexectomy for uterus myomatous in September 2015, where an IVL limited to the veins of the uterus and the right adnex had been diagnosed. No further medical treatment had been implemented. IVL of the inferior vena cava was diagnosed when a CT scan of the abdomen was performed due to an infected abdominal seroma in June 2016. Although histologically benign, we found this case of IVL to be clinically aggressive because of its expansion to the heart. This may lead to thromboembolic complications (e.g. pulmonary embolism) or signs of right sided cardiac failure. The patient was asymptomatic, but because of the extension of the intracaval thrombus to the heart, we decided to operate and performed thrombectomy via a median laparotomy. The patient left the hospital shortly after on newly started oral anticoagulation.

DISCUSSION: For caval IVL without intracardiac attachment, the extraction via laparotomy without sternotomy is the treatment of choice. It calls for an interdisciplinary approach and careful surgical planning. **CONCLUSION:** There is no inherent need for sternotomy in IVL extending to the right atrium. A one-year follow-up with sonographic control is advised. Medium term oral anticoagulation should be considered.

This work has been reported in line with the SCARE criteria Agha et al. (2016). The SCARE Statement: Consensus-based surgical case report guidelines. Agha RA, Fowler AJ, Saeta A, Barai I, Rajmohan S, Orgill DP; SCARE Group. *Int J Surg.* 2016 Oct;34:180-186 [1].

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1. Introduction

Leiomyomatosis is a benign muscle cell tumor. Its primary localisation is the uterine venous system, from where it can spread into the inferior vena cava. IVL was first described in 1896 [2], a first case of intracardiac extension of IVL was published by Dürck in 1907 [3]. Growth of the tumor sometimes extends as far as intracardially, intrahepatically or intrapulmonary, leading to serious complications such as pulmonary embolism or signs of right heart failure [4,5]. Clinical manifestation includes respiratory and cardiac symptoms (dyspnoea, orthopnoea, tachycardia, syncope), typically as late-onset. The aetiology of the tumor has not been conclusively settled: both growth from the vein wall and invasion

of the venous system originating from intrauterine leiomyoma are being discussed [5].

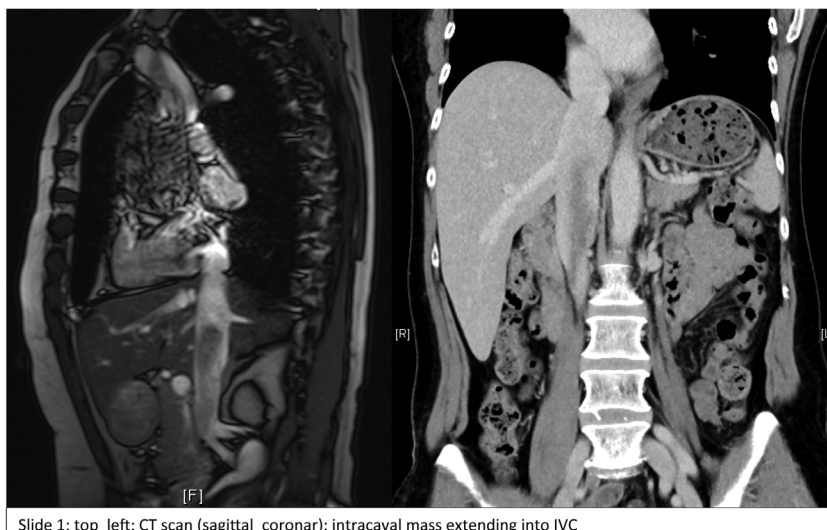
2. Presentation of case

Our patient was transferred to University Hospital Basel from an external regional hospital for treatment of an intravenous mass in the inferior vena cava (IVC). Initially, she had been hospitalised for treatment of a wound infection. The diagnosis of IVL was an incidental finding.

In September 2015, the patient had undergone hysterectomy and bilateral adnexectomy because of a myomatous uterus. The histology at that time showed intravenous leiomyomatosis in the veins of the uterus as well as the right adnex. There was no histopathologic sign of malignancy. Following an aesthetic surgical intervention, the patient developed an infected abdominal seroma. The CT scan showed a mass occluding the right internal iliac vein which extended into the IVC (Fig. 1). Although extending well intrahepatically, the tumor mass did not entirely occlude neither the right common iliac vein nor the IVC.

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Slide 1: top left: CT scan (sagittal coronar): intracaval mass extending into IVC

Fig. 1. Top, left: CT scan (sagittal, coronar): intracaval mass extending into IVC.

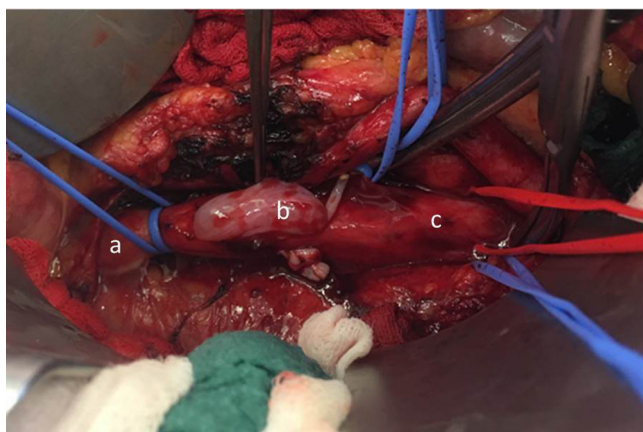


Fig. 2. Intraoperative site. a: proximal inferior vena cava. b: inferior vena cava with protruding thrombus through cavotomy. c: distal inferior vena cava.

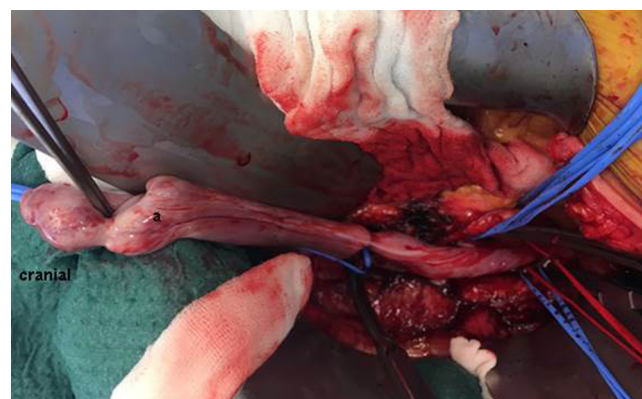


Fig. 3. Intraoperative site. a: intravascular tumor, extracted from inferior vena cava.

Upon admittance, there was no clinical evidence of caval thrombosis; the patient had no complaints of swelling, pain, cardiac or pulmonary symptoms. We started her on unfractionated heparin.

A biopsy of the intravenous thrombus was inconclusive. For the planning of the operation, it was crucial to determine the proximal extension of the thrombus. Therefore, a transoesophageal echocardiography was performed as well as a cardiac MRI. The findings were ambivalent, which is why we prepared for two possible intraoperative scenarios: 1) a freely floating thrombus easily extractible without further surgical exploration; 2) extension of the thrombus intracardially and/or adherent to the vessel wall, necessitating a sternotomy and cardiac exploration. The operation was prepared with the cardiac surgical team in standby and the patient draped for additional sternotomy and circulatory arrest. Median laparotomy was performed, followed by preparation of both common iliac veins. We then prepared the IVC and right internal iliac vein. This was followed by venotomy of the IVC, the thrombus protruding out of the vessel upon cavotomy (Fig. 2).

The major part of the tumor was freely floating, two more, very slender tumorous formations extended as far as 25 cm cranially and appeared fast attached, but could ultimately be removed by extraction. Next, the right internal iliac vein was cut and the tumor extracted. Figs. 3, 4. The vessel was ligated as far distally as possible.



Fig. 4. Resections. From top to bottom: wall adherent thrombus in internal iliac vein; freely floating intracaval thrombus (extracted cranially to cavotomy); wall adherent thrombus (extracted from vena cava); freely floating caval thrombus, protruding upon venotomy.

Closure of the cavotomy by running suture. Closure of the retroperitoneal space and the abdomen. The patient was stable during the entire procedure, markedly crossclamping of the vena cava was well tolerated hemodynamically. Postoperatively, the patient

was started on oral anticoagulation (Marcoumar). Gynaecological oncologists advised against the use of aromatase inhibitors. She made an uneventful recovery and was discharged from the hospital 10 days after surgery, with a follow-up planned for 6 months postoperatively. There was a delay of 20 days between hospitalisation and surgery, which was due to extensive diagnostic procedures and intricate surgical planning. The surgery was performed by the head of the vascular surgical department.

In the immunohistochemical study, the tumor cells were positive for smooth muscle actin and desmin, coherent with the histological diagnosis of an IVL [6]. No signs of malignancy could be found.

3. Discussion

IVL is a relatively rare entity. In hindsight, it cannot be determined if the intracaval mass had already been present at the time of hysterectomy in September 2015. Thus, we do not know if it is to be regarded as a remnant of the leiomyomatous thrombus found in the internal iliac vein. If it appeared only after hysterectomy, histological it must be considered a dissemination of the original thrombus found during hysterectomy in the right uterine vein. Drawing from intraoperative presentation and radiology, growth seemed to be originating from the internal iliac vein (IIV), extending proximally well intrahepatically. Curiously, vessel wall attachment was stronger in the proximal and distal parts of the thrombus, whereas the medium part in the inferior vena cava was entirely free-floating. In the IIV, where the IVL was first observed, attachment was strongest. In a patient with pre- or intraoperatively diagnosed IVL, total resection should be aimed at during radical hysterectomy in order to prevent recurrence [7]. In the pathology report of the hysterectomy in September 2015, it is unclear whether the IVL of the right adnexa is confined to the resected area or has already spread beyond. It remains to be seen whether ligation of the IIV will prevent reformation of the leiomyomatosis.

Intracardiac extension of IVL occurs in approximately 10% of cases described and is often clinically undetectable [8]. Clay et al. report single stage sternolaparotomy as the most common surgical treatment in intracardiac IVL extending to the right atrium [6]. In order to reduce surgical strain, we recommend a single stage laparotomy whenever oncologically feasible. Despite extensive diagnostic investigation, cardiac attachment of the thrombus could not be ruled out. However, we believe that a primary abdominal approach is rational as long as preparations for surgical escalation are made. Especially in view of the origin of the disease in the reproductive organs, we find the abdominal approach more promising regarding the complete resection of the tumor. Therefore, a “cardiac only” surgical approach (8.8% of cases reported in the literature) [6] seems counterintuitive.

To our knowledge there is no standardized follow-up protocol for IVL. Some authors confine controls to a one-year follow-up [7], while others recommend long term monitoring because of possible recurrence [3,5]. Due to the late, but serious clinical manifestations we advocate the latter approach. Sonography/TTE should be the diagnostic of choice, reducing radiation exposure and unequivocally determining cardiac involvement [9]. Medical treatment such as a gonadotropin-releasing hormone agonist and aromatase inhibitor is advised by some authors [7,10]. As our patient is already postmenopausal, aromatase-inhibitor treatment is less promising than in premenopausal women [5,11].

4. Conclusion

Removal of IVL with extension to the right atrium is feasible via single stage median laparotomy in standby for sternotomy. A one-year follow-up with sonographic control is advised.

Conflicts of interest

There are no conflicts of interest between this article and its contributors.

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Ethical approval

No ethical approval was necessary. The patient gave her consent to publishing data relating the case anonymously.

Consent

The patient agreed to the use of anonymised data.

Authors contribution

Lorenz Guerke: editing, main surgeon.

Andrej Isaak: editing, photography, assistant surgeon.

Hannah Schäfer: writing of article, literature research, assisting surgeon.

Guarantor

Hannah Maria Schäfer.

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