



### Leiomyosarcoma of the Right Iliac Veins Presenting as a Pelvic Mass: A Case Report

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#### Abstract

**Background:** Leiomyosarcoma of the iliac veins is a very uncommon tumour. Prognosis is usually poor and the best treatment is unknown.

**Case report:** We report the case of a 74-year-old woman presenting a right leg oedema. Venous colour doppler was normal. Abdominal ultrasound and magnetic resonance scanning demonstrated a 70 mm mass in the right pelvis with a well-defined profile, which was not homogeneous after contrast enhancement. The lesion was in close contact with the iliac vessels and the right iliac veins profile was inside the mass. Serum markers were in the normal range. The patient underwent an exploratory laparotomy by a team of Gynecologic Oncologists for the suspicion of ovarian cancer, but a retroperitoneal tumour on the wall of the right common iliac and external iliac veins was found. With the help of a vascular surgeon the tumour was resected en bloc and polytetrafluoroethylene grafts were used to reconstruct the vessels. The postoperative course was uneventful. On histological examination a leiomyosarcoma of the iliac veins was found. The patient received adjuvant chemotherapy. Three years after surgical resection no signs of recurrence or postoperative complications were detected.

**Conclusions:** Leiomyosarcoma of the iliac veins is a rare option to consider in the management of a pelvic mass. Radical surgical excision with polytetrafluoroethylene grafts reconstruction followed by adjuvant chemotherapy was feasible and effective.

#### Keywords

Leiomyosarcoma, Pelvic mass, Iliac veins, Polytetrafluoroethylene grafts

#### Introduction

Leiomyosarcomas are a relatively rare group of smooth muscle soft tissue sarcomas. Leiomyosarcomas involving large blood vessels are quite unusual, comprising of less than 2% of all leiomyosarcomas [1]. Vascular leiomyosarcomas originate five times more often in arteries than in veins. Primary venous leiomyosarcomas are rare slow-growing tumours that originate from the smooth muscle tissue of vessels, usually the inferior vena cava. Since the first case reported by Perl in 1871, few cases (mostly single case reports or small series) have been described, with little information on follow-up [2]. This type of tumour has late diagnosis because of the varying clinical presentation and generally has a poor prognosis. Data on optimum

treatment is limited due to the paucity of cases. However, radical surgical excision appears to offer the best chance of cure. The possible benefits of adjuvant therapy have not been proven.

We report the case of a primary leiomyosarcoma of the right iliac veins presenting as a pelvic mass.

#### Case Report

A 74-year-old woman presented an oedema of the right lower limb. She was not a smoker and had no risk factors for venous thrombosis.

Venous colour doppler displayed no signs of deep vein thrombosis. Abdominal ultrasound showed a hypo-anechoic image of 46x45mm at the level of the uterus, in the right paramedian position of the pelvis. Magnetic resonance imaging of the inferior abdomen and pelvis showed a 70mm mass localised at the right side of the pelvic cavity with a well-defined but irregular profile. At contrast enhancement the mass impregnation was not homogeneous. The lesion was in close contact with the iliac vessels with an apparent greater involvement of the right iliac veins that presented themselves as incorporated in the context of the mass. A modest extrinsic compression of the iliac muscle was observed, but the lesion seemed to be cleavable. The radiologists hypothesized an adnexal tumour, so CT/MR angiogram was not performed. The patient also underwent a computerised tomography of the thorax and upper abdomen as preoperative staging: metastatic lesions were not found.

Laboratory findings, including the serum concentration of the oncologic markers (Ca125, Ca15.3, CEA and Ca19.9), were in the normal range.

The patient underwent an exploratory laparotomy by a team of Gynecologic Oncologists for the suspicion of ovarian cancer. At the exploration of the abdominal cavity both the uterus and adnexa were regular and no adnexal masses had been detected. A large retroperitoneal tumour on the wall of the right common iliac and external iliac veins was found (Figure 1). With the help of a vascular surgeon they proceeded to separate the right iliac vessels (common iliac, external iliac and internal iliac veins). Then they isolated and removed the mass, which was well encapsulated and dissociable from the surrounding plans (Figure 2). At frozen section examination a leiomyosarcoma of the common and

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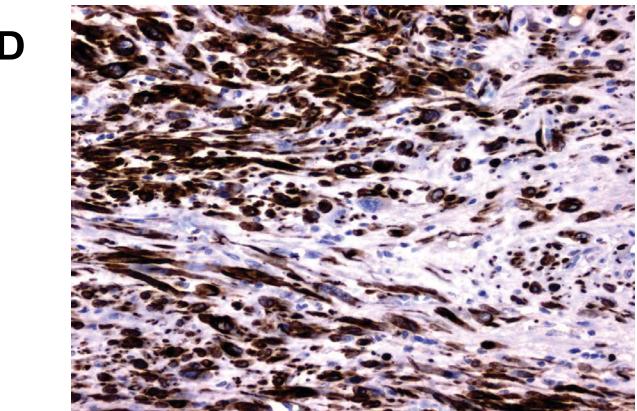
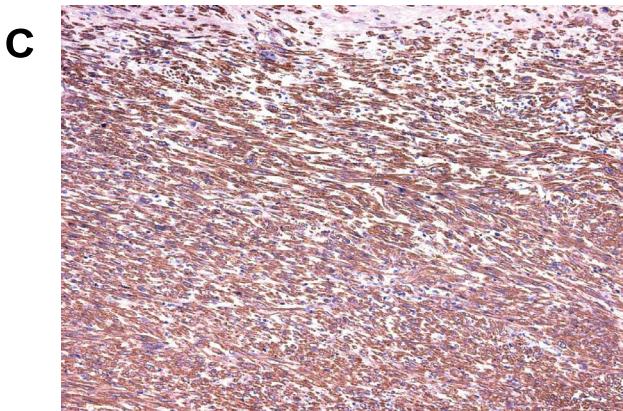
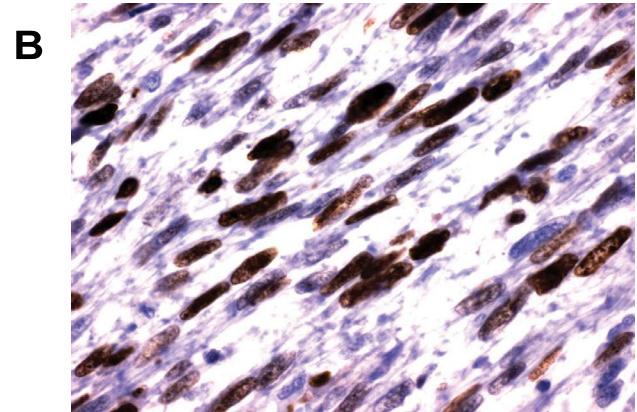
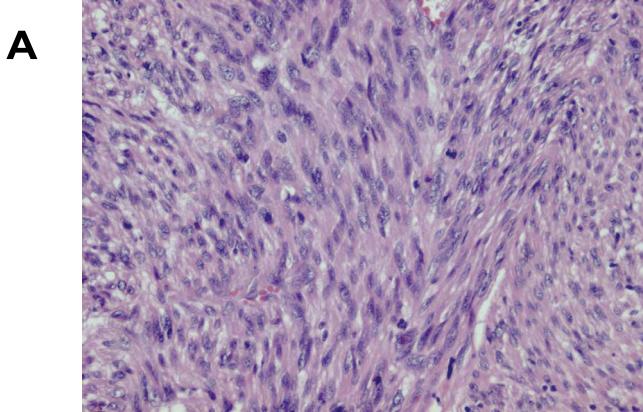
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**Figure 1:** Intraoperative finding: a large retroperitoneal tumor adherent to the right iliac vessels.



**Figure 2:** The resected specimen: a segment of the iliac veins wall excised en bloc with the tumor.



**Figure 3:** Pathological features. **A:** proliferation of atypical spindle cells with elongated, blunt-ended, or cigar-shaped nuclei and densely eosinophilic cytoplasm arranged in interlacing bundles, admixed with scattered bizarre cells and mitotic figures (H&E,  $\times 20$ ). **B:** high expression of Ki-67 ( $\times 40$ ). **C:** strong reactivity for smooth muscle actin ( $\times 10$ ). **D:** strong reactivity for desmin ( $\times 20$ ).

external iliac veins was detected. Polytetrafluoroethylene grafts were used to reconstruct the vessels.

Post-surgery evaluation of the pathology specimen confirmed a leiomyosarcoma originating from the right iliac veins, positive for actin and desmin at immunochemistry (Figure 3A-3D). The lesion was 7,2cm large, and grade 3 according FNCLCC (score 7). Lymphovascular space involvement and perineural invasion were negative. Resection margins were free from tumour.

The patient tolerated the surgery well and had an uneventful recovery. Venous thrombus-embolic prophylaxis with low molecular weight heparin was performed (6000 UI twice a day).

It was considered greater the risk of distant relapse than local recurrence, so she received three cycles of adjuvant treatment with Epirubicin (50mg/mq) and Ifosfamide (1800mg/mq in 5 days).

The patient undergoes lower limb venous colour doppler and clinical examinations every six months. Three years from surgery she has no recurrence and remains without signs of lower extremity venous congestion. Polytetrafluoroethylene grafts are patent.

## Discussion

Leiomyosarcomas represent 10% to 15% of connective tissue sarcomas, 45% of retroperitoneal tumours [3] and 0.5% of sarcomas in adults [4]. Primary vein leiomyosarcoma is a rare type of vascular sarcoma that arises from the spindle cells of the venous wall; growth pattern may progress from intramural to endoluminal, extraluminal, or mixed forms [5]. Approximately half of primary venous leiomyosarcomas originate in the inferior vena cava [6]. Less frequently, great saphenous vein (25%), femoral vein, internal jugular vein, and iliac vein involvement have been reported [7]. Primary

venous leiomyosarcoma of the inferior vena cava is more frequent in women in their sixth decade [5] although reports of younger patients are found [8]. Pathological classification follows retroperitoneal leiomyosarcoma criteria based upon mitotic figure count [9]. There are less than 250 cases reported to date, with a 60 % 5 year overall survival rate [10].

Clinical presentation is usually aspecific and physical examination yields normal findings. Thus, its diagnosis is frequently overlooked. However, at time of diagnosis, more than one half of the cases already present pulmonary metastases [11]. When symptoms are present, they depend on the location, size and growth rate. Acute venous thrombosis is the most common presentation of the iliac vein leiomyosarcoma. Other symptoms may occur due to compression of the iliac artery and pelvic organs. Primary venous leiomyosarcomas are clinically divided into non-occlusive, occlusive or terminal [12]. Other frequent clinical presentations include abdominal pain [13], consumptive symptoms (loss of weight, anorexia, or asthenia), fever, night sweats, nausea, vomiting, and dyspnoea [14]. On physical examination an abdominal mass may be detected. Other less frequent presentations include haematuria [15], a renal mass [16], or a cardiac tumour [17]. In most cases, the tumours are diagnosed in the occlusive stage. In our case, the patient had only an aspecific like symptom as oedema of the right lower limb, so it was difficult to reach the correct diagnosis. Preoperative imaging techniques were not definitive in identifying the origin of the mass in our patient.

Although hematogenous spread commonly leads to secondary deposits in the lung, liver, and brain in primary venous leiomyosarcoma and in later stages, lymphatic pathways may be involved in metastasis [18]; in our patient no metastatic lesions have been found.

Due to the limited cases of primary venous leiomyosarcoma, optimal treatment is yet to be established [19]. However, an aggressive surgical approach assuring a free margin *en bloc* tumour resection can be curative [19] despite reported local recurrence rates in selected cases of 40 to 60% [17]. Our patient underwent radical surgery and 3 years after, the local response is excellent.

The tumour may present difficulty of vascular reconstruction after *en block* resection. [20,21] The controversy in these types of cases was whether to reconstruct the iliac veins and how it could be done. The veins can be managed with primary repair, ligation, or prosthetic graft. The current published data suggests a high rate of polytetrafluoroethylene graft occlusion, with a patency of 62% at 3 years reported by Jost and 60% at 12 months in case series by Caldarelli [22,23]. At the same time, patency of the autologous vein graft appears to be higher [22]. Postoperative morbidity such as leg oedema with pain and tenderness, ulcerative sequelae, and venous claudication has been documented in a number of cases [21,23]. Reconstruction using a polytetrafluoroethylene graft can be indicated to bridge wide gaps in the vein, but clinical outcomes with prosthetic grafts in peripheral vascular reconstruction are inferior to those with autologous vein bypass grafts, in part because of anastomotic neointimal hyperplasia. Polytetrafluoroethylene grafts were used to reconstruct the vessels in our case, the patient had no postoperative complications and after 3 years there are no signs of obstruction or evidence of neointimal hyperplasia.

The role of chemotherapy and radiation therapy for all retroperitoneal sarcomas in general and for venous sarcoma in particular is not clear [24]. In our case the patient received 3 cycles of chemotherapy with Epirubicin (50mg/m<sup>2</sup>) and Ifosfamide (1800mg/m<sup>2</sup> in 5 days), the treatment was well tolerated with no toxicity greater than grade 2.

Patients with primary venous leiomyosarcoma have a mean survival of 3.5 years, and a favourable outcome relies greatly upon eligibility and achievement of a free-margin resection [25], as has been achieved in the patient that we treated at our centre. Comparatively to other sarcomas, primary venous leiomyosarcoma has a shorter metastases free interval and survival [26].

## Conclusion

Primary malignant tumours of vascular origin are rare. Leiomyosarcoma of the iliac veins is a rare option to consider in the management of a pelvic mass and is important to know in the differential diagnosis of gynecologic pelvic diseases. A mass causing venous stasis, with oedema, swelling, or thrombosis of the lower limbs, is frequently the manifestation of a leiomyosarcoma of the iliac veins. Preoperative diagnostic procedures may not always successfully identify the origin. Therefore, definitive diagnosis can only be possible with surgery and subsequent histopathologic examination.

Optimal treatment strategies are unclear. Complete surgical resection with negative margins offers the only chance of long-term survival [24]. The reported case concerns a leiomyosarcoma of the right iliac veins with extraluminal growth. Radical surgical excision of the lesion with polytetrafluoroethylene grafts to reconstruct the vessels and adjuvant chemotherapy allowed our patient to have three year without recurrence.

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