Multifocal Sarcoma Mimicking Superficial Vein Thrombosis of the Leg: A Case Report

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Patient: Female, 61-year-old
Final Diagnosis: Leiomyosarcoma
Symptoms: Pain • redness • tenderness • warmth
Medication: —
Clinical Procedure: Excision of tumor
Specialty: Surgery

Objective: Unusual clinical course
Background: Leiomyosarcoma is a common tumor found in soft tissue. In relation to the vascular system, leiomyosarcoma appears as the most common malignancy characterized by poor prognosis. Leiomyosarcomas of the leg large vessels often occur late, and their appearance can imitate vein thrombosis with symptoms such as soft tissue swelling or mild pain, and can be misdiagnosed. Peripheral vascular leiomyosarcomas are rare. Especially leiomyosarcomas of the great saphenous vein are uncommon. The tumors develop on the media basis and grow from endovascular to exovascular order. Distant metastasis can be identified and worsen prognosis.

Case Report: We present a case of a 61-year-old female patient with varicose vein disease complicated by recurrent superficial vein thrombosis. After 2 months of conservative treatment, while waiting for admission to the department of surgery, she developed additional symptoms. Clinical examination on the day of admission revealed several tumors along and near the great saphenous vein on the left limb below the knee. The diagnosis of leiomyosarcoma was confirmed after the surgery, involving excision of the saphenous vein, including tumors formed on its course. Preoperative clinical and ultrasound findings did not suggest malignancy.

Conclusions: Leiomyosarcoma of the great saphenous vein is an extraordinarily rare tumor originating from the middle layer of the vessel, mimicking unspecific symptoms and complicating and delaying diagnosis. In every case of vascular or perivascular lesions, a detailed examination and diagnosis it is required, and even unlikely clinical scenarios should be considered.

Keywords: General Surgery • Leiomyoma • Varicose Veins • Venous Thrombosis

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Background

Soft tissue sarcomas (STS) account for less than 1% of all malignant neoplasms diagnosed in adults [1]. The incidence in Poland is estimated at around 900 cases per year, almost equally in men and women. Superficial vein thrombosis (SVT) of the legs are often seen in association with varicose veins. Many of the SVT risk factors are also correlated with other thrombotic conditions, including cancer [2].

Leiomyosarcoma (LMS) is an aggressive and relatively rare form of STS. Vascular leiomyosarcoma is an ultra-rare disease, accounting for <2% of all smooth-muscle tumors. It is 5 times more likely to occur in veins than in arteries, with 30% of all the cases attacking the great saphenous vein (GSV). Because of its asymptomatic nature, leiomyosarcoma diagnosis and treatment are often delayed and have poor prognosis [3-5]. LMS can originate from any smooth muscle, including the tunica media of blood vessels, and thus can be found in any part of the body that contains these structures [6].

In the legs, the GSV is the most common site of origin for primary vascular LMS, with the majority of cases occurring proximal to the knee near the inguinal region [7]. There have been few recorded cases of LMS of the greater saphenous vein found at the level of the ankle. Due to the rareness of the disease, information regarding this form of cancer relies on case studies or small case series [8].

At present, there are no specific image or laboratory tests that could help diagnose LMS. Thus, a histological examination of the sample seems to be the only way to make the proper diagnosis [9,10].

Case Report

A 61-year-old woman with no significant past medical history, including no drinking, no smoking, and no diabetes, presented to a surgeon with symptoms suggesting SVT. She had experienced exacerbation of symptoms for 1 year. She reported having pain, warmth, redness, and tenderness over a superficial vein of her left leg. Clinical presentation was accompanied by “cord-like” and “tumor-like” structures less than 1 cm in diameter upon palpation, suggesting superficial thrombophlebitis. An ultrasound (US) exam revealed that thrombus in the saphenous vein almost reached the ostium of the femoral vein at the saphenofemoral junction. Thus, the patient was referred to our department for surgery. The time between surgical consultation and admission was 2 months, but at the moment of intervention, the clinical presentation of SVT was not typical. The primary “tumor-like” thrombosed veins were much bigger (5 and 10 cm in diameter) and suggested a malignant background rather than simple SVT (Figure 1, 2). The great saphenous vein (GSV) was ligated in the proximal part and its main trunk was removed with all thrombosed veins with a Nabatoff probe. “Tumor-like”, enlarged veins close to the popliteal fossa and under the ankle were excised independently via additional incisions with safe tissue margins, and the patient recovered well from the surgery. Due to their unusual appearance, we performed a pathological examination. The surgery was uneventful, and the patient left hospital on the second postoperative day. The final histopathologic diagnosis was LMS (Figure 3). Thus, the patient was referred to computed tomography and magnetic resonance imaging for detection of possible metastasis. The exams revealed no metastatic
However, the patient was managed by an oncologic surgeon for further monitoring. The last follow-up was done 1 year after surgery.

Discussion

The first description of venous LMS was presented by Perl in 1871. This malignancy originates from the artery smooth muscle layer [11]. The most frequent site for LMS is the retroperitoneum, accounting for up to 50% of all cases, 50% of reported vascular LMS originates from the vena cava inferior and 15% from the iliac veins. The localization in veins is 5 times more common than in arteries [12]. In the legs, the GSV is the most common site of vascular LMS. Most cases are found proximal to the knee near the femoral bundle, and it is more common in females. In 2016, Cangiano et al reported a total of 39 documented cases of primary vascular LMS of the GSV in the literature. After the publication of this work, the total number grew to 42 cases. The most common presentation involves a slow-growing, non-painful, palpable mass along the course of a major vessel in the legs. Primary vascular STS originating in superficial leg veins usually present as a painless mobile mass, but those arising from deeper veins tend to be fixed in the surrounding tissue. Vascular LMS is asymptomatic in most cases, thus it can be diagnosed late in the terminal stage of the disease when metastases have developed. Nonetheless, even 10% of vascular LMS develop metastases in the early stage of disease [13-15]. Similarly, our patient did not present with severe symptoms. The crucial moment in diagnosis was admission to the hospital prior to intervention. Unfortunately, the 2 months between the first surgical consultation and intervention proved crucial for LMS asymptomatic growth. The patient had developed alarming symptoms – pain, warmth and redness – scarcely 1 week before admission, and this asymptomatic course of the disease led to a considerable delay in diagnosis.

With an experienced operator, US is a non-invasive examination that can differentiate between simple varices of the leg and leiomyosarcoma. However, our patient had a recurrent SVT; the last was 2 months prior to surgery from a series performed in the previous years. All descriptions were very similar and suggested pure SVT. Such rapid tumor growth during 1 month suggests that the last US is sufficient for correct diagnosis.

Generally, LMS is treated by surgical excision of the lesion (tumor) with adequate margins, but metastatic cases are treated as incurable, and surgical excision is intended to decrease tumor dimensions and pain. This approach can prolong survival and improve quality of life. Although a multidisciplinary treatment is advised, the effectiveness of adjuvant radiotherapy, chemotherapy, and hormonal therapy is unknown [11].

In everyday practice, physicians should be alert when patients report such symptoms. Surgery is a first-line treatment for these patients, but they should also be referred to an oncologist. Hence, surgery is the treatment of choice, with close monitoring for recurrence and formation of metastases.

Conclusions

LMS of GSV is a very rare entity. Clinical presentation varies among patients and by localization of the tumor. Asymptomatic
cases are the most dangerous, and their prognosis for survival is the worst. The first signs of LMS can be detected by US. Additional imaging, such as CT or MRI, are used for staging the disease. Despite routine surgical practice in the classical treatment of leg varices, every additional finding requires histopathologic diagnosis and further therapy may be necessary. Despite many efforts and being aware of imperfections, surgical excision of LMS still continues to be the only effective method of treatment.

References:


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