GIANT LEIOMYOSARCOMA WITHOUT SYMPTOMS
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Abstract
Of all types of sarcomas, about 25% are leiomyosarcomas.
We report a case of a young woman aged 35 years with asymptomatic leiomyosarcoma, who came for an examination due to hypochromic anemia.
Due to the patient's age and the asymptomatic course of the disease, this case is specific and differs from previously published cases of giant LMS.

Key words: leiomyosarcoma, giant, asymptomatic

Introduction
Of all types of sarcomas, about 25% are leiomyosarcomas (LMS) [1].
LMS originates from smooth muscle cells that line small blood vessels, but can also be of uterine, gastrointestinal, or soft tissue origin [2,3].
The prognosis is poor, the survival rate is one of the lowest of all types of sarcomas. Women are affected more than men (2 : 1) and the disease most often occurs in the 5th to 6th decade of life. The most common localization of LMS is the retroperitoneum, which accounts for 12-69% of all localizations [4].
Vascular LMS originates from smooth muscle cells from the medial layer of blood vessels. These tumors most often occur in the veins, often arising from the smooth muscle cells of the inferior vena cava (IVC). About 400 papers on this topic have been published in the literature so far [5].
Like other LMSs, IVC LMS manifests in middle age, predominantly in women. Symptoms vary depending on the location of the tumor relative to IVC. LMS that occurs in the central segment of IVC, i.e. the infrahepatic or suprarenal part are more common, about 40% [6,7].
LMS that include the lower part of the IVC, i.e. the infrarenal segment is second in frequency, with a frequency of about 30% [8].

Case report
We report a case of a young woman aged 35 years, who came for an examination due to hypochromic anemia. Because of the disease, she was taking therapy for hypochromic anemia for several months. Laboratory findings were normal, Hb 10 g/l, serum Fe 13.5 µmol/l.
An ultrasonographic examination was performed, where a large tumor mass was detected in the retroperitoneum, heteroechoic, and on Doppler the mass was vascularized.
The liver and right kidney are compressed, and the other organs in the abdominal cavity were normal. In the following act, a contrast MSCT examination was performed A large tumor in the retroperitoneal cavity was seen, measuring 23 x 11 x 14 cm, with dominant heterodensity and necrotic zones, which in the postcontrast sequence were emphasized as hypodense areas, with scar lesions.
The mass had a compressive effect on the surrounding structures, so that the head of the pancreas was not well followed, and the right kidney was pushed down. The mass included IVC adventitia in the infrarenal segment in a length of about 3 cm. There was no heterogeneous filling defect in the venous phase, which would indicate the possible existence of a thrombus.
Due to the compressive effect and spread of the tumor in the superior direction, dilatation and tortuosity of the left renal vein were monitored, but without repercussion on renal function.
There was a clear demarcation of the tumor from the liver parenchyma. No metastases were found in the parenchymal organs, as well as in the lung parenchyma. An MSCT core biopsy was also performed; two cylinders of 22 mm material were taken. Histopathology confirmed retroperitoneal
LMS, which was also confirmed by a radiologist’s report. The patient underwent neoadjuvant chemotherapy, followed by a surgical treatment.

**Discussion**

Vascular IVC LMS are relatively slow-growing tumors that are typically attached to the venous wall and show extraluminal growth, while intraluminal tumor penetration is mainly manifested by thrombosis [9,10].

According to the IVC segment and because of the implications for surgical planning and prognosis, retroperitoneal LMS are classified into:

- Type 1 - which includes the suprahepatic segment (6-24%);
- Type 2 - includes the suprarenal segment (42-50%);
- Type 3 - covering the infrarenal segment (34-44%) (1,5).

In the diagnosis of these tumors the algorithm is generally well established, as in other malignant diseases: US, MDCT and MRI. On ultrasound, the retroperitoneal LMS looks like a lobulated solid mass. Irregular cystic spaces may appear simple or contain low-level echoes [11].

The solid mass is iso- to hyperechoic compared to the liver. Tumors can be homogeneous when they are smaller, but as their heterogeneity increases they become more and more dominant. Intravascular tumor growth leads to IVC dilatation [12]. On MSCT, retroperitoneal LMS is presented as a large lobulated mass [13].

Necrosis is characterized by irregular areas of hypoattenuation in 28-38% of cases. Calcifications are not specific for LMS [14,15].

Collateral blood vessels are a common finding, which is noticed in about 61% of cases. Metastases occur in half of the cases during the basic examination, while hematogenous in the lungs (59-65%) and liver (50-53%) are more common than lymphatic metastases (14-35%) [16].

In intravascular LMS, the findings are consistent with tumor thrombus. Typical tumor thrombus findings include dilatation of a blood vessel with heterogeneous amplification of the intraluminal lesion [17].

Giant retroperitoneal LMS is rarer; there is a small number of cases in the literature. Elderly patients, 6th to 7th decade, are most often affected and the anamnesis is always positive, depending on the location and origin of the tumor. Takeda et al. described a giant retroperitoneal LMS, with metastatic deposits in the liver and lungs [18].

Due to the patient's age and the asymptomatic course of the disease until diagnosis, this case is specific and differs from previously published cases of giant LMS.

The tumor includes the renal and infrarenal segment of IVC, type 3, it has extraluminal growth, and upon adventitious IVC its length is about 3 cm. MSCT phlebography showed no intraluminal tumor penetration into the IVC or the presence of a thrombus.

MSCT is more favorable than MRI for the following reasons: better defines the relationship of the primary tumor to the environment, the relationship and assessment of soft tissue infiltrations and neurovascular structures, which allows a better operative plan and approach, the presence of peritoneal or liver metastases, and most importantly allows a complete appearance of the primary tumor to determine the origin and degree of invasiveness [19].

In addition to standard MSCT protocols, MSCT or MRI phlebography may be performed to assess IVC venous basin.

In retroperitoneal IVC LMS type 1, and due to the assessment of perihepatic structures, MRI with the liver protocol may be a complementary modality of imaging, especially when tumors involve the retrohepatic segment of IVC.

This modality is also useful for assessing the degree of liver infiltration. FDG-PET scan is not recommended in a routine diagnostic procedure [20].

Since IVCs are LMS of exophytic growth, CT core biopsy is the gold standard [21] in establishing the diagnosis, which was also made in our patient. The approach to resolving IVC LMS is multimodal, and treatment is surgical or in combination with neoadjuvant therapy [22-25].
Conclusion
Retroperitoneal IVC LMS in general, especially the giant type 3, is a highly aggressive tumor that occurs less frequently, but has an extremely poor prognosis.

Therefore, timely diagnosis is crucial for the most favorable outcome, with MSCT as a modality, which provides great possibilities in diagnosis. CT core biopsy is the gold standard to the final pathohistology, and a multimodal approach to the final decision to enable a longer and better patient life.

Figure 1. Giant retroperitoneal IVC LMS, coronal view
Figure 2. Axial section, cranial segment IVC LMS - impression of liver and spleen

Figure 3. Extraluminal growth of IVC LMS, right kidney is compressed and pushed down


