JMS 2020;3(3):43-48
UDC:616-006.36

GIANT LEIOMYOSARCOMA WITHOUT SYMPTOMS

Antuleska-Belceska Gordana, Bundovska-Kocev S
[1]University Institute of Radiology, Faculty of Medicine, University „ Ss. Cyril and Methodius", Skopje, R.of North Macedonia

Abstract

Of all types of sarcomas, about 25% are leiomyosarcomas.

We report a case of a young woman aged 35 years with asymptomatic leiomiosarcoma, 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: leiomiosarcoma, giant, asimptomatic

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 5^{th} to 6^{th} 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, 6^{th} to 7^{th} 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.

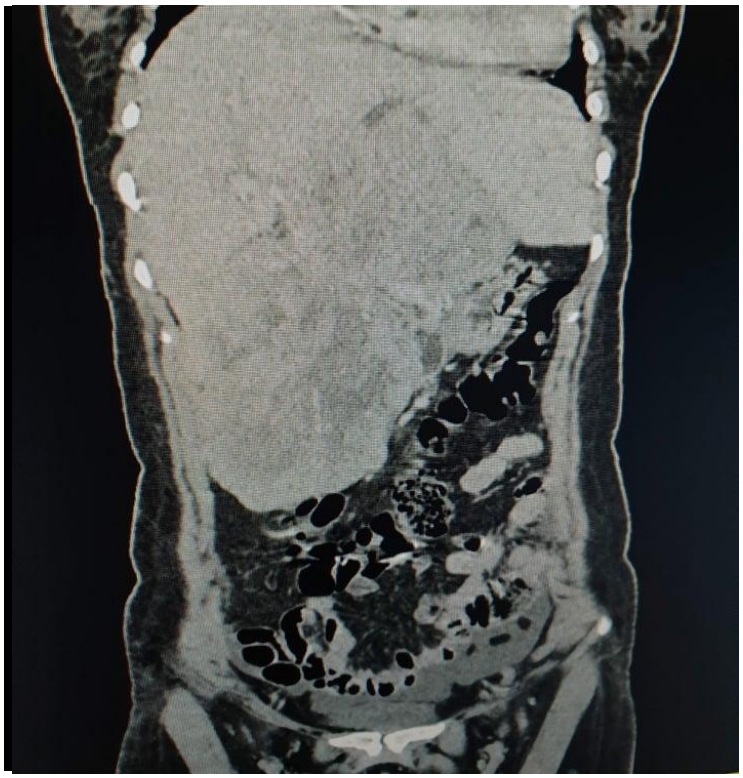

Figure 1. Giant retroperitoneal IVC LMS, coronal view

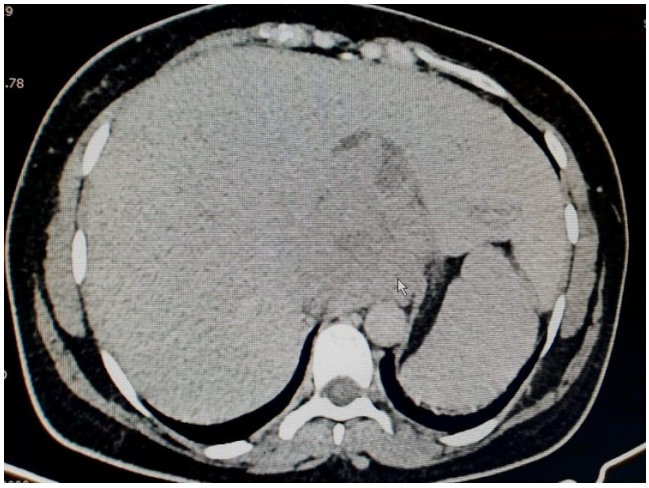

Figure 2. Axial section, cranial segment IVC LMS - impression of liver and spleen

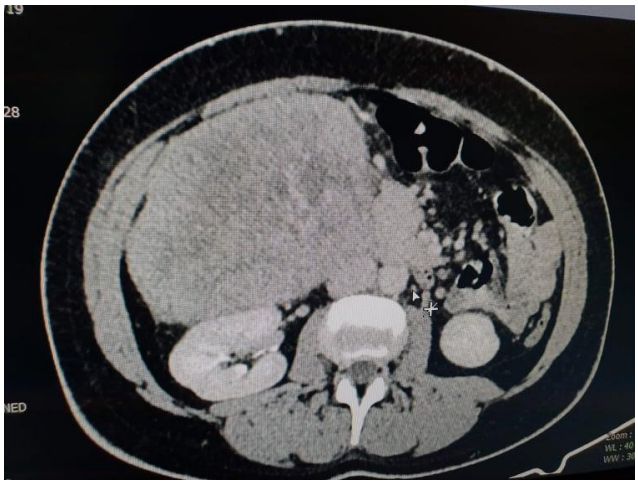

Figure 3. Extraluminal growth of IVC LMS, right kidney is compressed and pushed down

References

1. Mangla A, Yadav U.:Cancer,Leiomyosarcoma.,StatPearlsPublishing; 2020Mar 24. available from: https://www.ncbi.nlm.nih.gov/books/NBK551667/, PMID: 31869131
2. Potti A, Ganti AK, Tendulkar K, Sholes K, Chitajallu S, Koch M, Kargas S.: Determination of vascular endothelial growth factor (VEGF) overexpression in soft tissue sarcomas and the role of overexpression in leiomyosarcoma. J Cancer Res Clin Oncol. 2004 Jan;130(1):52-6. PMID: 14600832 DOI: 10.1007/s00432-003-0504-0
3. Messiou C, Morosi C.: Imaging in retroperitoneal soft tissue sarcoma, J Surg Oncol. 2018 Jan, 117(1): 25–32. PMID: 29193092
4. Rajiah P, Sinha R, Cuevas C, Dubinsky TJ, Bush WH Jr, Kolokythas O.: Imaging of uncommon retroperitoneal masses. RadioGraphics 2011;31(4):949–976.
5. Mann GN, Mann LV, Levine EA, Shen P.: Primary leiomyosarcoma of the inferior vena cava: a 2-institution analysis of outcomes. Surgery. 2012;151(2):261-267. doi: 10.1016/j.surg.2010.10.011.
6. Rajiah P, Sinha R, Cuevas C, Dubinsky TJ, Bush WH Jr, Kolokythas O.: Imaging of uncommon retroperitoneal masses, RadioGraphics 2011;31(4):949–976.
7. Cooley CL, Jagannathan JP, Kurra V et al.: Imaging features and metastatic pattern of non-IVC retroperitoneal leiomyosarcomas: are they different from IVC leiomyosarcomas?, J Comput Assist Tomogr 2014;38(5):687–692.
8. Ganeshalingam S, Rajeswaran G, Jones RL, Thway K, Moskovic E.: Leiomyosarcomas of the inferior vena cava: diagnostic features on cross-sectional imaging. Clin Radiol 2011;66(1):50–56.
9. Wiseman JT. Grignol V.: Location, Location, Location: Approaches to Retroperitoneal Vascular Leiomyosarcoma, Sarcoma Oncology Journal, January 15, 2020, 34(1)
10. .Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F.: WHO classification of tumours of soft tissue and bone. Lyon, France: IARC, 2013. Google Scholar
11. Messiou C. Morosi C.: Imaging in retroperitoneal soft tissue sarcoma, J Surg Oncol. 2018 Jan 1; 117(1): 25–32 doi: 10.1002/jso.24891
12. Bathan AJ, Constantinidou A, Pollack SM, Jones RL.: Diagnosis, prognosis and management of leiomyosarcoma: recognition of anatomic variants, Curr Opin Oncol 2013;25(4):384389
13. Ganeshalingam S, Rajeswaran G, Jones RL, Thway K, Moskovic E. Leiomyosarcomas of the inferior vena cava: diagnostic features on cross-sectional imaging. Clin Radiol. 2011 Jan;66(1):50-6. doi: 10.1016/j.crad.2010.08.004
    Hartman DS, Hayes WS, Choyke PL, Tibbetts GP.: From the archives of the AFIP: leiomyosarcoma of the retroperitoneum and inferior vena cava—radiologic-pathologic correlation. RadioGraphics 1992;12(6):1203–1220.
14. Demetri GD, Baker LH, Beech D, et al.: Soft tissue sarcoma clinical practice guidelines in oncology. J Natl Compr Canc Netw. 2005;3(2):158-194.
15. Mateo F,VallejoM, Reinado DR et al: Giant retroperitoneal leiomyosarcoma. Multiorgan block removal.International Journal of Surgery Case Reports Volume 5, Issue 12, 2014**:**1050-1053
16. Demetri GD, Baker LH, Beech D, et al: National Comprehensive Cancer Network. Soft tissue sarcoma clinical practice guidelines in oncology. J Natl Compr Canc Netw. 2005;3(2):158-194.
17. Takeda T, Asaoka D, Fukumura Y, Watanabe S: Asymptomatic giant retroperitoneal mass detected at a medical checkup. Clin Case Rep. 2017 Oct 16;5(12):2148-2150. doi: 10.1002/ccr3.1231.
18. Emily M, Zhen J,Wang Antonio C et al: Can CT Features Differentiate Between Inferior Vena Cava Leiomyosarcomas and Primary Retroperitoneal Masses?, AJR:200, January 2013, 205–209 DOI:10.2214/AJR.11.7476
19. Nishino M, Katsumi H, Manabu M et al.: Primary Retroperitoneal Neoplasms: CT and MR Imaging Findings with Anatomic and Pathologic Diagnostic Clues, Radiographics 23, Jan 1, 2003,1: 45-57. doi:10.1148/rg.231025037

20. Berger-Richardson D, Swallow CJ.: Needle tract seeding after percutaneous biopsy of sarcoma: risk/benefit considerations. Cancer. 2017;123(4):560-567. doi: 10.1002/cncr.30370.
21. Joon Hyuk Choi , Jae Y Ro :Retroperitoneal Sarcomas: An Update on the Diagnostic Pathology Approach. Clin Case Rep. 2017 Oct 16;5(12):2148-2150. doi: 10.1002/ccr3.1231
22. Jamie M, Darcy JW: Retroperitoneal Leiomyosarcoma From the Radiologic Pathology Archives, RadioGraphics,Vol. 38, No. 5 Sep 12 2018 doi.org/10.1148/rg.2018180006
23. Mullinax JE, Zager JS, Gonzalez RJ.: Current diagnosis and management of retroperitoneal sarcoma. Cancer Contr 2011; 18(3):177–187
24. Siva Kumar S, Ranjan RV, Nikithamani A et al.: Anaesthetic management of a case of large retroperitoneal leiomyosarcoma, Anesthesiol Case Rep. 2018;1(2):27-29.