CASE REPORT

Annals of Agricultural and Environmental Medicine
www.aaem.pl

Leiomyosarcoma of the right renal vein with tumour thrombus extending to the right atrium – Case Report and literature review

Michał Godzisz[1,A-F], Andriy Yadlos[1,A-E], Katarzyna Szklener[2,C,E-F], Iga Kuliniec[1,B,E], Damian Sudoł[1,C,E], Przemysław Mitura[1,B,E-F], Krzysztof Bar[1,E-F]

[1] *Department of Urology and Urologic Oncology, Medical University, Lublin, Poland*
[2] *Department of Clinical Oncology and Chemotherapy, Medical University, Lublin, Poland*
A – Research concept and design, B – Collection and/or assembly of data, C – Data analysis and interpretation, D – Writing the article, E – Critical revision of the article, F – Final approval of the article

Godzisz M, Yadlos A, Szklener K, Kuliniec I, Sudoł D, Mitura P, Bar K. Leiomyosarcoma of the Right Renal Vein with Tumour Thrombus Extending to the Right Atrium – Case Report. Ann Agric Environ Med. doi: 10.26444/aaem/203784

Abstract

Leiomyosarcoma (LMS) is a mesenchymal neoplasm accounting for 20% of all soft tissue sarcomas (STS), with a particularly rare localization in the muscular layer of the venous vessels. The case report presents a patient with non-metastatic LMS of the right renal vein, spreading intraluminally, and reaching the right atrium of the heart, supplemented with a review of the state of knowledge. The patient underwent radical treatment, including radical nephrectomy with *en bloc* thrombectomy under extracorporeal circulation without circulatory arrest and deep hypothermia. Histopathological examination diagnosed LMS and negative surgical margins. A computed tomography (CT) scan performed on the 45th postoperative day detected no early recurrence or metastatic features. The world literature describes 78 cases of renal vein (RV) LMS. Surgical treatment has the greatest overall survival (OS) impact. Adjuvant systemic treatment and radiotherapy do not improve OS outcomes, having only palliative significance.

Key words

nephrectomy, review, thrombectomy, case report, leiomyosarcoma, renal vein, tumour thrombus

INTRODUCTION

LMS originating from a venous vessel is an extremely rare tumour which can occur at any anatomical site. The three main locations are limbs, retroperitoneum, and uterus. Venous LMS most often originates from the inferior vena cava (IVC) [1]. LMSs outside IVC are exceptionally rare and reported in the renal vein (RV), great saphenous, pulmonary, and femoral veins [2]. To date, 78 cases of LMS of the renal vein have been published. RV LMS is classified by growth pattern into extravascular, intravascular, and both extravascular and intravascular types.

CASE REPORT

A 61-year-old female patient was admitted to the Emergency Department (ED) due to shortness of breath, cough, lower extremity oedema, and high blood pressure. Symptoms persisted for two weeks. Urgent echocardiography (Fig. 1) showed tumour tissue in the right atrium of the heart and IVC, as well as suspected pulmonary embolism. In ED, angiography CT of the pulmonary arteries and non-contrast computed tomography (NCCT) of the abdomen and pelvis were performed. A pulmonary embolism was ruled out. Imaging revealed a tumour thrombus in IVC, extending from the right kidney into the right atrium (Fig. 2).

✉ Address for correspondence: Michał Godzisz, Department of Urology and Urologic Oncology, Medical University, Jaczewskiego 8, 20-954 Lublin, Poland
E-mail: michal.godzisz92@gmail.com

Received: 05.12.2024; accepted: 07.04.2025; first published: 16.04.2025

The patient was admitted to the Department of Urology and Urologic Oncology. During hospitalisation, abdominal and pelvic MRI (Fig. 3), cardiac CT, and abdominal and pelvic CT with contrast were performed. A tumour was found originating from the right kidney hilum with a tumour thrombus extending into the right atrium. There were no obvious features of metastatic disease. MRI showed a tumour of the renal hilum region (dimensions 5cm x 6cm x 5cm) with a thrombus extending into IVC (10cm) and the right atrium (3cm) (Fig. 3–4).

The case of a tumour thrombus reaching the right atrium require cooperation with cardiothoracic surgeons. The

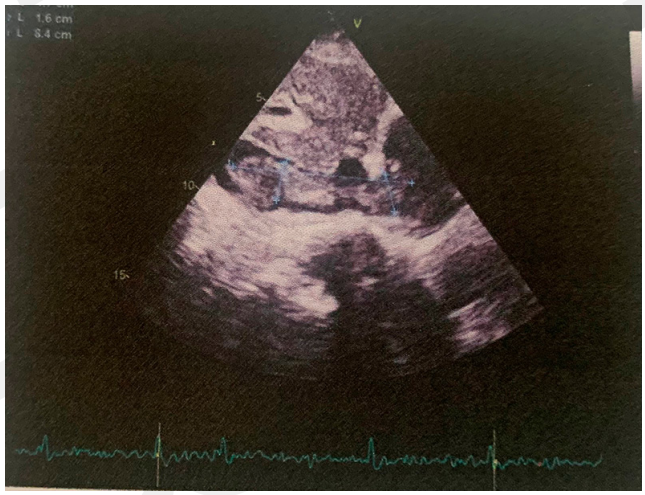

Figure 1. Echocardiography performed at pre-hospital diagnosis, visible thrombus in VC

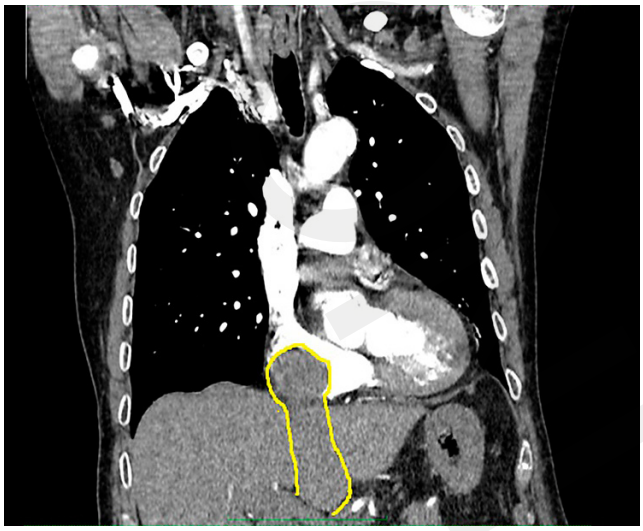

Figure 2. CT angiography of the pulmonary arteries with visible right atrial tumour thrombus (yellow contour)

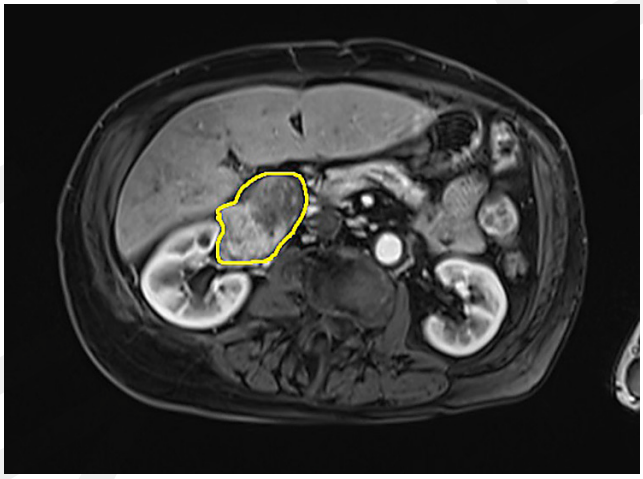

Figure 3. MRI image of the IVC filling thrombus (yellow contour) originating from the right renal vein

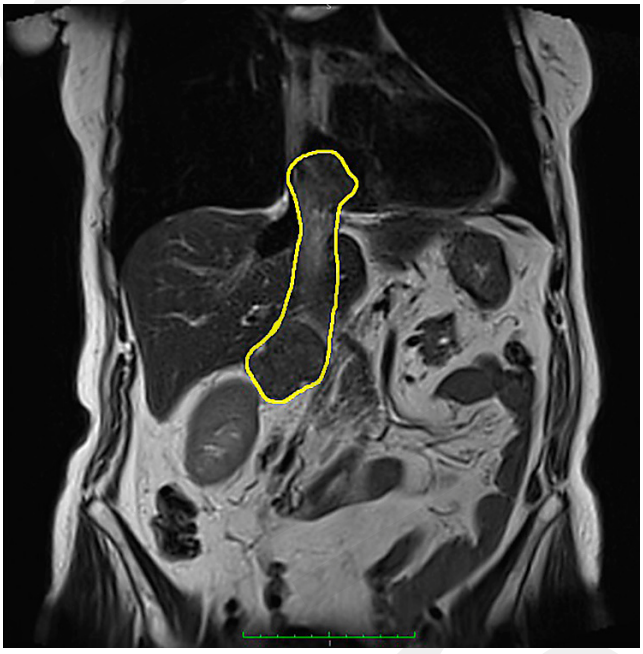

Figure 4. MRI image, the thrombus (yellow contour) extends into the right atrium

patient was qualified for a right-sided nephrectomy with cavotomy, combined with simultaneous sternotomy with a right atriotomy. In the facility where the patient was treated, atriotomy and thrombus manipulation are performed without cardiac arrest and deep hypothermia, which is beneficial for the patient.

Laparotomy was performed with a right hemi-chevron incision. The retroperitoneal space was opened through Toldt's line and the ascending colon with hepatic flexure was mobilized and moved medially. The Kocher's manoeuver provided access to the inferior IVC which was dissected distally inferior to the left renal vein orifice, and proximally to the level of hepatic veins, which involved clamping short hepatic veins (Fig. 5). Lumbar veins on the posterior aspect of IVC were ligated and cut.

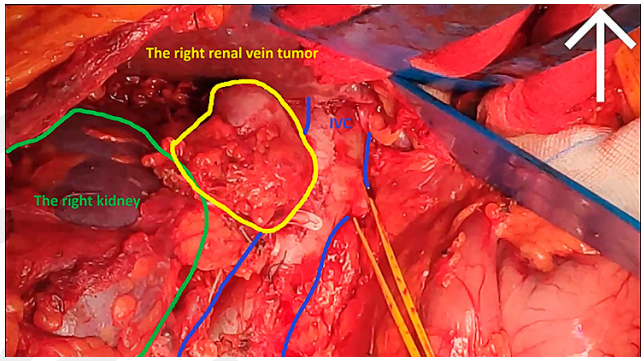

Figure 5. Intraoperative image of right retroperitoneal space with visible dissected IVC (blue line), right kidney (green line), and the right renal vein tumour (yellow line). A yellow rubber band marks the left renal vein

Next, the right kidney with the adrenal gland was mobilized outside the fat capsule. The right renal artery and ureter were dissected, clamped with Hem-o-lock clips, and severed. The hepatoduodenal ligament was mobilized and marked to perform the Pringle manoeuver later. At this stage, the cardiothoracic team performed a sternotomy and initiated extracorporeal circulation, incorporating suction from the operative field into the circuit. When the two surgical teams were ready, the cavotomy and right atriotomy were performed and the thrombus successfully pushed down from the right atrium and dissected from the IVC endothelium. The next step included resecting IVC surrounding the ostium with a safe margin of macroscopically healthy tissue. The right kidney was removed *en bloc* with the thrombus. After the removal of the tumour thrombus from IVC hepatic veins ostia were no longer blocked, but intensive bleeding from the hepatic veins occurred. This made IVC reconstruction difficult and Pringle manoeuver (temporary clamping of the haepatoduodenal ligament, which carries the blood supply to the liver) had to be performed. Such bleeding is expected and the common result of complex vascular surgery involving IVC at this level, and is therefore incorporated into the surgical plan. Due to the immediate suction of the blood back into the circuit patients outcome was not affected. IVC was reconstructed with the 5.0 polypropylene vascular suture, with no significant lumen narrowing (Fig. 6).

Immediately after surgery, the patient was transferred to the Cardiothoracic Intensive Care Unit, where she remained for five days, until the pleural drainage was removed, and she was transferred to the Urology Department. In the post-operative period, due to delusional and delirious

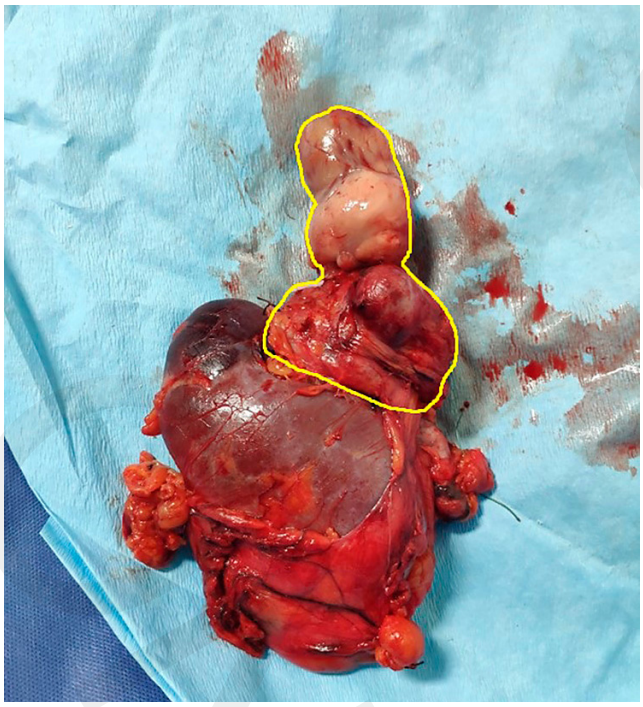

Figure 6. The right kidney with the renal vein tumour and the tumour thrombus (yellow contour) removed *en bloc*

disorders, the patient was consulted by a neurologist and underwent cerebral imaging and carotid Doppler ultrasound; morphological changes were excluded and the symptoms resolved spontaneously. The patient also developed atrial fibrillation which was successfully treated with electrical cardioversion (ECV). On post-operative day 18, the patient was discharged home in good general condition. CT was performed on the 45th post-operative day with no signs of local recurrence or metastasis.

Histopathology of the tumour showed renal vein LMS G2 (FNCLCC), vimentin (+), desmin (+), Smooth Muscle Actin (SMA; +), and caldesmon (+).

DISCUSSION

LMS of the renal vein is an extremely rare tumour of the retroperitoneal space. So far, the literature describes 78 cases of cancer in this localization. 67 cases up until 2017 [3] and 11 cases from 2017 to date [4–14]. In 11 cases, the cancer spread in the IVC lumen and formed a tumour thrombus [3,5,6]. In only one case, the disease reached the right atrium, but the patient was not eligible for surgical treatment [6]. Occasionally, the clinical presentation mimics renal cell carcinoma (RCC) [4]. In the presented case, the LMS closely resembled RCC, presenting with a tumour thrombus in the IVC.

The most important prognostic factors for overall survival (OS) are surgical margins and the absence of metastasis at the time of diagnosis, and for the local recurrence free survival (LRFS) prognostic factors are intraluminal extension and the degree of malignancy according to the FNCLCC scale [1,3].

Surgical treatment. The lack of effective adjuvant systemic treatment and radiotherapy limits the therapeutic options to radical resection [15,16]. According to the Gage et al. analysis, 5-year OS in patients with non-IVC LMS is 32%, with an incidence peak between 60–69 years of age. Females were affected more often. 95% of patients in this group were treated surgically, and 51% received adjuvant treatment (RT and chemotherapy) [2]. Hollenbeck et al. analyzed the results of 25 patients with IVC LMS [17]. After radical resection, 3-year and 5-year OS were 76% and 33%, respectively. Patients with non-radical resection did not survive 3 years. In the Hines group, the 5-year OS of patients with IVC LMS was 53% after surgery and chemoradiotherapy [18]. IVC LMSs frequently metastasize to the liver and lungs, despite aggressive multimodality treatment. Serrano et al. reported that 5-year OS is 33% in general and 68% after radical treatment with negative surgical margins [1], consistent with the results of the presented case report.

The longest observation of patients with IVC LMS was provided by Laskin et al. He reported 5-year and 10-year OS of 50% and 22%, respectively, in patients after radical resection with negative surgical margins. Moreover, he stated that IVC LMS intraluminal spread of the tumour suprahepatic or into the right atrium is correlated with an OS of less than 2 years [19].

It is important to emphasize that all the above studies were underpowered because of small groups of patients. Moreover, the data pertain to IVC LMS rather than RV LMS, and while these tumours share similar histology, directly extrapolating the results is problematic. Nonetheless, given the rarity of RV LMS, the available information has to be relied on.

The surgical treatment with negative surgical margins remains a challenge due to the location of the tumour at great vessels and critical vital structures, e.g. kidney, IVC with its tributaries, aorta, right atrium, and hepatic vessels. Radical resection is the only treatment option for LMS confined to an organ or anatomical location which provides a chance for full recovery [17].

Non-surgical treatment. Neoadjuvant radiotherapy, compared to primary surgery, has no recurrence-free survival (RFS) benefit [15]. The role of radio- and chemotherapy in LMS treatment is limited. Similarly, in 2012, Woll et al. performed a large, randomised trial under the aegis of EORTC, that showed no benefit of adjuvant chemotherapy in the form of doxorubicin and isofosfamide after radical resection of STS in improving RFS and overall survival (OS) [16].

LMS is one of the most aggressive STS cancers. According to the Surveillance, Epidemiology and End Results (SEER) database about 16% of patients present with metastatic disease at diagnosis, and 50% of patients develop metastasis during the course of the disease [20]. Due to the LMS tendency for haematogenous spread, systemic therapy is crucial for patients with metastases [21]. In 2007, Dileo et al. presented the results of their phase II trial on gemcitabine and vinorelbine combination chemotherapy for patients with unresectable or metastatic STS. They concluded that there were low effects of systemic treatment on improving OS, and clinical benefit was observed only in 25% of patients [22].

Chemotherapy is a palliative treatment reserved for patients with unresectable LMS and metastatic disease, directed at reducing symptoms, reducing tumour masses, and improving the quality of life. Anthracycline chemotherapy is the main systemic treatment for metastatic LMS [1,21], while targeted therapy and immunotherapy have no therapeutic effect [23].

This case report and review of existing literature emphasize the critical necessity for pioneering research aimed at creating more effective systemic therapies, given the significant risk of haematogenous spread and the unfavourable prognosis associated with metastatic leiomyosarcoma (LMS). Enhancing our comprehension of oncogenesis and discovering new targeted treatments are vital for improving the outcomes for patients diagnosed with leiomyosarcoma.

SUMMARY

Leiomyosarcoma of the renal vein is a rare disease, therefore knowledge of diagnostic and therapeutic approaches is primarily derived from case reports and case reviews. The presented case illustrates a successful short time outcome following radical resection, which is crucial in preventing progression and metastatic spread. Radical surgical treatment is a major factor in improving overall survival in patients without metastatic disease. The literature emphasizes the aggressive nature of LMS and the limited efficacy of systemic treatments, such as adjuvant chemotherapy and radiotherapy. In this context, achieving negative surgical margins represents the best chance for prolonging survival and enhancing the quality of life of the patient.

REFERENCES

1. Serrano C, George S. Leiomyosarcoma. Hematol Oncol Clin North Am. 2013;27(5):957–974. https://doi.org/10.1016/j.hoc.2013.07.002
2. Gage MJ, Patel AV, Koenig KL, et al. Non-vena cava venous leiomyosarcomas: a review of the literature. Ann Surg Oncol. 2012;19(11):3368–3374. https://doi.org/10.1245/S10434-012-2379-2
3. Novak M, Perhavec A, Maturen KE, et al. Leiomyosarcoma of the renal vein: analysis of outcome and prognostic factors in the world case series of 67 patients. Radiol Oncol. 2016;51(1):56–64. https://doi.org/10.1515/RAON-2016-0051
4. Fekkar A, Elouazzani H, Jahid A, et al. Leiomyosarcoma of the Renal Vein Mimicking a Primitive Renal Cell Carcinoma: Case Report of an Unusual Presentation. Case Rep Pathol. 2021;2021:1–6. https://doi.org/10.1155/2021/6637533
5. Konno M, Osawa T, Hotta K, et al. Primary renal leiomyosarcoma with a tumour thrombus in the inferior vena cava. IJU Case Rep. 2021;5(1):66–69. https://doi.org/10.1002/IJU5.12396
6. Lindblad G, Prater S, Chaniotakis SE, et al. Inoperable Left Renal Vein Leiomyosarcoma Refractory to Chemotherapy Invades Inferior Vena Cava and Right Atrium: A Case Report. Cureus. 2021;13(2):e13182. https://doi.org/10.7759/CUREUS.13182
7. Manoj Kumar G, Nirmal KP. Leiomyosarcoma of renal vein: A case report. Urol Case Rep. 2020;31:101186. https://doi.org/10.1016/J.EUCR.2020.101186
8. Lee JY, Kim SS. Intravascular leiomyosarcoma in the left renal vein. J Belg Soc Radiol. 2019;103(1):56. https://doi.org/10.5334/JBSR.1911
9. Suneetha KP, Gudaganatti SB, Gayathri J. Rare Case Presentation of Leiomyosarcoma as IVC Thrombus. Indian J Surg Oncol. 2019;10(3):540–541. https://doi.org/10.1007/S13193-019-00939-0
10. Hafeez M. Leiomyosarcoma of Renal Vein with Bone Metastasis: The Radiological Findings. J Coll Physicians Surg Pak. 2019;29(9):902–903. https://doi.org/10.29271/JCPSP.2019.09.902
11. Sekito S, Nishikawa K, Kageyama T, et al. [LEIOMYOSARCOMA ARISING FROM THE RENAL VEIN]. Nihon Hinyokika Gakkai Zasshi. 2019;110(4):244–248. https://doi.org/10.5980/JPNJUROL.110.244
12. Vicente E, Quijano Y, Duran H, et al. First case of complete full robotic surgical resection of leiomyosarcoma of the right renal vein. Surg Endosc. 2018;32(2):1072. https://doi.org/10.1007/S00464-017-5738-Z/METRICS
13. Ojha S, Nilkanthe R, Valecha J, et al. Leiomyosarcoma of Renal Vein – A Rare Case Report. J Clin Diagn Res. 2017;11(4):ED03–ED04. https://doi.org/10.7860/JCDR/2017/24684.9632
14. Trandem K, Aghazadeh MA, Goh AC. Robot-assisted Laparoscopic Resection of Renal Vein Leiomyosarcoma. Urology. 2017;103:e1–e2. https://doi.org/10.1016/J.UROLOGY.2017.02.008
15. Bonvalot S, Gronchi A, Le Péchoux C, et al. Preoperative radiotherapy plus surgery versus surgery alone for patients with primary retroperitoneal sarcoma (EORTC-62092: STRASS): a multicentre, open-label, randomised, phase 3 trial. Lancet Oncol. 2020;21(10):1366–1377. https://doi.org/10.1016/S1470-2045(20)30446-0
16. Woll PJ, Reichardt P, Le Cesne A, et al. Adjuvant chemotherapy with doxorubicin, ifosfamide, and lenograstim for resected soft-tissue sarcoma (EORTC 62931): A multicentre randomised controlled trial. Lancet Oncol. 2012;13(10):1045–1054. https://doi.org/10.1016/S1470-2045(12)70346-7
17. Hollenbeck ST, Grobmyer SR, Kent KC, et al. Surgical treatment and outcomes of patients with primary inferior vena cava leiomyosarcoma. J Am Coll Surg. 2003;197(4):575–579. https://doi.org/10.1016/S1072-7515(03)00433-2
18. Hines OJ, Nelson S, Quinones-Baldrich WJ, et al. Leiomyosarcoma of the inferior vena cava: Prognosis and comparison with leiomyosarcoma of other anatomic sites. Cancer. 1999;85(5):1077–1083. https://doi.org/10.1002/(SICI)1097-0142(19990301)85:5<1077::AID-CNCR10>3.0.CO;2-0
19. Laskin WB, Fanburg-Smith JC, Burke AP, et al. Leiomyosarcoma of the inferior vena cava: clinicopathologic study of 40 cases. Am J Surg Pathol. 2010;34(6):873–881. https://doi.org/10.1097/PAS.0B013E3181DDF569
20. Surveillance, Epidemiology, and End Results (SEER) Program. SEER*Explorer: An interactive website for SEER cancer statistics. https://seer.cancer.gov/statistics-network/explorer/ (accessed 2024.12.01)
21. von Mehren M, Kane JM, Agulnik M, et al. Soft Tissue Sarcoma, Version 2.2022, NCCN Clinical Practice Guidelines in Oncology. J Natl Compr Canc Netw. 2022;20(7):815–833. https://doi.org/10.6004/JNCCN.2022.0035
22. Dileo P, Morgan JA, Zahrieh D, et al. Gemcitabine and vinorelbine combination chemotherapy for patients with advanced soft tissue sarcomas: results of a phase II trial. Cancer. 2007;109(9):1863–1869. https://doi.org/10.1002/CNCR.22609
23. Chibon F, Darbo E, Perot G. Leiomyosarcomas: whole genome sequencing for a whole biology characterization. Curr Opin Oncol. 2019;31(4):317–321. https://doi.org/10.1097/CCO.0000000000000550