Medicinska istraživanja 2024; 57(4):131-136 | DOI: 10.5937/medi57-50984

OPEN ACCESS

Medical Research | Published by Faculty of Medicine University of Belgrade

CASE REPORT

Primary leiomyosarcoma of the inferior vena cava – radical resection and vascular reconstruction

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Recived: 12 May 2023

Revised: 17 September 2024

Accepted: 24 September 2024

Check for updates

Funding information:
The authors received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Competing interests:
The authors have declared that no competing interests exist

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Summary

Introduction: Sarcomas are rare tumors that account for less than 1% of malignant tumors in adults. Primary leiomyosarcoma (LMS) of the inferior vena cava (VCI) is a very rare tumor with an incidence of <1/100,000 of all malignant diseases in adults. This paper presents the case of a woman with LMS of the VCI who underwent radical tumor resection with vascular reconstruction.

Case report: A 71-year-old woman went to the doctor because of painless hematuria. As part of the diagnostic evaluation, in addition to US and CT examination of the abdomen, CT angiography with 3D reconstruction was performed. The conclusion of the radiologist was that the described tumor mass first corresponds to a primary LMS of the VCI VCI that propagated laterally into the lumen of the left renal vein, while the right renal artery was infiltrated by the tumor. The patient was presented to the multidisciplinary team at the Institute for Oncology and Radiology of Serbia (IORS), which decided for operative treatment. The tumor was completely removed en bloc with the right kidney, right suprarenal gland and the confluence of the left renal vein into the VCI. The VCI was reconstructed with a graft with end-end anastomosis, while the left renal vein was implanted in the graft with end-side anastomosis.

Conclusion: LMS of the VCI is an extremely rare tumor. Surgical resection of the tumor and the involved blood vessel with negative resection margins is the only therapeutic option that improves survival. Very often, in addition to complicated vascular reconstructions, surgery also includes multivisceral resections, in order to achieve the best possible therapeutic effect

Key words: leiomyosarcoma, vena cava inferior, sarcomas, surgery

Cite this article as: Nikolić S, Petrović O, Kocić M, Babić A, Jokić V, Pejnović L, Vučić N, Gačić S, Rajačić L, Đurišić I. Primary leiomyosarcoma of the inferior vena cava – radical resection and vascular reconstruction – a case report; Medicinska istraživanja 2024; 57(4):131-136 DOI: 10.5937/medi57-50984

INTRODUCTION

Primary LMS of the VCI is a very rare tumor. Early diagnosis is usually difficult and the prognosis is poor, due to very often advanced stage. We present a case of a woman with primary LMS who underwent radical resection of the tumor, along with vascular reconstruction.

CASE REPORT

A 71 year- old female presented with painless haematuria.

Ultrasonography of the abdomen was performed and showed a thick wall cystic mass in the right kidney, as well as avascular mass outside of the kidney, 60x40mm in size. The renal vein was thrombosed, and proximally the VCI was filled with a tumor mass, with partial patency. What was described distally was stasis or thrombosis of the VCI – differential diagnosis was RCC of the right kidney.

Subsequently, CT of the abdomen and the pelvis was performed. It showed a tumor mass in the area of the VCI that extended from L2 to porta hepatis and which dislocated and compressed the VCI forward, with unclear demarcation of the posterior wall of the vein – differential diagnosis - a conglomerate of lymph nodes para-aortic on the right, 64x41mm in size. The suprahepatic portion of the VCI was dilated.

The patient was then presented to a multidisciplinary team at IORS, which made a decision for CT angiography of the VCI, in order to discuss surgery resection.

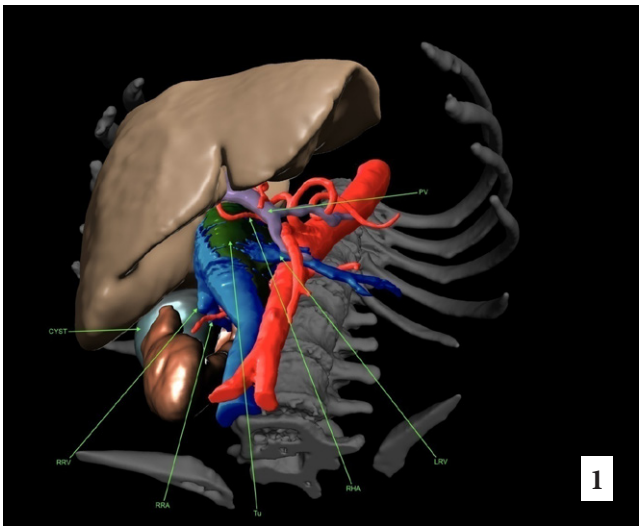
CT angiography was performed and showed a lobular, infiltrative, heterodense retroperitoneal mass 74x69x51mm, dominantly at the posteromedial wall of the VCI, irregularly narrowing the lumen. In the upper part it was reaching a caudal part of intrahepatic segment of the VCI, 4cm from the hepatic veins confluence. With the lower part, the mass was 85mm above common iliac veins. Lateral aspect of the tumor mass propagated into the lumen of the left renal vein. With its anterolateral aspect, it was very close to the right hepatic artery which was in this case originating from the upper mesenteric artery. The tumor made a compression on the main trunk of the portal vein, as well as suppression on the right renal vein, without any visible signs of intraluminal propagation. The right adrenal gland, and the right renal artery were infiltrated with the tumor.

According to the CT conclusion, described mass was in the first place LMS of the VCI **(Fig. 1-8)**.

There were no distant metastases at the moment of diagnosis.

The patient was once again presented to the multidisciplinary team, which decided on a surgical procedure. She was admitted to hospital on 21st January 2021, and the surgery was performed four days later. Preoperative urea and creatinine values were 9,7 mmol/L and 120 μmol/L.

We entered the abdomen through a bilateral subcostal laparotomy. Exploration confirmed the retrohepatic tumor, around 8x7cm in diameter. It was located about 3cm below the hepatic veins confluence, reaching the level of renal veins **(Fig. 9)**.



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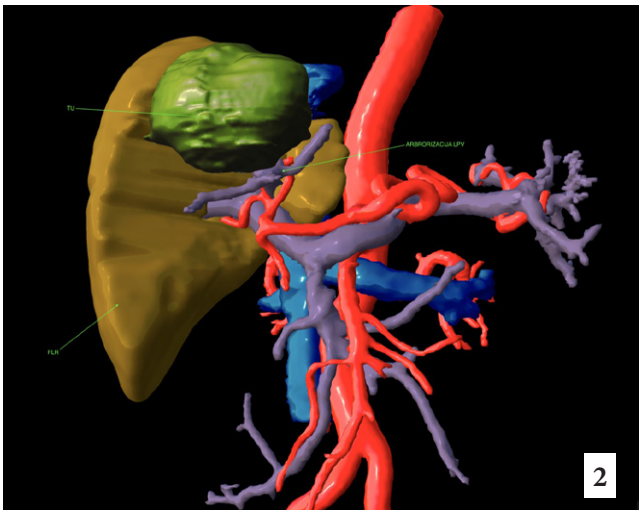

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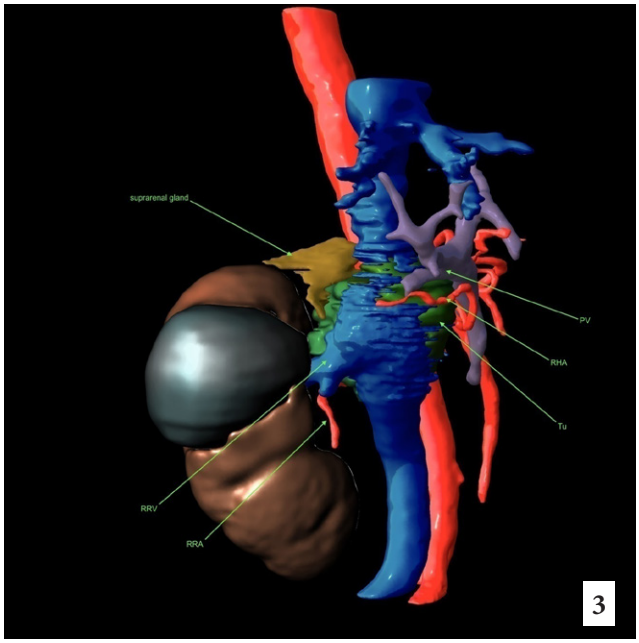

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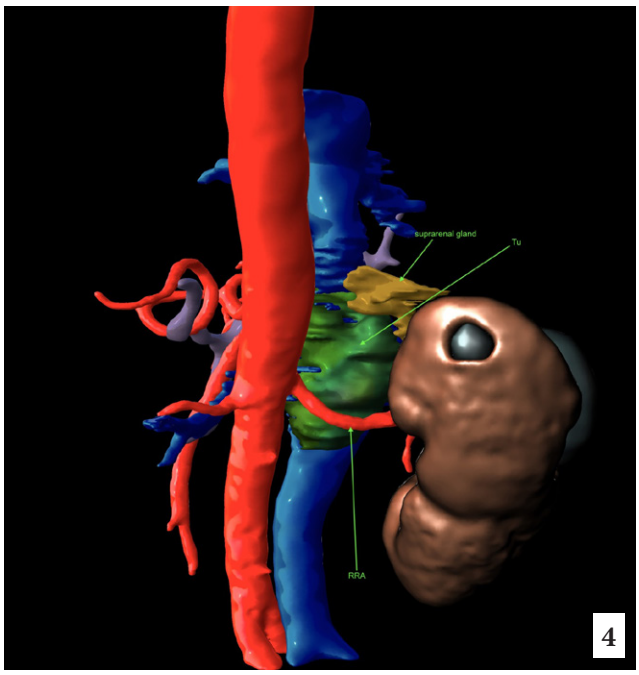

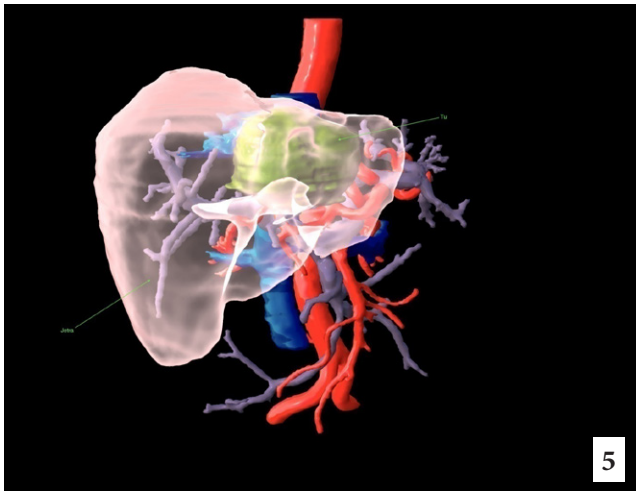

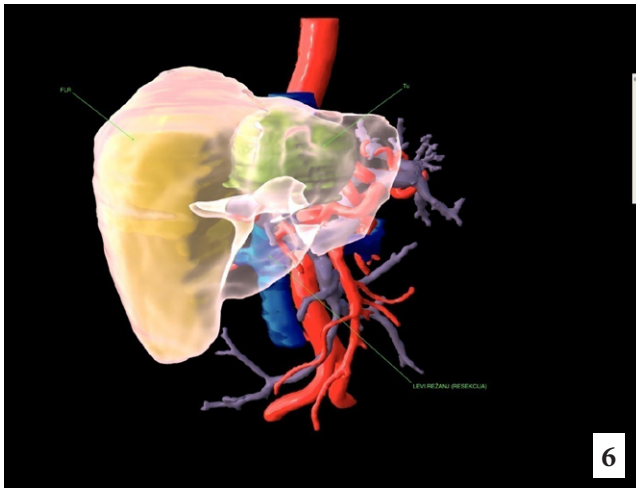

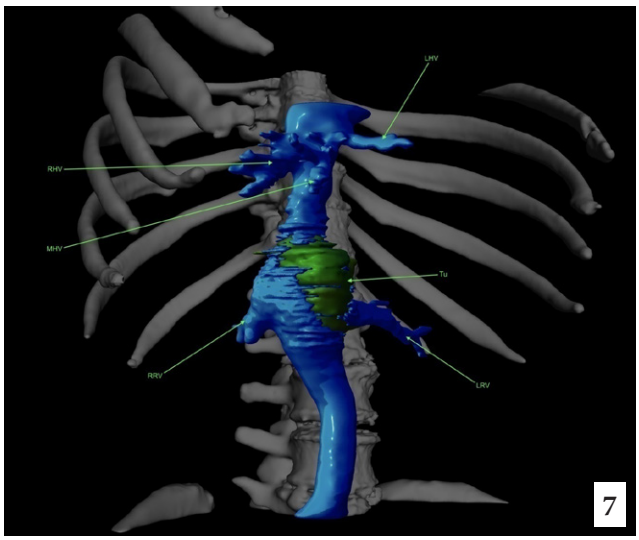

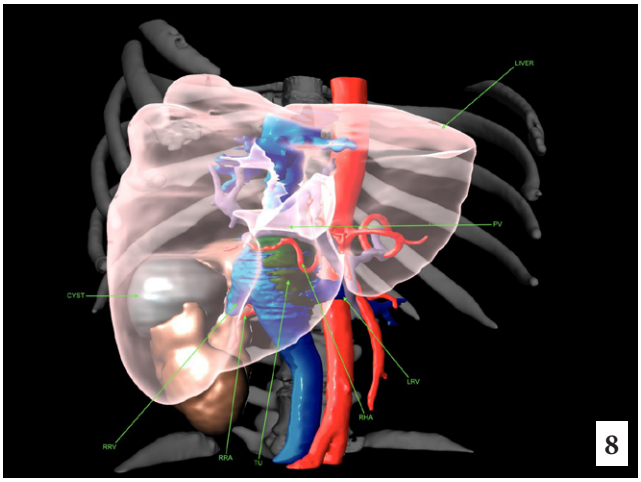

Fig. 1-8. CT angiography presenting the VCI tumor (green) and surrounding structures (PV – portal vein; LRV – left renal vein; RHA – right hepatic artery; RRA – right renal artery; RRV – right renal vein; LRV – left renal vein; LHV – left hepatic vein; MHV – middle hepatic vein; RHV – right hepatic vein).

There was an obvious thrombus identified in almost entire right renal vein as well as in the small portion of the left renal vein. The tumor had completely dislocated the renal vein confluences. The right kidney was enlarged and cystic. No other tumor changes were verified in the abdomen and pelvis by careful exploration. The right colon, duodenum and the head of the pancreas were mobilized and the access was made to the retroperitoneum and the VCI. After that we mobilized the liver, accessed also the retrohepatic portion of the VCI and made a proximal control. Several lumbar veins were ligated to free the VCI. We also made the distal control of the VCI, as well as the control of the left renal vein. The tumor with the involved right kidney, right suprarenal gland, and the left renal vein confluence was removed **(Fig. 10)**.

We used polyester (Dacron) graft for the reconstruction of the VCI, in end-to-end fashion. An end-to-side anastomosis was created with the left renal vein **(Fig. 11)**.

The overall operative time was three and a half hours. The patient was discharged on postoperative day eight, with postoperative urea and creatinine values of 8 mmol/L and 164 μmol/L.

Histopathological examination showed vascular leiomyosarcoma, G2, 75x75x60mm in size and R0 resection **(Fig. 12)**.

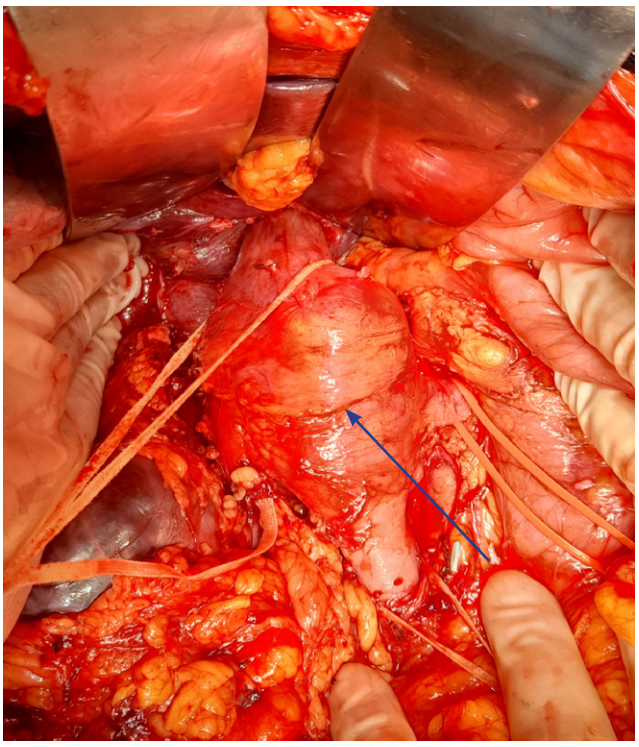

Fig. 9. Intraoperative finding of the VCI tumor (blue arrow).

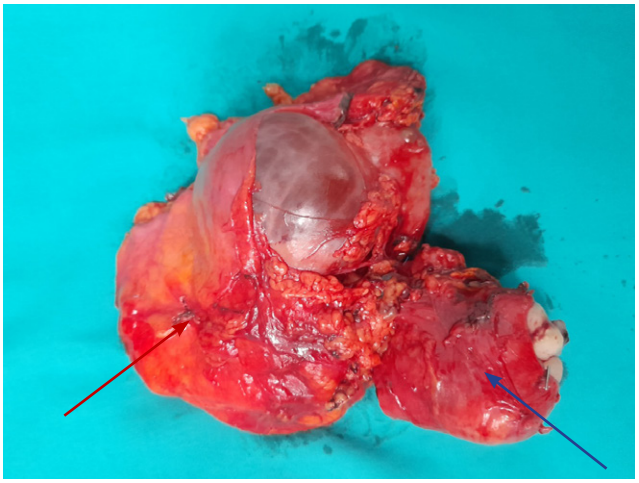

Fig. 10. VCI tumor (blue arrow) and the right kidney (red arrow) after removal

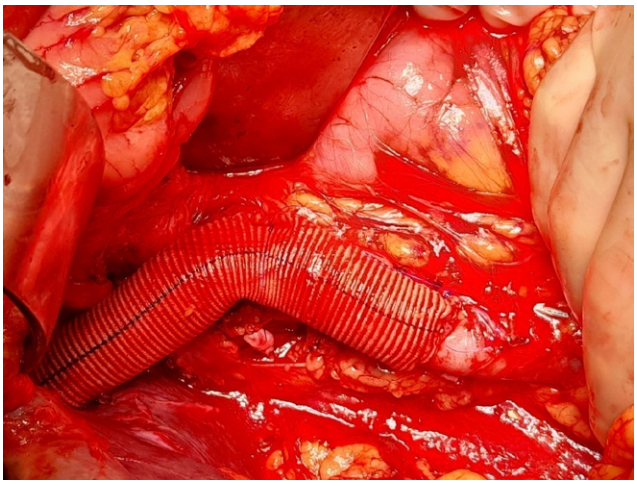

Fig. 11. Intraoperative photo after the reconstruction of the VCI with a graft

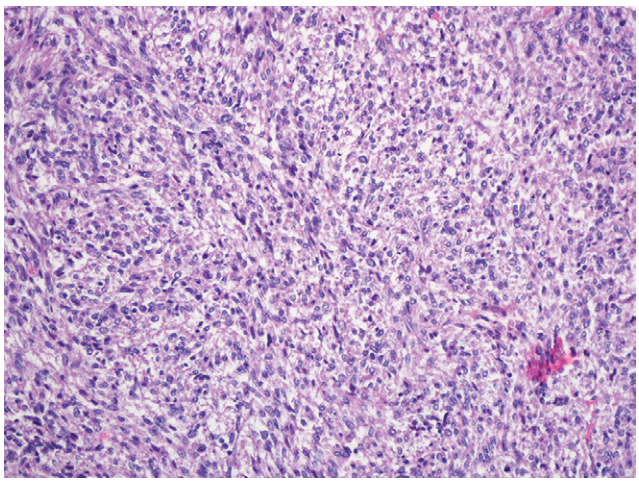

Fig. 12. Microscopic pathological finding.

Adjuvant therapy was not administered.

It has been three and a half years since the operation and the patient is still undergoing regular monthly check-ups.

DISCUSSION

Sarcomas are rare tumors accounting for less than 1% of cancer in adults and represent 15% of cancers in children. Majority of sarcomas are soft tissue tumors, including primary LMS which account for 0.5 to 1% of all malignant tumors. Most sarcomas are of unknown cause, but there are some known risk factors, such as radiation exposure, occupational chemical exposure, trauma, and chronic lymphedema (1).

VCI is the most common site of vascular LMS. Primary LMS of the VCI is a rare tumor accounting for around 0.5% of soft tissue sarcomas in adults. The incidence is <1/100 000 of all malignant diseases in adults. The prognosis is poor. Patients have intra- or extra-luminal tumor growth, very often with invasion of neighboring structures (2). In 1871, Perl et al. were the first to describe this tumor at an autopsy. It was a case of LMS of the VCI originating from the smooth muscles of a blood vessel. Melchior was the first to report a case of this tumor in 1928, which was treated surgically, by resection of the distal third of the VCI (3).

According to a recent pooled data analysis, fewer than 400 cases of LMS of the VCI have been reported, with most studies limited to single case reports (4). Patients are predominantly women (80%), mean age of 54 (3, 5).

The clinical behavior of most soft tissue sarcomas is determined by anatomic location, hystological subtype, grade and size. Presentation depends on the involved segment of the VCI. In relation to the localization of the malignant process, VCI is according to Kulaylat et al. divided into three segments:

1. Segment I or infrarenal segment (lower segment), involved in 36% of cases.

2. Segment II or inter- and supra- renal segment, from the hepatic veins to the renal veins (middle segment), affected in 44%.
3. Segment III or suprahepatic segment, from the right atrium to the hepatic veins, with potential intracardiac propagation (upper segment) (12).

The most common segment affected by the tumor is the segment between the renal and hepatic veins, which is also associated with the best prognosis, while the involvement of the upper segment of the VCI, intraluminal growth as well as potential obstruction of VCI are associated with poor prognosis (13).

The diagnosis is often made late, due to a long asymptomatic period, non-specific abdominal symptoms caused by tumor compression on surrounding structures. Very often patients present with abdominal pain and discomfort, as well as a palpable mass in the abdomen. In case of middle segment involvement, nephrotic syndrome or renal hypertension can occur. Almost a third of patients have lower extremity edema, due to obstruction of the VCI. Often there are also non-specific symptoms and signs such as nausea, vomiting or fever. Thrombotic mass can cause pulmonary embolism, tricuspid valve insufficiency, cardiac arrhythmias, as well as failure of the liver and kidneys due to "outflow" obstruction (10, 14, 15, 16). There can also exist symptoms due to metastatic disease.

Diagnostic imaging includes ultrasonography, CT and MRI. Chest CT should be performed at presentation and before any radical treatment, to evaluate for lung metastasis, knowing that distant metastases occur most often in the lungs, which 80% of the time occur within 2 to 3 years of the initial diagnosis (1). Other sites include the liver, bones and brain (17). Lymph node metastases in sarcomas are rare (less than 5%), but the incidence is higher in some subtypes, including angiosarcomas (1, 6).

Surgical resection of the tumor and affected vessel with negative margins is the only treatment shown to improve survival (7, 8, 9,10). Very often, along with complicated vascular reconstructions, surgery involves multivisceral resections, in order to achieve best therapeutical effect (11). Surgery is considered for patients without metastatic disease and with resectable primary tumor. Pre-operatively, high quality imaging is reviewed to determine the likely extent of resection, specifically including the need for potential en-bloc resection of adjacent organs. In cases where up-front surgical approach would expose the patient to excessive morbidity (such as bilateral nephrectomy, multi-visceral resection, or prohibitively high risk of positive margins), neoadjuvant chemotherapy and/or chemoradiotherapy is considered (18).

Overall 5-year survival rate for patients with all stages of soft tissue sarcoma is 50% to 60%. There is no much difference in LMS of the VCI, with the 5-year survival rate between 31% and 67% with R0 resection (1, 2).

CONCLUSION

LMS of the VCI is an extremely rare tumor entity. Surgical resection of the tumor and affected vessel with negative margins is the only treatment shown to improve survival. Very often, along with complicated vascular reconstructions, surgery involves multivisceral resections, in order to achieve the best therapeutical effect. Also, patients with LMS of the VCI should be treated in experienced centers by experienced surgeons.

Statements and Declarations

The patient's consent to the publication of the paper has been obtained.

The paper has never been published or concurrently submitted for publication to any other journal.

All authors who have met the authorship criteria have read and approved the manuscript.

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PRIMARNI LEJOMIOSARKOM DONJE ŠUPLJE VENE - RADIKALNA RESEKCIJA I VASKULARNA REKONSTRUKCIJA

Nikolić Srđan[1], Petrović Ognjen[1], Kocić Milan[1], Babić Anđela[1], Jokić Vladimir[1], Pejnović Luka[1], Vučić Nikola[1], Gačić Stefan[1], Rajačić Lila[1], Đurišić Igor[1]

Uvod: Sarkomi su retki tumori koji čine manje od 1% malignih tumora kod odraslih. Primarni lejomiosarkom donje šuplje vene je veoma redak tumor čija je incidencija <1/100 000 svih malignih bolesti kod odraslih. U ovom radu predstavljen je slučaj žene sa lejomiosarkomom donje šuplje vene kojoj je urađena radikalna resekcija tumora uz vaskularnu rekonstrukciju.

Prikaz bolesnika: Žena stara 71 godinu, javila se lekaru zbog bezbolne hematurije. U sklopu dijagnostičke evaluacije pored UZ i CT pregleda abdomena učinjena je i CT angiografija sa 3D rekonstrukcijom. Zaključak radiologa je bio da opisana tumorska masa najpre odgovara primarnom lejomiosarkomu donje šuplje vene koji se lateralno levo propagirao u lumen leve bubrežne vene, dok je desna bubrežna arterija bila infiltrisana tumorom. Pacijentkinja je prikazana konzilijumu na Institutu za onkologiju i radiologiju Srbije koji je doneo odluku o operativnom lečenju. Tumor je u celosti odstranjen u bloku sa desnim bubregom, desnom nadbubrežnom žlezdom i ušćem leve bubrežne vene u donju šuplju venu. Donja šuplja vena je rekonstruisana uz pomoć grafta termino-terminalnom anastomozom, dok je leva bubrežna vena implantirana u graft termino-lateralnom anastomozom.

Zaključak: Lejomiosarkom donje šuplje vene je ekstremno redak tumor. Hirurška resekcija tumora i zahvaćenog krvnog suda sa negativnim ivicama resekcije je jedina terapijska opcija koja poboljšava preživljavanje. Vrlo često uz komplikovane vaskularne rekonstrukcije, hirurgija uključuje i multivisceralne resekcije, kako bi se postigao najbolji mogući terapijski efekat.

Ključne reči: lejomiosarkom, donja šuplja vena, sarkomi, hirurgija

Primljen: 12.05.2024. I **Revizija:** 17.09.2024. I **Prihvaćen:** 24.09.2024.

Medicinska istraživanja 2024; 57(4):131-136