American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2022; 23: e938009
DOI: 10.12659/AJCR.938009

Received: 2022.08.04
Accepted: 2022.09.12
Available online: 2022.09.20
Published: 2022.10.27

Successful Right Hepatectomy for Recurrent Liver Tumor Originating from an Inferior Vena Cava Leiomyosarcoma: A Follow-Up Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABDEF 1 Atsushi Nanashima
BE 2 Hiroki Takamori
B 1 Naoya Imamura
F 3 Koji Furukawa
D 1 Masahide Hiyoshi
D 1 Takeomi Hamada
BD 1 Koichi Yano
B 1 Yuuki Tsuchimochi
EG 2 Toshiyuki Kamoto

1 Division of Hepato-Biliary-Pancreas Surgery, Department of Surgery, University of Miyazaki, Miyazaki, Japan
2 Department of Urology, University of Miyazaki, Miyazaki, Japan
3 Division of Cardiovascular Surgery, Department of Surgery, University of Miyazaki, Miyazaki, Japan

Corresponding Author: Atsushi Nanashima, e-mail: a_nanashima@med.miyazaki-u.ac.jp
Financial support: None declared
Conflict of interest: None declared

Patient: Female, 58-year-old
Final Diagnosis: Leiomyosarcoma
Symptoms: None
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Rare disease
Background: Leiomyosarcoma (LMS) of the inferior vena cava (IVC) is a rare malignancy, and complete resection may provide better patient survival. Hepatectomy for intrahepatic tumor recurrence has not been previously reported.
Case Report: A 58-year-old woman underwent resection of an IVC-LMS with en bloc nephrectomy, adrenalectomy, and retroperitoneal soft tissue resection without IVC reconstruction 3 years 8 months ago. Twenty-nine months after the primary operation, a solitary intrahepatic liver tumor was found adjacent to the right and middle hepatic veins during imaging follow-up. The patient was diagnosed with LMS recurrence. As her liver functional parameters permitted major hepatectomy, right hepatectomy combined with resection of the vena cava wall leaving a tumor-free margin and securing the confluence of the middle hepatic vein were successfully accomplished via an anterior approach, without adverse events. Intrahepatic metastasis of LMS invading the vena cava wall has been diagnosed historically. Her postoperative course was uneventful, and at 1-year follow-up after the second surgery, she was observed to have no tumor relapse without any adjuvant treatment. Previous reports have shown that IVC-LMS is often observed, and operative risk or prognosis is based on the extension of the LMS toward the hepatic veins or cardiac atrium.
Conclusions: Radical hepatectomy for recurrent IVC-LMS has not been previously reported, and our case experience revealed that a challenging surgical intervention resulting in complete tumor removal can provide good survival outcomes.

Keywords: Hepatectomy • Leiomyosarcoma • Neoplasm Recurrence, Local • Vena Cava, Inferior

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/938009

1787 — 5 12

This work is licensed under Creative Common Attribution-NonCommercial-NoDerivatives 4.0 International (CC BY-NC-ND 4.0)

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

Background

Leiomyosarcoma (LMS) is a rare malignant neoplasm primarily composed of cells exhibiting features of smooth muscle degeneration, and mainly occurs in the extremities, trunk wall, and body cavities. Retroperitoneal LMS is more common in women, in whom it often arises from the inferior vena cava (IVC), which originates from smooth muscle cells of the vascular walls. Only 5% of LMS cases directly arise from large blood vessels, primarily from the IVC [1,2]. Although LMS can only be cured by radical surgical resection, complete resection is difficult because of the expansive appearance of IVC-LMS at the initial diagnosis [1-3]. After removal of an IVC sarcoma, tumor recurrence is common because of hematogenous metastasis or local dissemination [4]. Control of recurrent sarcoma is also considered to be associated with patient survival. We previously reported a rare case of successful en bloc resection of IVC-LMS in the right kidney, adrenal gland, and retroperitoneal tissues [5]. Following this publication, the clinical course of intrahepatic recurrence treated surgically in the same patient is described below.

Case Report

A 58-year-old woman underwent en bloc IVC tumor resection that included the right kidney, adrenal gland, and surrounding retroperitoneal soft tissues via thoracoabdominal and retroperitoneal approaches, but the IVC was not reconstructed [5]. **Figure 1A** shows the resected specimens and tumor at that time, which appeared similar to the intraluminal tumor thrombus originating from the kidney. However, transected formalin-embedded tissue specimens showed tumor growth from the IVC wall with involvement of the surrounding organs (**Figure 1B**). The patient's postoperative course was uneventful, and she was followed-up by both urological and liver surgeons for 29 months without tumor recurrence, no clinical symptoms, and good general health status. The patient underwent imaging examination with ultrasonography every 3 months and enhanced computed tomography every 6 months as a follow-up schedule.

Physical examination revealed no abdominal findings, and the liver function was well preserved within the normal range. However, enhanced computed tomography in the venous phase showed a slightly enhanced solitary tumor measuring 2.5 cm in size in the axial and coronal views (**Figure 2A, 2B**). T1-weighted magnetic resonance imaging with gadoxetate sodium contrast medium also revealed a heterogeneous tumor (**Figure 2C**) in which ^{18}F-fluorodeoxyglucose positron emission tomography showed increased accumulation (**Figure 2D**). As this tumor had grown to 3 cm at the month follow-up examination, IVC-LMS recurrence in the liver or local recurrence at the IVC stump was diagnosed, and right hepatectomy with combined resection of the caval wall was scheduled.

The Department of Surgery policy at our institute requires that indocyanine green retention rate at 15 minutes (ICGR15) be determined preoperatively for the liver to be resected, and the estimated resected or remnant liver volume, excluding tumor volume (cm^3), is measured by computed tomography volumetry using Synapse Vincent Work Station (Fujifilm Medical Co., Tokyo, Japan). Essentially, in cases where the future remnant volume is less than 40%, even in a normal liver, preoperative portal vein embolization (PVE) is selected. Although ICGR15 was 2.1% and the comprehensive evaluation of liver function was Child-Pugh grade A, the estimated future remnant liver

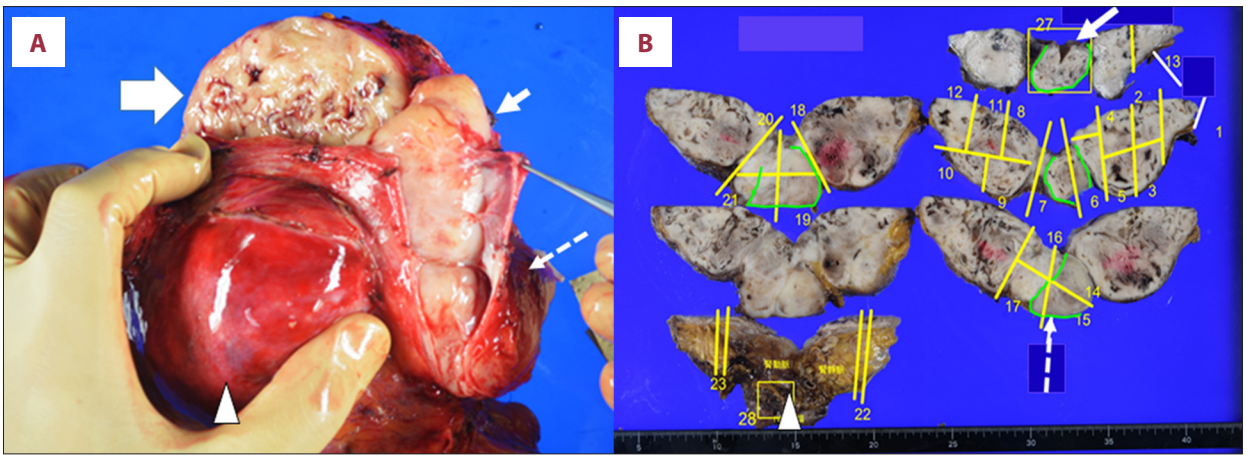

Figure 1. (**A**) Macroscopic findings of the primary inferior vena cava (IVC)-leiomyosarcoma (LMS) (thick arrows) by en bloc resection of the right kidney (arrowhead), adrenal (AD) gland and surrounding retroperitoneal soft tissue at the time of initial resection 4 years earlier [5]. The thin arrow indicates the tip of the LMS at the cut end of cranial-side IVC (dotted thin arrow). This image was not published in the previous report. (**B**) The transected formalin-embedded specimen. Green lines indicate the IVC wall with intraluminal filling by the LMS, which invaded the kidney and soft tissues.

This work is licensed under Creative Common Attribution-NonCommercial-NoDerivatives 4.0 International (CC BY-NC-ND 4.0)

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

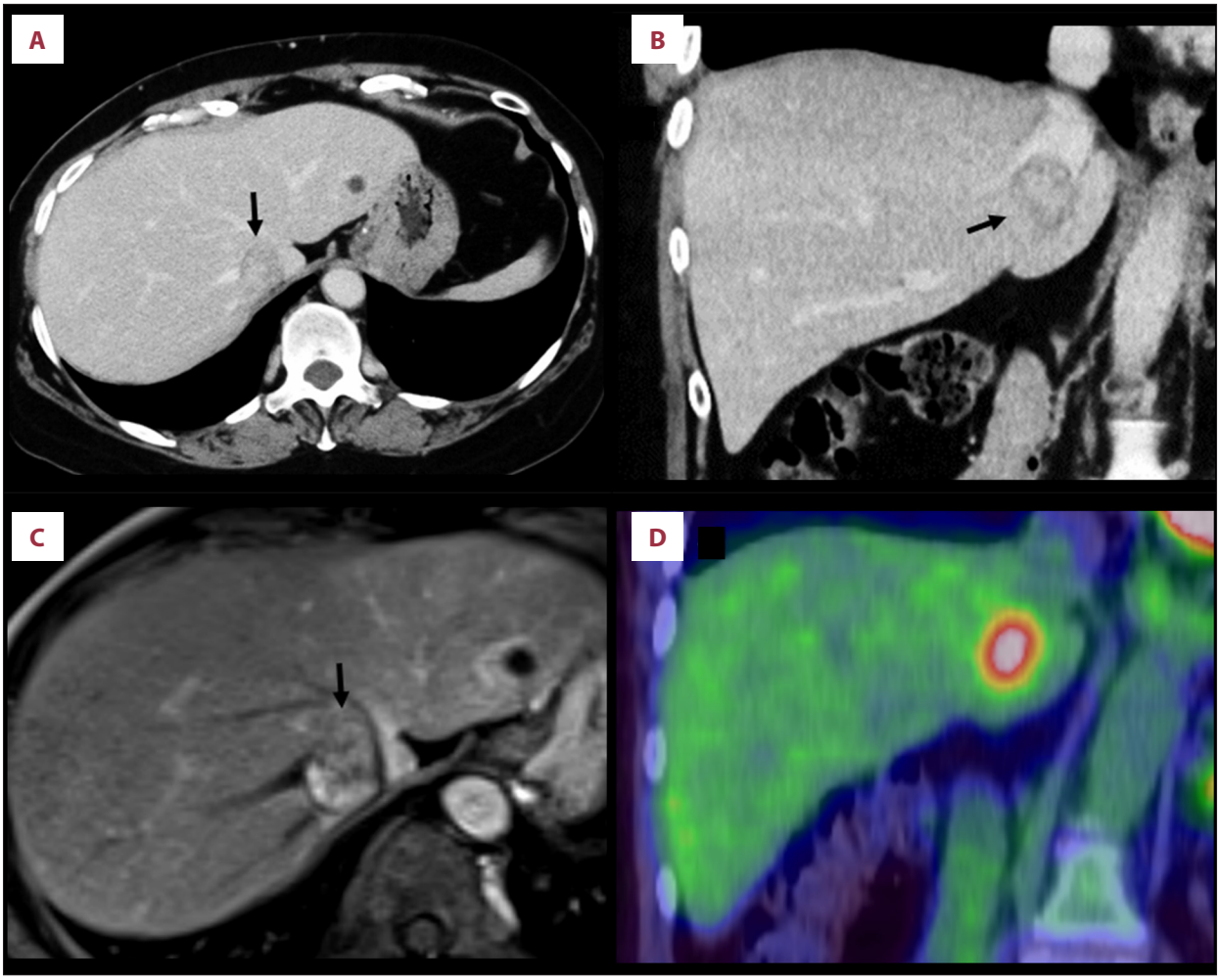

Figure 2. Image findings of tumor recurrence at 31 months after the first operation. (**A**) Axial and (**B**) coronal views of enhanced computed tomography images in the venous phase show a solitary, low-density, mass lesion 2.5 cm in size that was observed in the liver parenchyma adjacent to the right hepatic vein, middle hepatic vein, and the stump of the vena cava (black arrow). (**C**) Gadoxetate sodium-enhanced magnetic resonance image shows a partially hypervascular liver mass in the arterial phase (black arrow). (**D**) Positron emission tomography shows an accumulation of ^{18}F-fluorodeoxyglucose in the tumor lesion.

volume was 466 cm^3 (35% of the whole liver). Therefore, interventional radiologists performed preoperative transhepatic PVE. Substances used for embolization were 4 sheets of gelatin lipiodol, Serescue (Nihon-Kayaku Co., Tokyo, Japan), mixed in contrast media, and 2 permanent microcoils were subsequently placed in the right portal veins. The estimated future remnant liver volume was increased to 594 cm^3 (50% of the whole liver) was achieved on day 21 after PVE. ICGR15 was still well maintained at 3.0%, and the scheduled operation was performed at 3 months after the diagnosis of recurrence.

Surgical Procedures

Because we determined that mobilization within the retrohepatic space would be difficult due to the initial operation, parenchymal transection at the Rex-Cantlie plane via the anterior approach was selected. Laparotomy comprising an upper median plus transverse incision was performed (**Figure 3A**). No peritoneal dissemination or metastasis was observed in the abdominal cavity. Although the liver tumor did not infiltrate the liver surface, intraoperative ultrasonography showed that the tumor was compressing the confluence of the right hepatic vein (RHV) and IVC stump (**Figure 3B**). The confluence of the middle hepatic vein (MHV) and left hepatic vein could be dissected; however, the confluence of the RHV could not be dissected (**Figure 3C**). To prevent injury to the IVC, the pericardial IVC was taped to control hemostasis and prevent air embolism (**Figure 3D**). After the midline transection and division of the right Glissonian pedicle were completed, the tumor, including the RHV, could be encircled (**Figure 4A**), and the IVC could

This work is licensed under Creative Common Attribution-NonCommercial-NoDerivatives 4.0 International (CC BY-NC-ND 4.0)

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

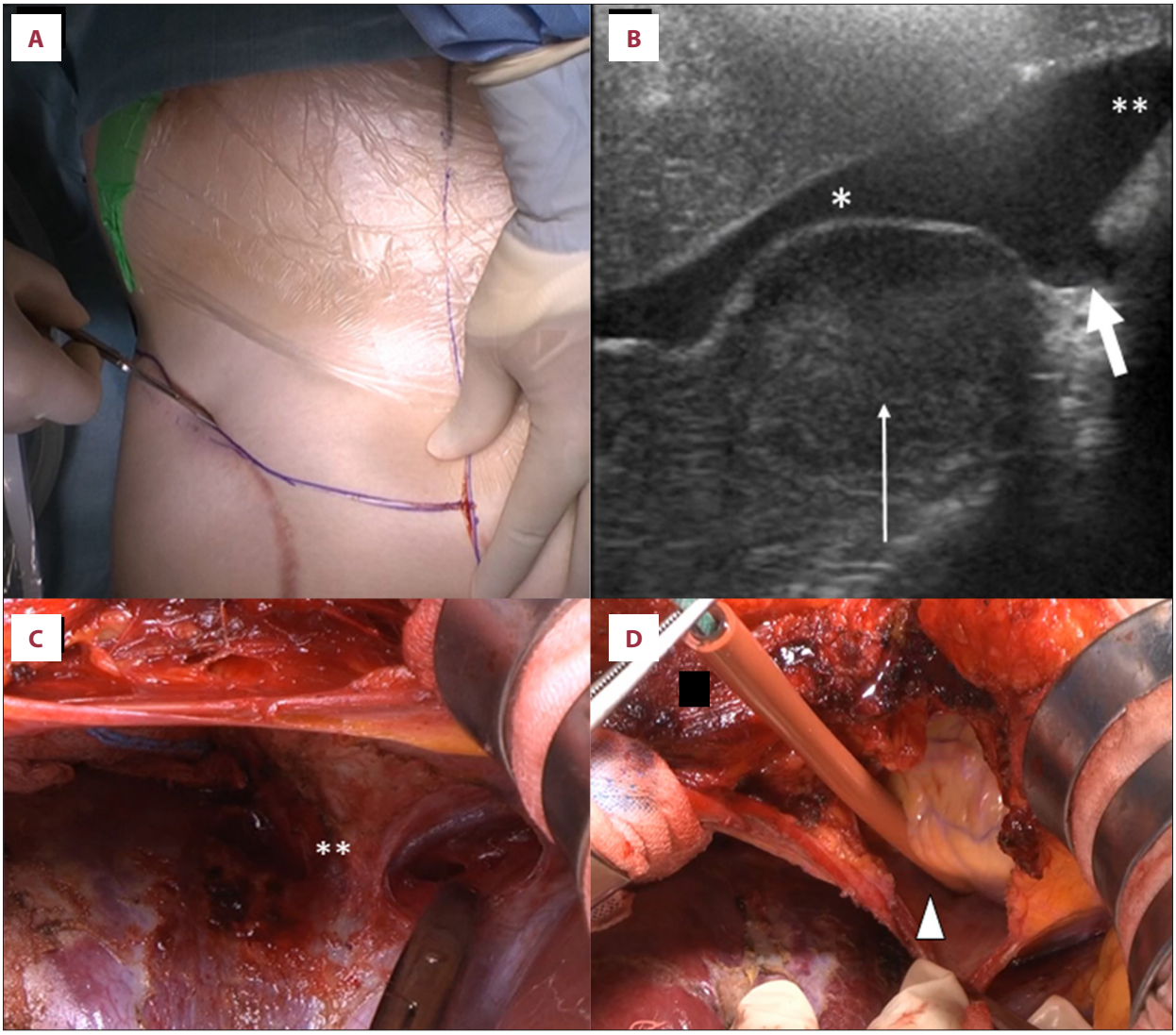

Figure 3. Reoperation for recurrent liver tumor was performed via (**A**) an upper median plus transverse incision. (**B**) Intraoperative ultrasonography showed the round mass (thin arrow) as a hypoechoic lesion compressing the right hepatic vein (*) and the area adjacent to the inferior vena cava (IVC) stump (thick arrow). The middle hepatic vein (**) was not compressed by the tumor. (**C**) The confluence of the MHV was isolated, but dissection around the RHV and the front of the IVC was difficult due to tumor compression. (**D**) To control injury to the IVC, the IVC in the pericardium was encircled by a tourniquet (arrowhead) before hepatectomy.

be side-clamped by the aortic clamp forceps without blocking the outflow of the MHV. We released the tumor without exposure from the confluence of the MHV during liver transection. Then, the IVC wall and liver tumor could be removed via R0 resection (**Figure 4B**), followed by safe closure with running sutures using a 5-0 polypropylene thread (**Figure 4C**). Thereafter, right hepatectomy was performed (**Figure 4D**) to resect the right liver and tumor, which resembled the original tumor compressing the IVC wall and RHV (**Figure 5A**). The operating time was 595 min, and blood loss was 1450 mL. An intra-abdominal drainage tube was placed for 3 days.

Histological findings showed that the tumor did not infiltrate the IVC wall (**Figure 5B, 5C**). The spindle cell-like LMS cells proliferated in a network structure, and nuclear mitosis was observed in 12 cells under high magnification in 10 fields, and MIB-1 positivity was detected in 29% of 1000 tumor cells by Ki-67 staining (**Figure 5D**). Some lesions were necrotic in the tumor, but no vessel infiltration was observed, and the surgical margin appeared to be cancer-free (R0). The final diagnosis was IVC-LMS with liver metastases. No postoperative complications were observed, and the patient was discharged on hospital day 13 because of prolonged ascites. No adjuvant therapy was administered after the second operation, and

This work is licensed under Creative Common Attribution-NonCommercial-NoDerivatives 4.0 International (CC BY-NC-ND 4.0)

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

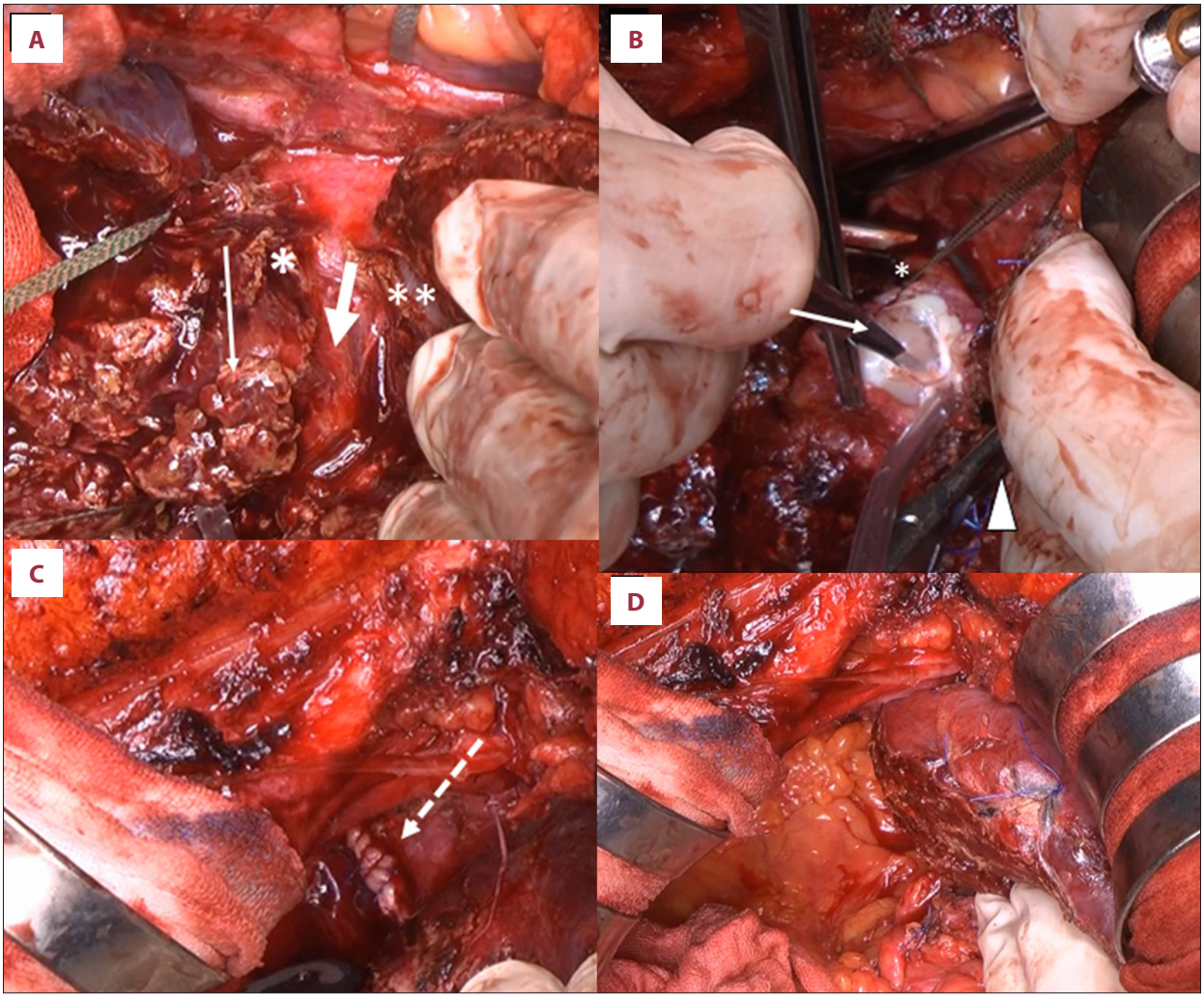

Figure 4. Findings of hepatic transection. (**A**) The liver tumor (thin arrow) was infiltrating the confluence of the RHV (*) and IVC stump (thick arrow). The MHV (**) was secured. (**B**) Side clamping of the IVC (arrowhead) by vascular forceps allowed the vena cava wall to be transected. (**C**) The transected stump was sutured (dotted arrow), and (**D**) the right hepatectomy was completed.

the patient had no tumor recurrence 12 months postoperatively. The patient still received imaging examination of the ultrasonography every 3 months and the enhanced computed tomography every 6 months for a follow-up, and she was still alive at 3 years 8 months (44 months) after the second aggressive surgery.

Discussion

To cure IVC-LMS, complete en bloc resection with negative tumor margins in the retroperitoneal organs should be performed during the initial operation [3]. As described in our previous case report [5], the patient's primary tumor was first diagnosed as a retroperitoneal sarcoma with IVC tumor thrombus, and en bloc resection of the kidney and adrenal glands was performed. After IVC-LMS was histologically diagnosed, distant metastasis disseminating to the lung was predicted because of the existence of viable intraluminal sarcoma lesions in the IVC. Regardless, no adjuvant chemotherapy or brachytherapy was administered, and the tumor did not recur for >2 years. In 1871, Perl first reported a case of IVC-LMS [6]. Approximately 125 years later, Mingoli et al, in a worldwide analysis of 281 patients with a mean age of 54 years, reported that 88% of the patients were women [7]. Wachtel et al later showed that in 377 patients with IVC-LMS, the rate of tumor-free survival at 1 year was 57%, and the overall survival rates at 1 and 5 years were 92% and 55%, respectively. Thus, although the tumor frequently recurred in the early period, the 5-year survival in resected patients was relatively good [8]. The present patient also showed long tumor-free survival, similar to that in the above reports.

Radical resection can be performed via aggressive surgery. LMS of the IVC is categorized by its anatomical location. Zone

This work is licensed under Creative Common Attribution-NonCommercial-NoDerivatives 4.0 International (CC BY-NC-ND 4.0)

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

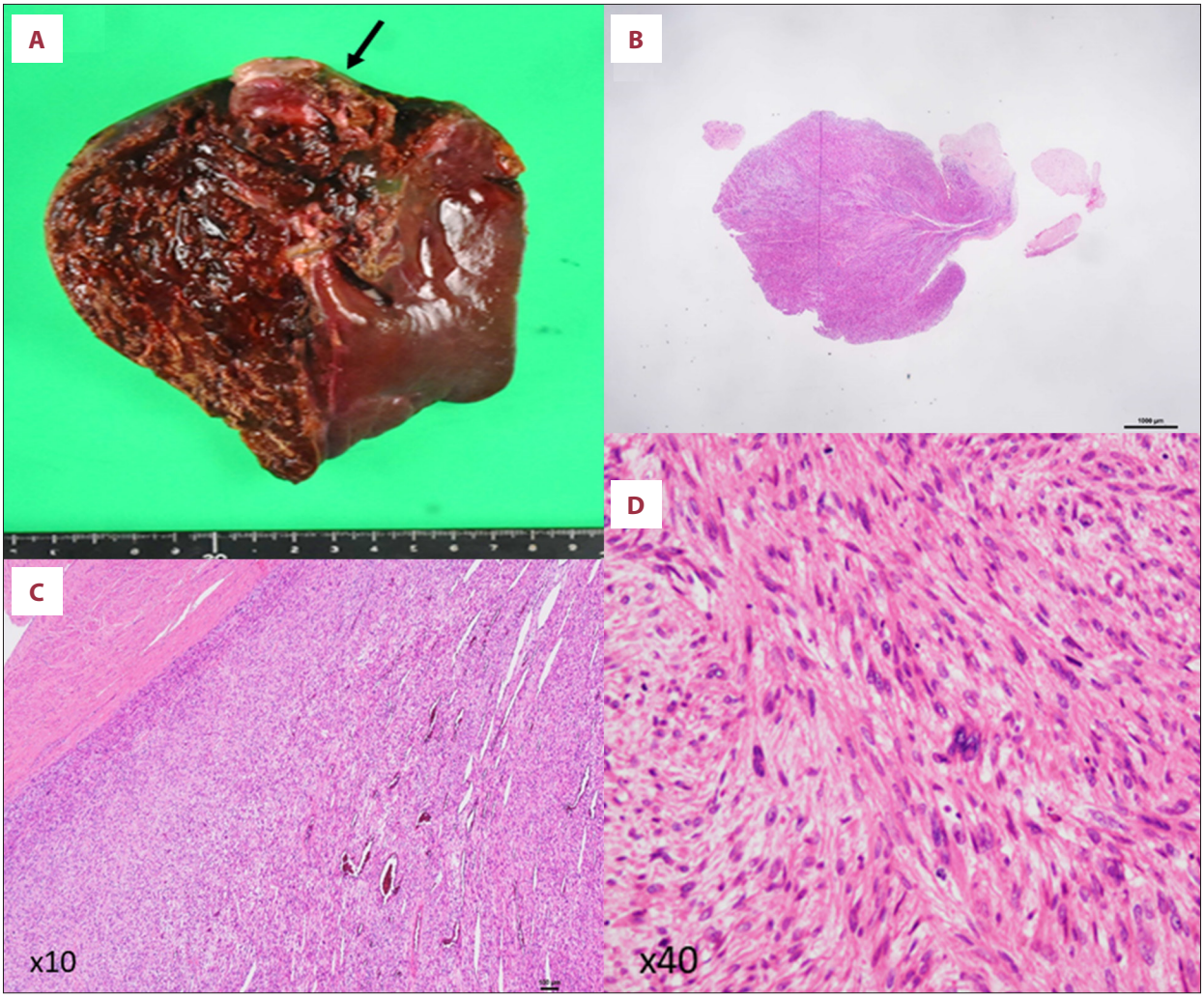

Figure 5. Macroscopic (**A**) and microscopic findings of the tumor of the intrahepatic mass lesion (black arrow). (**B**) Histological findings of hematoxylin and eosin staining by objective magnification: (**B**) ×4, (**C**) ×10 and (**D**) ×40. Intravenous infiltration can be observed by elastic fiber staining in **C and D**. Spindle cell-like leiomyosarcoma cells proliferated within a network structure.

I refers to the infrarenal portion of the IVC, zone II refers to the hepatic veins to the renal veins, and zone III refers to the right atrium to the hepatic veins. The rates of successful radical resection or survival appear to be related to tumor extension zones [9]. According to previous reports [10-12], in cases of tumor extension that spread to the confluences or tributaries of the hepatic veins, liver autotransplantation was necessary for IVC-LMS in zones II-III, and survival rates of over 1 year could be achieved. In the initial operation in the present patient, the tumor tip extended to the orifice of the hepatic vein trunk, and curettage of the floating piece of the tumor was performed by the cardiologist by opening the transected stump of the IVC. In fact, the present specimen showed that the tumor did not recur in the intraluminal space or as a distant lesion adjacent to the IVC stump or liver parenchyma. Although the kidneys were resected together and had a negative surgical margin, local recurrence was observed in the liver. However, no local recurrence was observed in the renal bed; therefore, it was considered that there should be no hesitation in the combined resection of the kidney with the tumor if the contralateral kidney is normal because of the characteristics of sarcoma. With respect to resectability for recurrent tumors, we believe that upfront surgery is the best choice because it is a solitary small lesion and the cancer-free period is over 2 years. In the case of IVC-LMS, sensitivity to preoperative chemotherapy unusual. This case was suspected to be surgically or oncologically resectable. Adjuvant chemotherapy will be selected when the second tumor recurrence is found under the oncologist's comments at our institute. At this stage, the patient had no tumor recurrence for 1 year. Thus, a challenging operation for macroscopic removal can be a promising treatment for long survival, and successful radical hepatectomy for liver metastasis has not yet been reported. However, when deciding on major hepatectomy, the operative indication

This work is licensed under Creative Common Attribution-NonCommercial-NoDerivatives 4.0 International (CC BY-NC-ND 4.0)

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

for bilobar multiple liver metastases should be carefully determined from oncological and liver function points of view.

Conclusions

We report a rare case of local hepatic recurrence originating from an IVC-LMS in a woman who had previously undergone en bloc resection of an IVC-LMS and was subsequently treated with follow-up surgery combining radical right hepatectomy with resection of the IVC stump. She survived for 12 months without tumor recurrence after the second operation (44 months after the first operation), and previous reports have indicated that challenging surgical removal can lead to a good outcome. Long-term follow-up was provided by urological and liver surgeons. Based on this experience, in cases of tumor recurrence after removal of IVC-LMS, it appears that upfront and radical surgery can provide the patient with long-term survival.

Acknowledgements

English has been edited by the language editing company Rise Japan (Number: 220812) at first, and Paperpal (Receipt #1168-2602) at the time of revision.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

References:

1. Dew J, Hansen K, Hammon J, McCoy T, Levine EA, Shen P. Leiomyosarcoma of the inferior vena cava: surgical management and clinical results. Am. Surg. 2005;71:497-501
2. Raut CP, Miceli R, Strauss DC, et al. External validation of a multi-institutional retroperitoneal sarcoma nomogram. Cancer. 2016;112:1417-24
3. 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;12:1203-20
4. Jeong S, Han Y, Cho YP, Kwon TW. Clinical outcomes of surgical resection for leiomyosarcoma of the inferior vena cava. Ann Vasc Surg. 2019;61:377-83
5. Kida K, Terada N, Nakamura E, Mukai S, et al. *En bloc* removal of a large leiomyosarcoma arising from the inferior vena cava using retroperitoneal laparoscopic dissection of posterior vessels followed by cavectomy in a thoracoabdominal approach. Urol Case Rep. 2019;26:100984
6. Perl L, Virchow R. [A case of sarcoma of the inferior vena cava] Arch F Path Anat. 1871;53:378-83 [in French]
7. Mingoli A, Cavallaro A, Sapienza P, et al. International registry of inferior vena cava leiomyosarcoma: analysis of a world series on 218 patients. Anticancer Res. 1996;16:3201-5
8 Wachtel H, Gupta M, Bartlett EK, et al. Outcomes after resection of leiomyosarcomas of the inferior vena cava: A pooled data analysis of 377 cases. Surg Oncol. 2015;24:21-27
9. Fernandez HT, Kim PT, Anthony TL, Hamman BL, et al. Inferior vena cava reconstruction for leiomyosarcoma of Zone I-III requiring complete hepatectomy and bilateral nephrectomy with autotransplantation. J Surg Oncol. 2015;112: 481-85
10. Brekke IB, Mathisen Ø, Line PD, Hauss HJ. Hepatic autotransplantation with ex situ neoplasm extirpation and vena cava replacement. Hepatogastroenterol. 2003;50:2169-72
11. Takatsuki M, Eguchi S, Hashizume K, et al. Liver autotransplantation for an inferior vena cava tumor. Transplantation. 2014;98:e92-94
12. Tuxun T, Li T, Apaer S, et al. Ex vivo liver resection and autotransplantation as surgical option for zone II-III leiomyosarcoma of IVC: A case report and literature review. Front Oncol. 2021;11:690617

This work is licensed under Creative Common Attribution-NonCommercial-NoDerivatives 4.0 International (CC BY-NC-ND 4.0)

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]