Cardio-Oncology

CASE REPORT

Open Access

Sternotomy and extracorporal circulation for fulminant Budd-Chiari syndrome due to leiomyosarcoma of the inferior vena cava

Maciej Wiewiora[1*], Hanna Wiewiora[4], Ewa Chmielik[2], Michal Jarzab[3], Michael Grynkiewicz[4] and Marcin Kubeczko[3]

Abstract

Background Budd–Chiari syndrome is a rare and severe vascular liver disease. We presented patient with fulminant liver failure secondary to leiomyosarcoma of the IVC and thrombosis.

Case presentation A 44-year-old female presented with fulminant liver failure secondary to inferior vena cava (IVC) thrombosis. Contrast-enhanced computed tomography subsequently revealed a thrombus within the IVC, extending cranially to the right atrium and caudally to the renal veins. The patient's condition, characterized by early comatose symptoms, necessitated surgical intervention. Under extracorporeal circulation, a right atriotomy with thrombus lesion removal and descending thrombectomy of the IVC was performed. Hepatic congestion resolved after the thrombus was removed. A pathological examination of the excised thrombus revealed the presence of high-grade leiomyosarcoma.

Conclusions In cases where a thrombus extends from the IVC to the right atrium, urgent surgical intervention with extracorporeal circulation should be considered.

Keywords Budd–Chiari syndrome, Thrombosis, Leiomyosarcoma of the inferior vena cava

Background

Budd–Chiari syndrome is a rare and severe vascular liver disease definied by the presence of partial or complete impairment of hepatic outflow in the absence of right heart failure or constrictive pericarditis. Leiomyosarcoma of the inferior vena cava (IVC) is an extremely rare mesenchymal tumor arising from the smooth muscle constituents of the middle layer with intra- or extraluminal growth. We presented patient with fulminant liver failure secondary to leiomyosarcoma of the IVC and thrombosis.

Case presentation

A 44-year-old female presented with fulminant liver failure secondary to inferior vena cava (IVC) thrombosis and was promptly admitted for treatment. Contrast-enhanced computed tomography (CE-CT) subsequently revealed a

*Correspondence:
Maciej Wiewiora
m-wiewiora@tlen.pl
[1] Department of Cardiac Vascular and Endovascular Surgery and Transplantology, Faculty of Medical Sciences in Zabrze, Medical University of Silesia in Katowice, ul. Sklodowskiej-Curie 9, Zabrze 41-800, Poland
[2] Tumor Pathology Department, Maria Sklodowska-Curie National Research Institute of Oncology, Gliwice 44-102, Poland
[3] Breast Cancer Center, Maria Sklodowska-Curie National Research Institute of Oncology, Gliwice 44-102, Poland
[4] Student Scientific Society, Department of Cardiac, Vascular and Endovascular Surgery and Transplantology, Faculty of Medical Sciences in Zabrze, Sklodowskiej-Curie 9, Zabrze 41-800, Poland

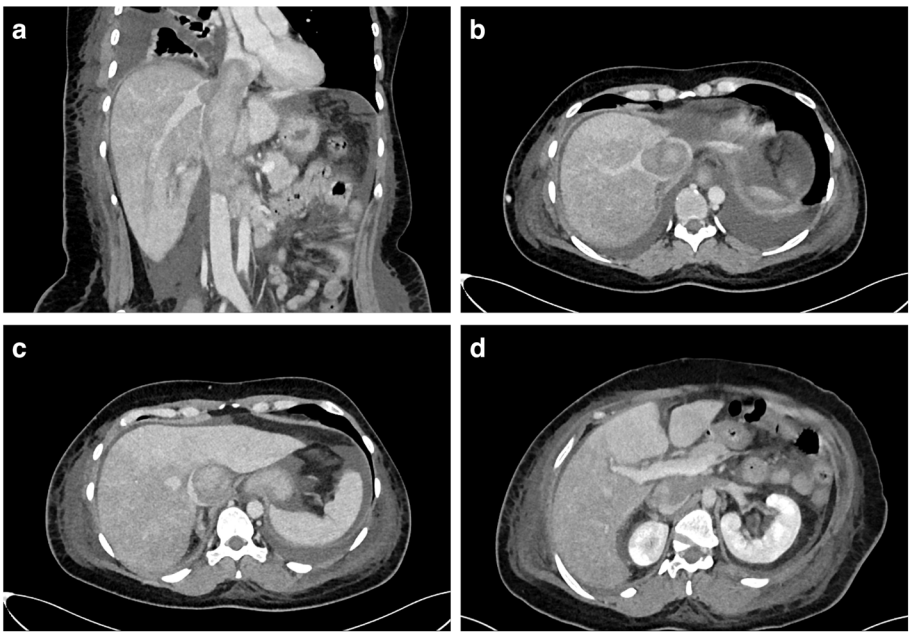

Fig. 1 Contrast-enhanced computed tomography scan: **a** the thrombus in the IVC extending to the right atrium and to the renal veins; **b** and **c** with diffuse patchy enhancement due to hepatic congestion and necrosis; **d** with thrombus in the renal veins

thrombus within the IVC, extending cranially to the right atrium and caudally to the renal veins. The CE-CT results are shown in Fig. 1. Laboratory tests revealed abnormalities in liver and renal function. The bilirubin, alanine transaminase (ALT), aspartate transaminase (AST), alkaline phosphatase (ALP), gamma-glutamyltransferase (GGT), and albumin levels were 34,4 μmol/L, 1381 U/L, 1028 U/L, 102 IU/L, 375 U/L, and 39 g/L, respectively. Coagulation tests relieved abnormalities associated with liver function: the prothrombin time and prothrombin activity were 26 s and 39%, the INR was 2.1, and the activated partial thromboplastin time (APTT) was 54 s. The

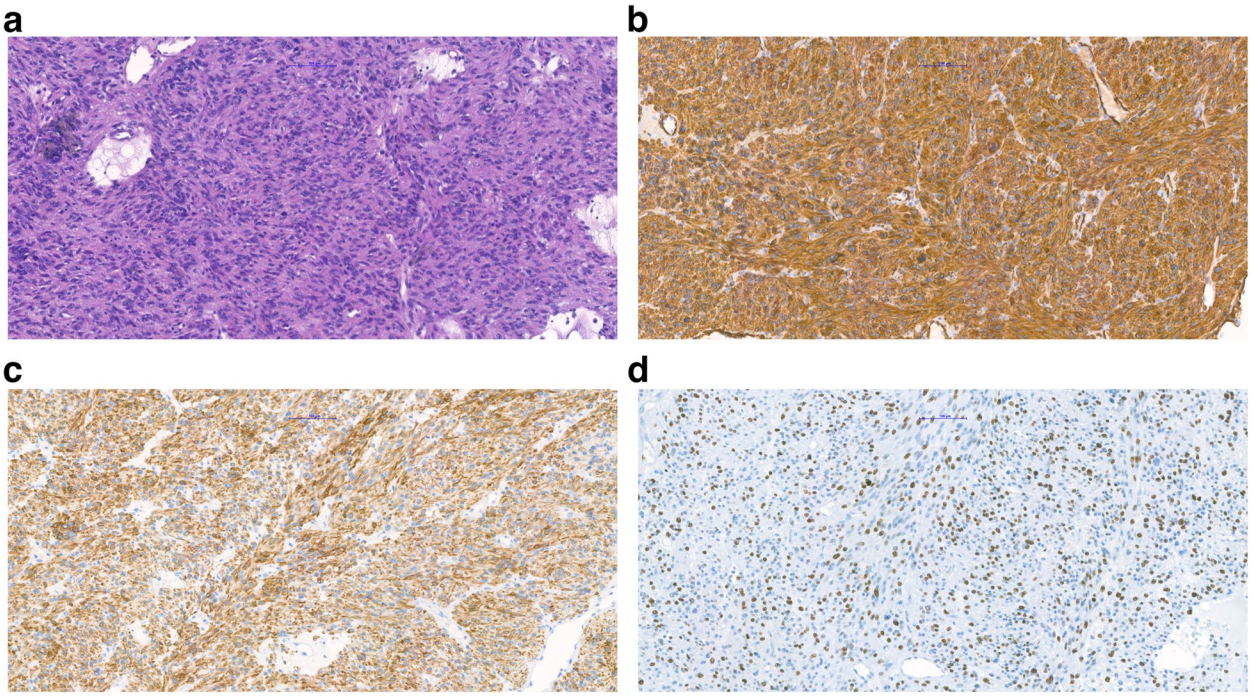

Fig. 2 Histopatological examination: **a** Leiomyosarcoma, hematoxylin & eosin staining **b**) Smooth Muscle Actin (SMA) positive expression **c**) Desmin positive expression **d**) Proliferative index Ki-67 75%

Child–Pugh score was 8. The creatinine, blood urea, and GFR levels were 129 µmol/L, 19,68 mmol/L and 45 ml/min/1,73 m^2, respectively. A complete blood count revealed no typical abnormalities except for elevated leukocyte counts; the white blood cell (WBC) count was 13.5 (10^9/L); the red blood cell (RBC) count was 5.23 (10^{12}/L); the PTL count was 194 (10^9/L); the platelet count (Hgb) was 15.6 (g/dL); the hematocrit count was 45%; the mean corpuscular volume (MCV) was 86 (fl); the mean corpuscular hemoglobin concentration (MCHC) was 29.8 (g/dL) (g/dL); and the mean corpuscular hemoglobin (MCH) count was 34.4 (pg). An arterial blood gas test revealed that acidosis was relieved, with a pH = 7.315 and a lactate level of 2.01 mmol/L.

The patient's condition, characterized by early comatose symptoms, necessitated surgical intervention. The procedure commenced with a median sternotomy. Under extracorporeal circulation, a right atriotomy with thrombus lesion removal and descending thrombectomy of the IVC was performed. Hepatic congestion resolved after the thrombus was removed. The patient was transferred to the Intensive Care Unit for follow-up and had an uneventful postoperative recovery. A pathological examination of the excised thrombus revealed the presence of high-grade leiomyosarcoma. These findings are illustrated in Fig. 2. Notably, 18F-fluorodeoxyglucose positron emission tomography (FDG-PET)showed no signs of malignancy. The FDG-PET scan relieved no findings indicative of a high-glucose-metabolism sarcoma. The results from the FDG-PET imaging study are presented in detail in Fig. 3.

The patient's treatment regimen included a sequential administration of gemcitabine with docetaxel, doxorubicin, and pazopanib.

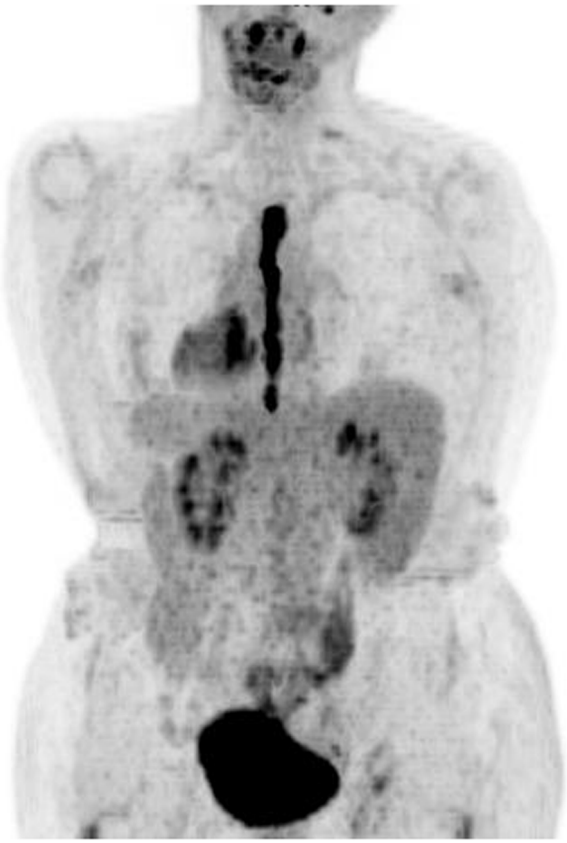

Fig. 3 Results of the 18F-fluorodeoxyglucose positron emission tomography (FDG-PET) scan. There is a very high accumulation of radiotracers along the entire length of the sternum, suggesting a reparative/regenerative process. No findings indicative of a high-glucose-metabolism sarcoma were observed. The positioning of the upper limbs during the scan was due to a recent sternotomy procedure

Discussion

Budd–Chiari syndrome (BCS) typically presents as a sub-acute or chronic condition; however, approximately 1–2% of BCS cases manifest as fulminant hepatic failure, which is associated with very high mortality [1]. The goal of surgical or endovascular intervention in BCS is to relieve hepatic congestion through the recanalization of the hepatic veins. Leiomyosarcoma of IVC is an extremely rare mesenchymal tumor, challenging to diagnose at an early stage, whereby the prognosis is generally poor [2]. Hepatic vein involvement can precipitate Budd–Chiari syndrome [3]. A comprehensive multidisciplinary approach is crucial for managing sarcoma cases, and treatment should be conducted in specialized reference canters [4]. For localized disease, surgical excision is critical for favorable outcomes [5], while systemic treatment remains the cornerstone for managing advanced-staged sarcomas [4].

The surgery was performed as a life-saving intervention due to rapidly progressing liver failure, with a preoperative diagnosis of IVC thrombosis. Preoperative CT angiography did not reveal any tumor-like changes within the IVC. Access through sternotomy and the right atrium provided good control during thrombus removal, minimizing the risk of pulmonary embolism. Given the patient's condition, simultaneous laparotomy and clearance of the entire IVC and renal veins were not performed. The goal of the procedure was to restore IVC patency, facilitating outflow from the hepatic veins. The patient did not require renal replacement therapy after the surgery, and her renal function is currently normal. Owing to the appearance of the removed thrombus, histopathological examination was performed, revealing that the cause of the thrombosis was sarcoma. Following oncological consultation, the patient was eligible for chemotherapy and later underwent resection surgery.

IVC thrombosis is one of the most common causes of Budd–Chiari syndrome, but it rarely presents as dramatically as it did in this patient. Typically, the initial approach is conservative treatment or the use of minimally invasive, usually endovascular, techniques. While IVC thrombosis involving the right atrium is observed in clinical practice, it very rarely necessitates urgent surgery.

In cases where a thrombus extends from the IVC to the right atrium, urgent surgical intervention with extracorporeal circulation should be considered.

Authors' contributions
Conception and design: Maciej Wiewiora Data collection: Maciej Wiewiora, Hanna Wiewiora Michael Grynkiewicz Analysis and interpretation: Maciej Wiewiora, Hanna Wiewiora, Marcin Kubeczko Writing the article: Maciej Wiewiora, Michael Grynkiewicz, Marcin Kubeczko Critical revision of the article: Ewa Chmielik, Michal Jarzab Overall responsibility: Maciej Wiewiora, Michael Grynkiewicz, Ewa Chmielik, Michal Jarzab Hanna Wiewiora, Marcin Kubeczko.

Funding
Not applicable.

Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
Not applicable.

Competing interests
The authors declare no competing interests.

Received: 15 July 2024 Accepted: 12 November 2024
Published online: 25 November 2024

References

1. Van Wettere M, Bruno O, Rautou PE, Vilgrain V, Ronot M. Diagnosis of Budd-Chiari syndrome. Abdom Radiol (NY). 2018;43(8):1896–907.
2. Wang MX, Menias CO, Elsherif SB, Segaran N, Ganeshan D. Current update on IVC leiomyosarcoma. Abdom Radiol. 2021;46:5284–96.
3. Kraft C, Schuettfort G, Weil Y, Tirneci V, Kasper A, Haberichter B, et al. Thrombosis of the inferior vena cava and malignant disease. Thromb Res. 2014;134:668–73.
4. Gronchi A, Miah AB, Dei Tos AP, Abecassis N, Bajpai J, Bauer S, et al. Soft tissue and visceral sarcomas: ESMO–EURACAN–GENTURIS Clinical Practice Guidelines for diagnosis, treatment and follow-up. Ann Oncol. 2021;32:1348–65.
5. Fatehi Hassanabad A, Ball CG, Kidd WT. Inferior vena cava tumor thrombus: clinical outcomes at a canadian tertiary center. Perfusion. 2023. https://doi.org/10.1177/02676591231202682. [online ahead of print]

Publisher's Note