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## Prevention of life-threatening bleeding complication from splenic venous ectasia by B-mode-, color Doppler- and contrast-enhanced ultrasound in a patient with ALL

Evelyn Martin<sup>1</sup>, Christian Görg<sup>2</sup>, Amjad Alhyari<sup>2</sup>, Ehsan Safai Zadeh<sup>2</sup>,  
Hajo Findeisen<sup>3</sup>, Corinna Trenker<sup>4</sup>

<sup>1</sup> Department of Anesthesiology, University Hospital Marburg, Marburg, Germany

<sup>2</sup> Department of Gastroenterology, University Hospital Marburg, Marburg, Germany

<sup>3</sup> Department of Internal Medicine, Red Cross Hospital, Bremen, Germany

<sup>4</sup> Department of Hematology/Oncology, University Hospital Marburg, Marburg, Germany

Corresponding author: Corinna Trenker; e-mail: Trenker@med.uni-marburg.de

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acute lymphatic leukemia

### Abstract

**Aim of the study:** Spontaneous splenic rupture is a serious complication of pathologically altered spleen tissue, associated with a high mortality rate. **Case description:** We describe a spontaneous splenic rupture in a patient with acute lymphoblastic leukemia undergoing chemotherapy. Ultrasound revealed splenomegaly, and diffuse splenic acute lymphoblastic leukemia-infiltration was suspected. In addition, only color Doppler sonography and contrast-enhanced ultrasound diagnosed splenic vascular ectasias with a venous-flow-profile. During therapy, short-term sonographic follow-up examinations were able to reveal an increase in the size of venous ectasias and the associated increased risk of spontaneous splenic rupture. Based on these sonographic findings, immediate surgical splenectomy was performed in the spontaneous splenic rupture case and the patient survived. **Conclusions:** Ultrasound is an important diagnostic method in patients with newly diagnosed malignant hematological diseases to detect disease-related splenic pathologies. Short-term follow-up examinations of splenic vascular pathologies can detect size progression and a potential risk of spontaneous splenic rupture with life-threatening bleeding.

## Introduction

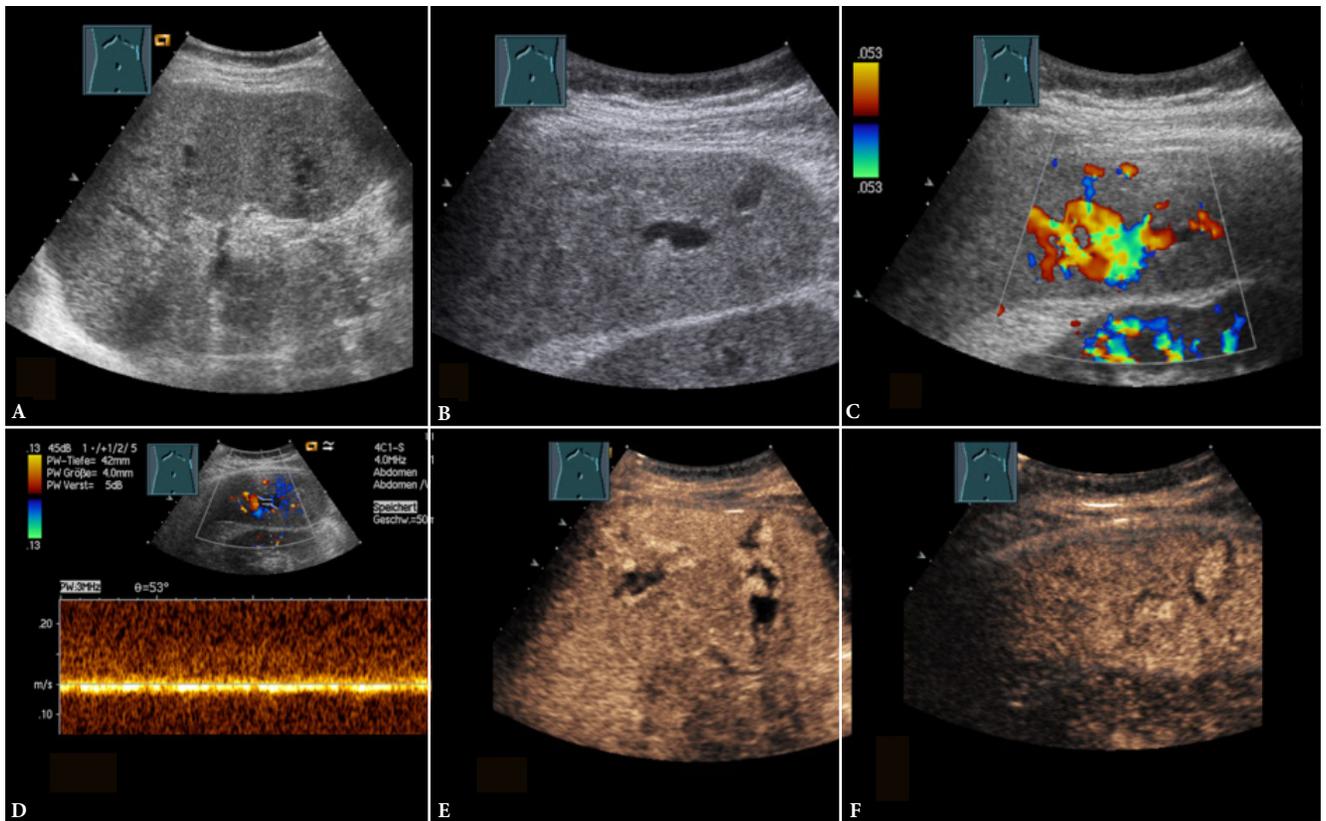
Spontaneous splenic rupture (SSR) is a rare but serious complication of pathologically altered splenic tissue, with an incidence of 0.1–0.5%<sup>(1,2)</sup> and a mortality rate of up to 38%<sup>(3)</sup>. In a retrospective analysis, SSR was most frequently observed in patients with hematological and oncological diseases (48.5%)<sup>(4,5)</sup>. The pathogenesis of SSR is variable, with splenic infarcts and splenic lacerations in minor trauma proposed as factors<sup>(6)</sup>. However, intralial arterial pseudoaneurysm is a known vascular complication and was observed in 12% of 41 patients with SSR in one study<sup>(3)</sup>. In patients with acute lymphoblastic leukemia (ALL), abdominal ultrasound is part of the recommended diagnostic staging prior to therapy<sup>(7)</sup>. Splenic ALL-involvement can be diagnosed in up to 50% of patients with ALL<sup>(8–10)</sup>. Different sonographic patterns of splenic lymphoma involvement have been reported<sup>(11)</sup>. Hypochoic nodular lymphoma lesions are the most commonly observed splenic pattern in B-mode ultrasound (B-US)<sup>(11)</sup>. Diffuse infiltration is characterized by inhomogeneous parenchyma and/or a splenomegaly<sup>(11)</sup>. Contrast-enhanced ultra-

sound (CEUS) is an established examination method in the diagnostic and further differentiation of splenic pathologies, e.g. in the demarcation of focal splenic lymphoma lesions<sup>(12)</sup>.

Below, we describe the first case of splenic venous varicose ectasia with secondary spontaneous splenic rupture diagnosed by B-US, color Doppler sonography and CEUS in a patient with ALL.

## Case presentation

A 35-year-old male patient with a diagnosis of ALL presented to our ultrasound center as part of the initial diagnostic staging prior to therapy. He had night sweats but no history of fever or weight loss. The patient was in poor general condition and had enlarged cervical lymph nodes. An abnormal blood count was diagnosed, with a leukocytosis of 276 Giga (G)/l with 80% lymphatic blasts, thrombocytopenia (17 G/l), and anemia (hemoglobin 114 g/l). The lactate dehydrogenase (LDH) level was 4,179 units (U)/l.



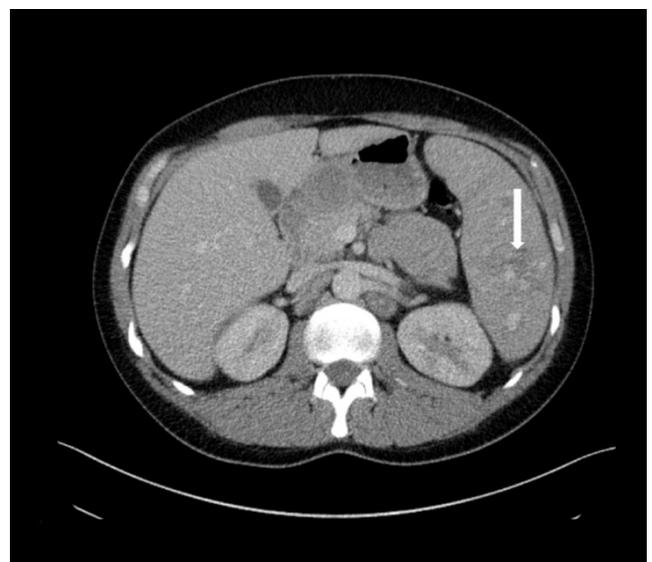
**Fig. 1.** B-US shows splenomegaly with inhomogeneous parenchyma and multiple cyst-like lesions (A, B). In color Doppler sonography (CDS), these lesions present low-impedance continuous venous flow signal (C, D). Contrast-enhanced sonography (CEUS) after one (E) and 6 minutes (F) revealed the echo-free lesions suspected for venous ectasias

B-US of the abdomen was performed with a Siemens Sequoia 512 (Siemens, Erlangen Germany) ultrasound machine, using a convex probe (4.0 MHz). Splenomegaly (with spleen length of 16 cm, thickness of 5 cm) and inhomogeneous parenchymal echotexture suspicious for diffuse lymphoma infiltration were observed. Moreover, within the parenchyma, multiple hypoechoic/anechoic “cyst like” focal lesions were seen in Fig. 1 A and B.

In color Doppler sonography (CDS), the lesions showed a characteristic low-impedance continuous flow signal, suspicious for a venous vascular pathology (Fig. 1 C and D). Contrast-enhanced ultrasound (CEUS) was performed to further explore the findings. The patient received an intravenous bolus injection of 2.4 ml of the contrast agent SonoVue (Bracco, Konstanz), followed by 10 ml sodium chloride in accordance with the European Federation of Societies for Ultrasound in Medicine and Biology (EFSUMB) guidelines<sup>(12)</sup>. A marked enhancement of the vascular pathology was observed. The findings were suspicious for intrasplenic venous ectasia (Fig. 1 E and F).

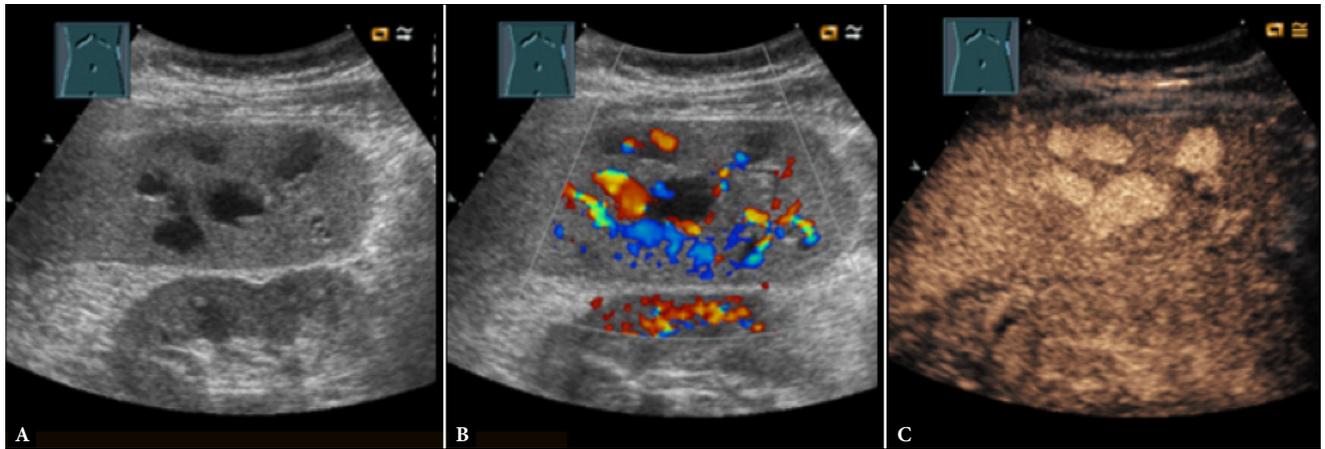
For staging, a contrast-enhanced computed tomography (CT) examination was performed on the same day. The CT scan confirmed splenomegaly with inhomogeneous contrast enhancement. However, reliable differentiation between lymphoma involvement, hemangiomas or multiple small infarcts with compensatory arterial hyperperfusion was not possible based on CT findings (Fig. 2).

The results of the ultrasound examination and the CT-scan concerning the splenic pathology were discussed with the surgical depart-

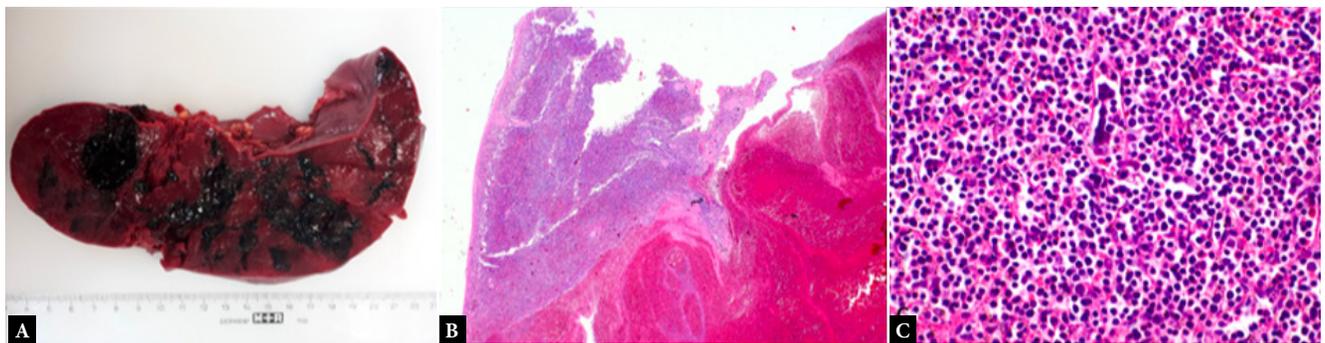


**Fig. 2.** Contrast-enhanced computed tomography (CT) presents a splenomegaly with inhomogeneous contrast uptake (arrow) (courtesy of Prof. Dr. Mahnken, Department of Radiology, Marburg)

ment, and sonographic follow-up examination was recommended. The next day, sonographic follow-up examination showed stable findings. In the meantime, pre-phase ALL-therapy with dexamethasone and cyclophosphamide according to the German Multicenter



**Fig. 3.** During short-term follow-up, B-US, CDS and CEUS showed size progression of the “cyst-like” vascular lesions



**Fig. 4.** Organ preparation after emergency splenectomy shows macroscopically organ rupture (A). Overview of the rupture site (top) with parenchymal bleeding (bottom right) (B). In HE 1:20, splenic rupture with capsule and parenchymal defect were seen. The overview shows no typical architecture of the spleen with red and white pulp. C. In HE (magnification  $\times 200$ ), diffuse infiltration of the red pulp/spleen parenchyma by ALL-blasts with enlarged vesicular nucleoli and only a small cytoplasm was observed (courtesy of Dr. A. Ramaswamy, Department of Pathology, Marburg)

Study Group on Adult Acute Lymphoblastic Leukemia (GMALL) protocol was started. On the second day of the pre-phase therapy, ultrasound showed size progression of the patient’s venous splenic lesions in B-US, CDS and CEUS (Fig. 3 A–C). Surgeons were informed about the sonographically diagnosed size progression of intrasplenic venous ectasia. Due to the patient’s hemodynamically stable situation and urgency of ALL therapy, it was decided to proceed conservatively with short-term sonographic and clinical follow-up. However, the patient suffered hemorrhagic shock the following night. Immediately after bedside sonography with detection of free abdominal fluid, emergency laparotomy with splenectomy was performed without further diagnostic CT imaging. A spontaneous two-sided rupture of the spleen was verified both intraoperatively and histopathologically (Fig. 4 A and B). In addition, the histopathological specimen showed diffuse ALL-blast infiltration of the spleen (Fig. 4 C).

The patient survived the emergency splenectomy procedure, and chemotherapy could be successfully continued shortly after the operation. At present, the patient is in complete remission.

Upon obtaining written informed consent from the patient for the publication of the case report, approval was subsequently granted by the ethics committee of the Philipps University of Marbur (approval number RS 22/60).

## Discussion

B-US of the abdomen is usually the first-line noninvasive diagnostic method in patients with hematological or oncological diseases for detecting splenomegaly or splenic involvement as well as other therapy-relevant pathologies<sup>(13,14)</sup>.

Lymphoma involvement is the most common reason for hypoechoic focal splenic lesions in these patients<sup>(15)</sup>, but splenic cysts, splenic infarctions, splenic rupture, and microabscesses due to splenic candidiasis represent other important differential diagnoses<sup>(15–19,20)</sup>.

CEUS provides more information about the perfusion of the lesions and helps exclude differential diagnoses such as hemorrhagic cyst<sup>(12)</sup>. Splenic lymphoma is characterized in CEUS by arterial iso- (46.3%) -or hypo (53.6%) enhancement of the focal lesions followed by parenchymal hypoenhancement (100%)<sup>(21)</sup>. Non-traumatic arterial splenic pseudoaneurysm (NTISP) is a rare differential diagnosis for focal lesions in a diseased spleen, showing arterial flow signals in CDS<sup>(3)</sup>. The development of NTISP could occur in 12% ( $n = 5/41$ ) of cases of nontraumatic spontaneous splenic rupture<sup>(3,5)</sup>. One case report describes NTISP diagnosed by B-US and CEUS, and confirmed by a CT scan in a patient with acute leukemia<sup>(22)</sup>.

In our report, we describe for the first time a case of intrasplenic venous ectasias with secondary splenic rupture in a patient with ALL and diffuse leukemic infiltration of the spleen diagnosed by ultrasound. The patient's venous ectasias were identified by a venous flow signal in CDS and contrast media uptake in CEUS. Short-term sonographic follow-up revealed size progression during ALL therapy and indicated an increased risk of rupture.

## Conclusion

The reported case highlights the importance of abdominal ultrasound in patients with newly diagnosed malignant hematological diseases and demonstrates the necessity of short-term follow-up examinations for splenic vascular pathology. In contrast to CT, B-US and CEUS had a capacity to reliably differentiate splenic lesions as venous ectasias. Size progression and thus an increased risk of two-stage splenic rupture with potentially life-threatening bleeding could be detected by short-term sonographic follow-up.

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## Conflict of interest

Christian Görg received fundings from Bracco Imaging for CEUS workshops for education. The other authors do not report any financial or personal connections with other persons or organizations which might negatively affect the contents of this publication and/or claim authorship rights to this publication.

## Author contributions

Original concept of study: CG, CT. Writing of manuscript: EM, CT. Final approval of manuscript: EM. Critical review of manuscript: EM, AA, ESZ, HF.