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Case Report

Spontaneous splenic rupture during induction therapy in acute myeloid leukemia: An unusual case. ☆,☆☆

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ABSTRACT

Spontaneous splenic rupture (SSR) is a rare life-threatening emergency. In hematological settings, it is uncommon in acute myeloid leukemia (AML). We report an atypical case of SSR in a 73-year-old male with AML where a prompt imaging ultrasound assessment played a key role. Performed noninvasively at bedside, it allowed rapid imaging diagnosis, confirming its essential role even in the presence of hematological disease.

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Introduction

Spontaneous splenic rupture (SSR) is a rare life-threatening emergency. In hematological settings, different etiologies have been reported, but they are uncommon in acute myeloid leukemia (AML), especially after induction chemotherapy initiation. [1–3] Several clinical associations have been reported,

but no clear risk factor has been identified. Prompt imaging diagnosis is required to reduce morbidity and mortality. Here we report a case of SSR due to an extremely rare condition known as peliosis in a 73-year-old male with AML. Ultrasound examination played a key role in this case, leading to rapid assessment and prompt, successful management. Performed noninvasively at bedside, ultrasound imaging has confirmed its role in hematological emergencies.

☆ Competing Interests: Nothing to disclose.

☆☆ Spleen peliosis_Inform Consent: Patient provided informative consent concerning his clinical informations and scientific purposes. This consent is signed at hospital admission for all recovered patients.

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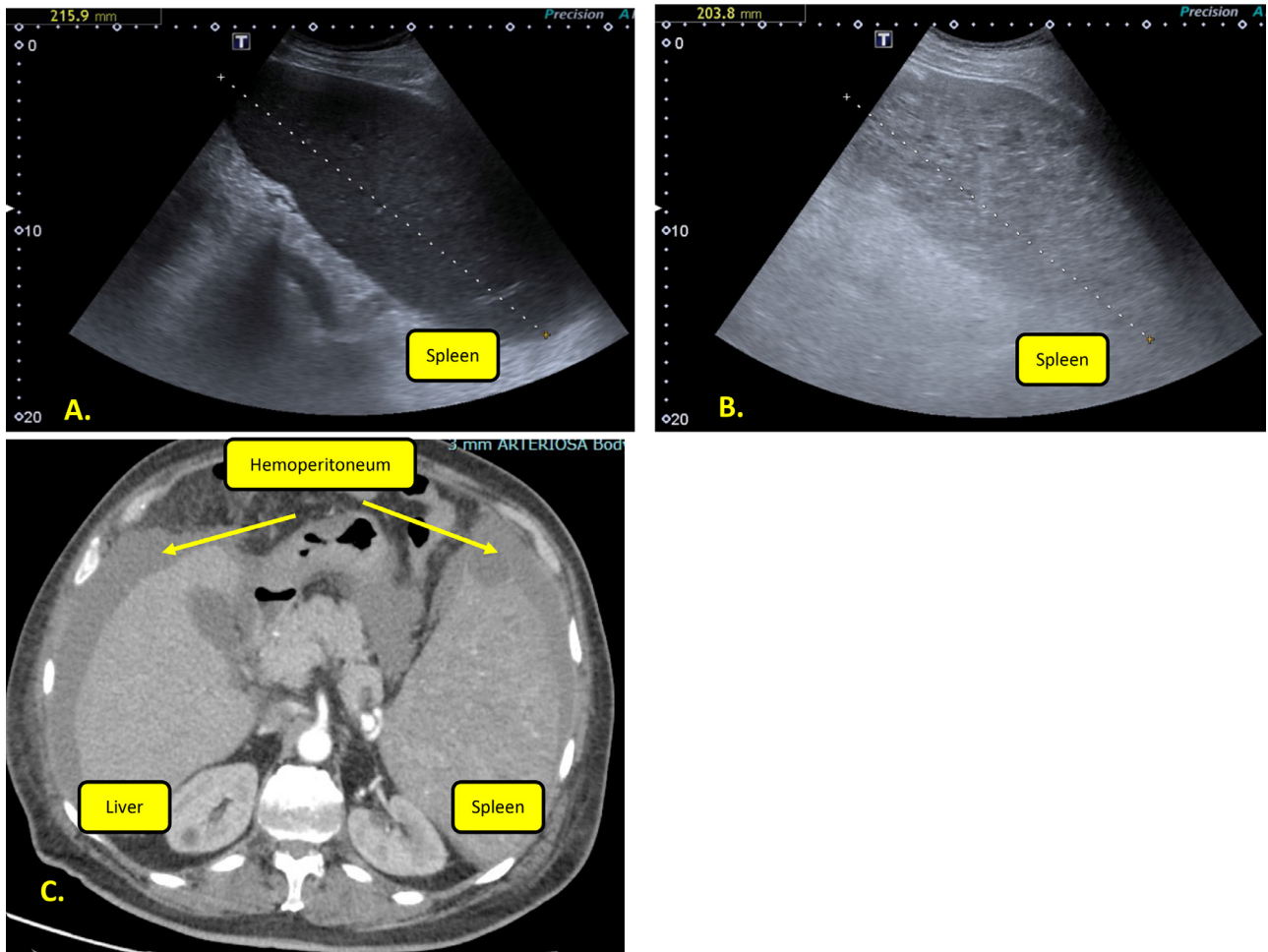


Fig. 1 – (A) Spleen ultrasound appearance at diagnosis. (B) Ultrasound spleen appearance in peliosis rupture. Ultrasonographic imaging showed inhomogeneous splenic parenchyma and increased size. (C) Abdominal computer tomography (CT) frame. CT showed hemoperitoneum and marked splenomegaly (with a pole-to-pole diameter of about 200 mm) with many hyperdense areas.

Case report

A previously healthy 73-year-old male presented at our emergency department with a 2-week history of a sore throat and chest pain. The patient had no hematological malignancy history. Upon physical examination, he was pale with skin bruising on his arms and mild epistaxis. Vital signs were normal, as were pulmonary, neurological, and abdominal examinations. Legs edema was reported. A blood sample was quickly collected showing severe leukocytosis with a white blood cell count (WBC) of 155.800/ μ L (normal range [NR] 4000–10 000), hemoglobin (Hb) of 10.3 g/dL (NR 14.0–18.0), and a platelet (PLT) count of 37.000/ μ L (NR 122 000–350 000). The differential formula presented 94% immature monocytes, 4% lymphocytes, and 2% neutrophils. Renal function, electrolytes, uric acid, and coagulation parameters were all within normal laboratory limits. A prompt peripheral cytofluorimetric assay confirmed the suspicion of acute monocytic leukemia, and the patient was admitted

to our hematological unit where a cytoreduction strategy with hydroxyurea was started. Whole-abdomen ultrasound imaging revealed homogeneous splenomegaly (pole-to-pole diameter of 21 cm) on examination (Fig. 1A). SARS-CoV-2 molecular analysis was negative.

The diagnosis was confirmed by a bone marrow examination that showed a marked increase in immature monocytic elements (bright CD33 and CD64 without CD14), hyper-cellularity, and minimal residual functional hematopoiesis. Cytogenetic analysis found hyperdiploidy with 51–52 chromosomes and deletion of 7q. The presence of a mixed lineage leukemia rearrangement (11q23) was also found during the molecular biology study. A diagnosis of high-risk acute monocytic leukemia was given.

After effective stabilization of hematological values with hydroxyurea, the patient underwent first-line treatment with Decitabine (20 mg/m²/day on days 1–5). Planned subsequent treatment should have consisted of Venetoclax on day four, delivered with a rump-up strategy with dose-escalations of 50 mg/day up to 400 mg/day.



Fig. 2 – (A) Spleen after surgical intervention. Spleen weighed 1300 g and measured 21 x 15 x 10.5 cm. A large portion of the spleen capsule was interrupted, and confluent nodular areas were present on the organ surface. (B) Spleen longitudinal section. Parenchyma showed multiple blood-filled cystic cavities; the red pulp was altered by histiocyte infiltration. The presence of extramedullary hematopoiesis was demonstrated. The described pattern agreed with ultrasound splenic peliosis appearance.

On day six, the patient experienced abdominal pain. Upon physical examination, he was pale and vital signs were normal. Pulmonary, neurological, and abdominal examinations were also normal. A prompt bedside abdominal ultrasound demonstrating the presence of ascitic effusion in both perihepatic and small pelvis confirmed splenomegaly (pole-to-pole diameter of about 20.5 cm), but an irregular structure was found. The parenchyma presented a highly nonhomogeneous appearance, with multiple large hypo-aneogenic areas. Doppler signal revealed peripheral enhancement pattern on spleen focal pathological areas. No evidence of focal hepatic or renal lesions was detected (Fig. 1B).

Blood exams revealed a marked worsening of Hb levels (from 8.1 g/dL to 6.8 g/dL) and PLT count was 6 000/ μ L; absolute neutrophil count was 0/ μ L. Coagulation test revealed prothrombin time 20.1 s (NR 0.80–1.20), activated partial thromboplastin time 39.4 s (NR 28.0–40.0), fibrinogen 100 mg/dL from 503 mg/dL (NR 150–450), and D-dimer 28.86 mg/L (NR 0.00–0.50). Clinical stabilization with blood product transfusions and fresh frozen plasma was started. Nevertheless, deterioration of clinical conditions and laboratory markers worsened. Acute intra-abdominal bleeding was suspected and confirmed by an urgent computer tomography (CT). CT showed hemoperitoneum and marked splenomegaly (with a pole-to-pole diameter of about 200 mm) with many hyperdense areas, suggesting a diagnosis of SSR (Fig. 1C). Following surgical consultation, an exploratory laparotomy with splenectomy was performed (Fig. 2A). Four liters of blood were removed from the abdominal cavity during surgery. To support vital functions, 38 units of red blood cells, 6 units of fresh frozen plasma, and 5 units of apheresis platelets were transfused. The post-surgery course was regular with no major complications, and the patient was extubated on day three and transferred back to the hematology unit.

The spleen weighed 1300 g and measured 21 x 15 x 10.5 cm. A large portion of the spleen capsule was interrupted, and confluent nodular areas were present on the organ surface. The parenchyma showed multiple blood-filled cystic cavities; the red pulp was altered by histiocyte infiltration. The presence of extramedullary hematopoiesis was demonstrated. Histological examination confirmed no leukemia substitution. The described pattern agreed with splenic peliosis diagnosis (Fig. 2B).

A repeat ultrasound on postoperative day 14 confirmed complete resolution of the hemoperitoneum. The patient was discharged one month after the initial admission date. The complete blood count (CBC) on discharge showed WBC 2120/ μ L, Hb 9.9 g/dL, and PLT count of 70 000/ μ L with a normal differential. The patient is currently undergoing outpatient management, continuing his Decitabine cycles. Venetoclax has not been resumed to date due to increased risk of infection.

Peliosis is an extremely uncommon disorder of unknown etiology, and only a few cases are described in hematological settings. [4–6] Many reports have associated clinical factors including malignant diseases, chronic infections, and ingestion of certain drugs such as anabolic steroids and long-term erythropoietin. [7,8] Although uncommon, some reports suggest that males, ages > 20 years, presence of splenomegaly, and use of cytoreductive therapy might be associated with increased risk of SSR in hematology. [1] In this case study, the patient had high-risk acute myeloid leukemia and was 73 years old. Additionally, he had splenomegaly at baseline ultrasound examination, but the spleen tissue was also described as homogeneous.

Usually, patients with splenic peliosis are asymptomatic, but they develop acute abdominal symptoms if the organ spontaneously ruptures. Only after receiving cytoreductive therapies did splenic peliosis rupture occur. Before SSR became

clinically diagnosed, we noted markedly worsening coagulation. The disseminated intravascular coagulation may be a result rather than a cause of SSR, as suggested by another report. [3]

Given the extreme rarity of the disorder in most cases, peliosis diagnosis is obtained after histopathological examination of the resected organ or at autopsy. Its histological appearance is characterized by multiple blood-filled cystic cavities primarily involving the mononuclear phagocytic system (liver, spleen, bone marrow, and lymph nodes). In this case, the parenchyma showed characteristic aspects, and the red pulp was altered by histiocyte infiltration. Of note, extramedullary hematopoiesis was found, and no leukemia substitution was confirmed. [3] It appeared during a spleen ultrasound after multiple large hypo-anaecogenic areas reflected multiple blood-filled cystic cavities upon histopathological exam.

Prompt initiation of massive transfusion played a leading role in the successful stabilization of SSR emergency. A high index of suspicion is required for early diagnosis and management, particularly when risk factors and rapid clinical deterioration are observed. Ultrasound examination plays a key role in this setting. Performed non-invasively at bedside, it allowed for rapid imaging assessment. As reported by observational data and confirmed by our case, multidisciplinary approaches reduce the morbidity and mortality associated with hemorrhagic emergency in ultra-high-risk patients.

Author contributions

RT, GC and ML: were major contributor in writing the manuscript. ML, GC, LD, MC, NM, BT, NP and RT: analyzed and interpreted haematologic data regarding SSR and peliosis condition. LD, MT, ML, VG, FR and GC: provided advice for treatment. ALL AUTHORS: read, revised and approved the manuscript.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at [doi:10.1016/j.radcr.2021.07.087](https://doi.org/10.1016/j.radcr.2021.07.087).

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