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Atraumatic Splenic Rupture as a Rare Complication of Acute Myeloid Leukemia: A Case Report and Literature Review

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Abstract

Introduction: Atraumatic Splenic Rupture (ASR) is a rare but life-threatening clinicopathological phenomenon with limited information on patient features, occurrence, or etiology. ASR is an uncommon and lethal complication that is observed in infectious (mainly mononucleosis) and hematological diseases (mainly malignant homeopathies) in more than half of cases. Mortality is approximately around 20%, and some deaths occur before the diagnosis is confirmed, while others occur after surgery due to delayed management and poor patient status.

Case Presentation: A 48-year-old man with no history of the underlying disease presented to the Emergency Department with abdominal pain. He was admitted with leukocytosis 145×103/µl, hemoglobin 6.4 g/dl, platelets 15×103/µl, erythrocyte sedimentation rate 89 mm/h, and D-Dimer 1043 ng/FEU ml. Sputum test through PCR ruled out severe acute respiratory syndrome coronavirus 2 infections. Due to peripheral blood smear and bone marrow aspiration/biopsy, acute myeloid leukemia was diagnosed for the patient. On the third day of hospitalization, the patient's abdominal pain intensifies. Ultrasound revealed medium free fluid inside the abdomen and pelvis. The patient was transferred to the operating room to undergo an emergency laparotomy. There was a large hematoma in the spleen with a rupture in its posterior surface. Splenectomy was performed, and the histopathological study of the spleen showed leukemic involvement, capsular ruptures, and subcapsular hematomas.

Conclusion: ASR can occur for a variety of reasons, including non-traumatic or idiopathic factors. In the absence of significant trauma, emergency physicians should be aware that splenic rupture can occur. ASR is more likely to present with symptoms similar to the underlying diseases.

Keywords: Acute myeloid leukemia, Atraumatic splenic rupture, Case report, Hematological diseases, Splenic rupture

1. Introduction

Atraumatic Splenic Rupture (ASR) is a rare but life-threatening clinicopathological phenomenon with limited information on patient features, occurrence, or etiology. Sneezing, coughing, vomiting, and straining during defecation or muscular effort can all cause an atraumatic rupture in a fragile spleen with pathological changes (1). The symptoms and signs of splenic rupture may be confused with those of the underlying disease or its associated consequences, making the diagnosis of splenic rupture difficult. At the time of rupture, nearly all patients (95%) complained of abdominal pain. However, biliary colic, aortic aneurysm, perforated viscous, pancreatitis, and pectoral angina are all symptoms that might be confused with this pain (2).

A total of six major etiological groups were identified for ASR, including neoplastic (30.3%), infectious (27.3%), inflammatory/non-infectious (20%), medication and treatment-related (9.2%), mechanical (6.8%), and normal spleen (6.4%). Males dominated females 2:1, and the mean age is 45 years (age range: 18-86 years).

Regardless of the treatment mode, the overall ASR-related fatality rate was 12.2%. Splenomegaly and an age of more than 40 years are strongly linked to an elevated ASR-related death rate (1). ASR is a rare complication, and therefore, we would like to present our clinical experience with this unusual patient for more familiarity with its various facets. Our study has been documented by the Surgical CAse REport guidelines.

2. Case Presentation

A 48-year-old man with no history of the underlying disease presented to the Emergency Department with abdominal pain and symptoms of nausea and anorexia. Moreover, he had no history of abdominal trauma and taking medication. He received the COVID-19 vaccine 10 days ago (Sinopharm vaccine). For two days, he had weakness and myalgia. Upon arrival, the arterial blood pressure, pulse rate, body temperature, and respiratory rate were 110/60 mmHg, 82/min, 36.6°C, and 14/min, respectively. The patient's blood tests were examined (Table 1). Sputum test through PCR

ruled out severe acute respiratory syndrome coronavirus 2 infections. Peripheral blood smear and bone marrow aspiration/biopsy were performed, which led to the diagnosis of acute myeloid leukemia (AML). Viral tests for hepatitis and AIDS were negative. Treatment was started with hydroxyurea, allopurinol, meropenem, and metronidazole. On the second day of hospitalization, two units of red blood cells were transfused. On the same day, he had two convulsions that were controlled with diazepam and phenytoin. The brain CT scan was normal. Subsequently, on the third day of hospitalization, the patient's abdominal pain intensifies. Ultrasound reported splenomegaly with multiple lymphadenopathies around the head of the pancreas and porta hepatis with a diameter of 12 mm. Medium-free fluid containing internal debris was also observed inside the abdomen and pelvis. Surgical consultation was performed. Due to the generalized tenderness and free fluid noticed in the ultrasound, peritoneal aspiration was performed under the ultrasound guide. Fresh blood was aspirated, and the patient was transferred to the operating room and underwent a laparotomy. There were about two liters of blood in the abdominal cavity, as well as a large hematoma in the spleen with a rupture in its posterior surface. Splenectomy was performed (Figure 1). The patient had an uneventful postoperative period, and he was transferred to the hematology ward for further treatment. The histopathological study of the spleen showed leukemic involvement, capsular ruptures, and subcapsular hematomas.

Table 1. Blood results

Blood	Result	Unit	Normal Range
WBC	145	$\times 10^3/\mu l$	4-10
HB	6.4	g/dl	13.3-17.2
HCT	19.2	%	38.9-50.9
PLT	15	$\times 10^3/\mu l$	150-450
ESR	89	mm/h	<20
CRP	29.5	mg/L	Up to 5.0
D-Dimer	1043	ng/FEU ml	<500
BUN	33	mg/dl	10-50
Cr	1.7	mg/dl	0.7-1.4
ALT	135	IU/L	Up to 41
AST	91	IU/L	Up to 37
PT	17.3	Seconds	10.5-14
PTT	45.1	Seconds	10-40
INR	1.39	_	_

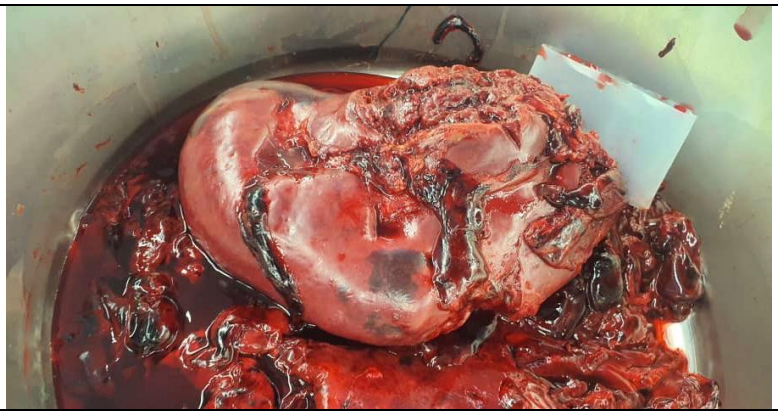

Figure 1. Macroscopic view of the spleen

3. Discussion

ASR is an uncommon and lethal complication that is observed in infectious (mainly mononucleosis) and hematological diseases (mainly malignant homeopathies) in more than half of cases. Mortality is approximately around 20%, and some deaths occur before the diagnosis, while others occur after surgery as a result of delayed management and poor patient status (3). Splenic infiltration related to hematological disorders, splenic infarction, and coagulation abnormalities are three potential reasons for splenic rupture in patients with leukemia (4). Renzulli et al. collected information from 845 patients with ASR from 1980 to 2008. The etiological analysis revealed that neoplastic reasons, infection, inflammation, and drug/treatment-related accounted for 30.3%, 27.3%, 20.0%, and 9.2% of the cases, respectively. The spleen was characterized as fully normal in 7% of the cases, and no etiological reason for ASR was found. A single etiological component was discovered in 84.1% of the cases. In 8.2% of the patients, the ASR was caused by a

combination of two etiological factors, and in 0.7% of the cases, the splenic rupture was caused by a combination of three etiological factors. Laparotomy, CT scan, ultrasonography, scintigraphy, laparoscopy, angiography, and post-mortem examination were taken in 42.3%, 32.4%, 18.6%, 0.7%, 0.5%, 0.3%, and 5.2% of all cases, respectively, to make the final diagnosis of splenic rupture (1). Similarly, there was one etiology in our patient, which was spleen involvement during acute leukemia. Although ultrasound and peritoneal aspiration helped hemoperitoneum be diagnosed, a definitive diagnosis was made by laparotomy.

Bassler et al. reviewed 613 cases of ASR between 1950 and 2011. Infectious, hematologic, and non-hematologic neoplasms were the most common related diseases. Amyloidosis and internal trauma, such as coughing or vomiting, and rheumatologic disorders are also less common. Colonoscopy was the most common procedure cited as a cause of the rupture. Anticoagulants, thrombolytic, and recombinant G-CSF are among the medications linked to rupture (5). In our patient, there was no history of taking any medications. In addition, no diagnostic procedures, such as colonoscopy, were performed for our patient before splenic rupture. AML diagnosis was the only thing done for the patient.

Bauer et al. in 1981 reported pain in the left hypochondrium with irradiation to the homolateral shoulder (Kehr's sign), which was considered a characteristic of a hemoperitoneum sign that was only noted in 17% of 48 cases of spleen ruptures. Furthermore, hypotension, fever, and tachycardia were reported in 66%, 74%, and 75% of the cases, respectively. There was no consensus between spleen size and risk of rupture since 40% of patients with splenic rupture did not have splenomegaly (2). Our patient also had no Kehr's sign and splenomegaly.

Odabas et al. described a 62-year-old man who was complaining of abdominal pain, nausea, vomiting, and diarrhea. The results of the vital sign were as follows: the blood pressure 100/60 mmHg, pulse rate 100/min, temperature 36.4˚C, and respiratory rate 22/min. He had taken chemotherapy for AML three years earlier and had healed without problems. His leukocyte and platelet count, as well as hemoglobin level, were 9630/mm^3, 99×10^3/µl, and 7.7 g/dl, respectively. FAST revealed a large amount of free fluid in the perihepatic and perisplenic regions. Following a clinical follow-up, the patient's arterial blood pressure steadily decreased, and the patient, who had become anuric, was diagnosed with a spleen rupture and operated on (6). Fortunately, our patient's surgery was performed before the deep and severe hemorrhagic shock, and therefore, the patient survived.

Hajri et al. presented a 48-year-old man with no recent trauma who was referred to the hospital's emergency room for acute abdominal pain while getting chemotherapy for AML. The patient's vital signs were as follows: heart rate of 120/min, respiratory rate of 30/min, arterial pressure of 90/60 mmHg, and body temperature of 37.4˚C. The abdomen was flexible at the time of admission. Ultrasound revealed a large amount of hemoperitoneum. A splenic rupture with extensive hemoperitoneum and bilateral pleural effusion was discovered on an abdominal CT scan. Emergency laparotomy decided to be performed, and the splenectomy procedure went well.

Unfortunately, the patient had cardiovascular decompensation after surgery and died on the seventh day (7). Our patient had an uneventful postoperative period and was discharged from the surgical ward in good general condition.

De Santis et al. presented a 66-year-old man with AML, fatigue, minor left upper abdominal pain, fever, and tachypnea. The patient was pale on examination, with a blood pressure of 130/80 mmHg and a pulse rate of 125 /min. Lab test showed hemoglobin, white blood cells, and platelet at 8.3 g/dl, 278×10^9/L, and 367×10^9/L, respectively. On the day of the diagnosis, leukapheresis was started, and two sessions were conducted at a 12-h interval. The patient developed severe tachypnea and hemodynamic instability by the end of the second session, culminating in circulatory shock, unresponsive to fluid infusion and dopamine. The electrocardiogram revealed ST depression at this time. The patient died two hours later since cardiac resuscitation was not responded to (8). Fortunately, in our experience, the patient's life was saved by a quick decision to have surgery.

Clinical symptoms and imaging modalities are used to diagnose ASR. Although paracentesis is a useful diagnostic tool, negative results cannot conclusively rule out hemorrhage. The FAST is a low-cost and quick diagnosis approach for intraperitoneal fluid formation or hematoma that can be performed at the patient's bedside in an emergency room. The FAST is chosen as a fast and non-invasive diagnostic imaging approach for hemodynamically unstable patients (6). The combination of paracentesis and FAST also helped us to rapidly diagnose the hemoperitoneum in this case.

Even in hemodynamically stable patients, the idea of performing a total splenectomy can be justified for three reasons. First, the etiology of the ASR, as well as any underlying systemic diseases, will be determined by histological examination of the spleen. Second, ASR can be caused by a wide range of malignancies, making any organ preservation strategy impossible. Third, a pathological change or infiltration of the splenic parenchyma may have already affected splenic function, resulting in functional hyposplenism. (1)

Splenic artery embolization is a safe procedure that allows the patient to be stabilized quickly and, if necessary, to have a safer and more elective splenectomy. This new therapeutic alternative in the treatment of ASR can provide both the benefits of splenectomy and the conservative treatment benefits, making it an attractive therapeutic option (9).

4. Conclusion

Patients admitted to the Emergency Department with abdominal pain and distension, negative history of trauma, and anemia with unknown etiology should be evaluated for ASR. It is important to know that ASR can occur for a variety of reasons, including non-traumatic or idiopathic factors. In the absence of significant trauma or previously known splenic pathology, emergency physicians should be aware that splenic rupture can occur. ASR is more likely to present with symptoms similar to the underlying disease. We expect that by raising awareness of these events, emergency clinicians would be better able to diagnose similar cases of splenic rupture on time.

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Footnotes

Authors' contributions: MEK performed the surgery and reviewed the literature, TZ and YA wrote the manuscript and reviewed the literature. HH and AKB reviewed the literature. All authors read and approved the final manuscript.
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