

# **Annals of Case Reports**

# **Case Report**

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# Interventional Radiology as Treatment of Massive Haemoperitoneum due to Atraumatic Spleen Rupture in Patient with Acute Myeloid Leukemia: Case Report and Literature Review

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# **Central Message**

Non-traumatic splenic rupture is a rare complication of acute myeloid leukemia. We report about an adult man who presented with sudden chest pain and dyspnea. The initial work-up was oriented towards a suspected diagnosis of pulmonary embolism. He showed leukocytosis with monocytosis, treatable abdomen and normal life parameters. In the following hours appeared a state of hemorrhagic shock. A surgical clinical evaluation, combined with ultrasound diagnostic support and CT scan enabled early diagnosis of haemoperitoneum which required close monitoring of the patient followed by a definitive optimal cure by interventional angiography. Atraumatic Splenic Rupture (ASR) is an uncommon pathologic condition in which bleeding from the spleen occurs for a variety of nontraumatic reasons, often in patients with hematological diseases. The Authors report the case of a ASR in a patient who had just received the diagnosis of acute Myelo-Monocytic Leukemia (AML) presenting with hypovolemic shock due to haemoperitoneum, and who was treated in emergency by artery embolization. This case offers the opportunity for a review of the literature and underlines the need for a multidisciplinary approach in the management of critically ill patient.

## Introduction

Atraumatic rupture of the spleen is a rare but life-threatening occurence because of the prognosis of the patients presenting with acute abdomen for the evolving haemoperitoneum, hypovolemic shock and electrolyte imbalance [1]. In this scenario a correct and less time-consuming diagnostic work-up as well as a multidisciplinary team are of a paramount importance for a rapid diagnosis to carry-out the best therapeutic approach. Nowadays, in patients with different comorbidities the classical open surgery can be replaced by non-invasive treatments with interventional-radiology.

A first distinction should be made between atraumatic

pathological spleen-rupture rather than that occurring in a healthy spleen. The pathologies of the spleen, which often cause voluminous spleen enlargement, range between infectious diseases, lysosomal storage diseases and neoplastic pathologies, first of all leukemias and lymphomas. Until a decade ago the treatment was exclusively performed by open surgery, and more recently by video-laparoscopic procedures, with high morbidity and mortality. The improvement of the techniques of interventional radiology has allowed a conservative and less life-threatening approach by the embolization of the proximal spleen vessels which guarantees both a good hemostasis creating vicarious collateral arterial circles up and the complete exclusion of the organ (iatrogenic asplenia).

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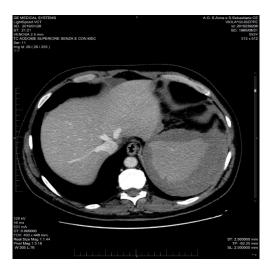
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# **Case Report**

A 54-year-old male, no smoke, no comorbidity, was admitted to the emergency department because of fever, asthenia, leukocytosis with monocytosis, mild anemia and thrombocytopenia. Clinical examination showed hepatomegaly and splenomegaly (6-8 cm bcm). Blood counts showed: Hb 11,8 g/L, Platelets 40000/uL, WBC 74000/uL, PA 120/70 FC 90bpm rhythmic, SpO<sub>2</sub> 98%. The patient was then referred to the Hematological Unit where received diagnosis of acute myelo-monocytic leukemia. At molecular analysis FLT3-ITD mutation was present. The patient starts cytoreduction with hydroxycarbamide 4,5 g per day.

Two days later,

- On morning, the blood count showed: GB 60,000 Hb 11 PLT 40,000
- In the afternoon, the patient feeled dizzy while in the bathroom, but did not lose consciousness; ECG and echocardiogram showing a picture compatible with hypovolemia at the level of the vena cava and the patient is treated with a plasmaexpander.
- At 6.00 pm the patient performed CT and a new US evaluation of the abdomen; at that time blood counts showed: Hb 7.9 g /dL, WBC 79000/uL and Platelets 32.000/uL. Hemorragic shock was suspected.
- The patient was stabilized with plasma-expander and Packed-Red Cells (PRC) and after angio-TC (Figure 1) was referred to the Interventional radiology.
- Arteriography showed an active bleeding (Figure 2) of the lower pole of the spleen, so he received a selective embolization of the splenic artery by puncturing the right femoral artery with subsequent catheterization of the celiac tripod. The splenic artery catheterization is performed with a cobra 5 f catheter. Catheterization of the splenic artery is carried out and 4 tungsten spirals were released near the hilum (2 mm 6 x 5.0 mmm, 1 5 mm x 4.5 mm and 1 mm 6 x 100 mm) (Figure 3).
- After the procedure the patient showed a rapid resumption of diuresis, elevation of the arterial blood pressure, a reduction in the heart rate.



**Figure 1:** Small air bubbles from centrilobular emphysema. No evidence of pleural and / or pericardial effusion. In abdomen abundant amount of free liquid in peritoneal cavity with periepatic site, perisplenic, along the parietocholic showers and in the pelvic excavation, with supra-fluid density from likely vertebral bone. Spleen of increased size (DL 159 mm) with evidence of a small and constant linear hypodensity scheme on the lower pole.

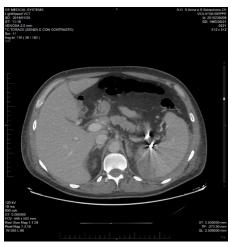


**Figure 2:** Pre-embolization arteriography.



Figure 3: Post-embolization arteriography.

The day after, the patient performed a new CT (Figure 4) showing the absence of bleedings. The patient was treated with induction chemotherapy including Cytarabine (200 mg/sm for 7 days), Daunorubicin (60 mg / sm on day 1,3,5) plus Midostaurin (100 mg BID from the day +8 to day +21). Complete remission was achieved and after consolidation treatment the patient received allogeneic stem-cell transplant. The patient is now in complete remission from leukemia, without complications (Figure 5).



**Figure 4:** Bilateral pleural effusion at posterior and apical basal course with maximum extension to supine patient of about 30 mm on the right and of 17 on the left with atelectasis of the contiguous lung parenchyma. Outcomes of splenic embolization with spleen of increased size and with lesion of the inferior pole, extended blood effusion, perisplenic, periepatic in abdomen and pelvis center.



**Figure 5:** No significant densitometric alterations to the lung parenchyma are appreciated. Disventilatory striae in basal bilaterally. Emphysematous bulla in apical site ds. Mediastino in axis. Non-visibility of lymph node swellings in the mediastinal ilo. Liver in place, with morphovolumetric limits, with a finely inhomogeneous structure, without evidence of focal lesions in place in its context. Gallbladder extended, alitiasic. The intra and extra hepatic bile ducts are not dilated. Results of splenic embolization with spleen of increased size, with inhomogeneous density and with presence of malacic hypodense area at the level of the lower pole, with coarse fluid perisplenic collection.

#### **Discussion**

Although it is a rare occurrence, atraumatic spleen rupture must be considered for differential diagnosis in patients with splenomegaly in whom signs of shock occurs. The experience derived from the rotation of general surgeons at the trauma center has meant that they acquired skills for a rapid response, both in terms of diagnosis as well as treatment in patients who present with hypovolemic shock. In this case, the haemoperitoneum was associated with an alteration of the hemocoagulative parameters because of acute leukemia, therefore a laparotomy aimed at splenectomy would have led to an increase in mortality and perioperative morbidity.

**Incidence:** the spleen is the most commonly injured solid organ in blunt abdominal trauma. It represents 31%-50% of cases. The prevalence of isolated splenic injury in blunt abdominal trauma decrease with age. The spleen is predispose o injury due to its fragile structure and position immediately beneath the left ribcage [1]. The incidence of rupture in diffuse B-cell non Hodgkin lymphoma is highly infrequent, despite reports of various non-traumatic splenic rupture in the literature [2].

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**Etiology:** Non traumatic splenic rupture is common and often related to a pathological spleen. Common causes of non-traumatic splenic rupture include myeloproliferative disease, vasculitis and infections (such as malaria, mononucleosis). However, diffuse large B-cell lymphoma remains an obscure cause of splenic rupture unique attention [3]. The major causative groups were classified as follows:

- -neoplastic (30,3%)
- hematological (13,7% diffuse large B-cell lymphoma, NHLs, HLs, acute lymphoblastic leukemia) (4)
- infectious (27,3% malaria, mononucleosis, cytomegalovirus [5], varicella zoster [6])

Inflammatory, non-infectious (20%)

Drug or medical treatment related (9,2%, cocaine [7], splenic rupture following colorectal surgery and hemodialysis [8], colonoscopy, pancreatitis [9))

Normal spleen - idiopathic (7%).

Medication related (anticoagulation and thrombolytics)

Pregnancy- related and others.

Chemotherapy.

**Symptomatology:** there is a variation in symptom manifestation in patients with splenic rupture. The presence of abdominal pain has been frequently reported. In 20% of the cases Kehr's sign (sharp radiating pain to the left shoulder) can be observed. In larger splenic injuries, signs of hypovolemic shock, such as tachycardia, rapid breathing, paleness, reduced capillary filling time and hypotension is a common presentation. Concomitant symptoms include nausea, alterations of consciousness, intestinal obstruction, and peritonitis.

**Diagnosis:** there are several nomenclatures describing the rupture of the spleen without trauma, such as atraumatic, pathological, atraumatic, idiopathic, occult rupture. Atraumatic splenic rupture was first described by Rokitansky [10] an Atkinson and it is also frequently used. Atraumatic is more accurate, and it can be classified as "Atraumatic-pathological splenic rupture" and "Atraumatic-idiopathic splenic rupture" according to the etiological factors and pathological changes in the spleen.

**Treatment:** splenectomy may be indicated in the management of selected patients with myeloid leukemia, but in the most of the cases, this procedure is burdened by a high morbidity and mortality, mainly due to severe sepsis. The postoperative adverse effects of the asplenic condition lead to the exclusion of surgical treatment in most patients with neoplastic diseases. Damage control interventional radiology is an aggressive and time-conscious algorithm that prioritizes saving life of the hemorrhaging patient which conventional emergency interventional surgery

cannot efficiently do. Hemodynamically unstable patient is a constant challenge to any trauma team. The imaging diagnostics and treatment decisions crucially influence patient's survival. Hemodynamic instability increase the probability of death by 1% for every 3 min and increase the risk of mortality by nearly 47% for every hour delay [11]. Damage control interventional radiology's main concern is that to control hemorrhage by any mean in order to save life in a extremely time-conscious, aggressive and minimally invasive way. It is assigned and implemented to any patient with either: SBP <80 mmHg or; SBP <90 mmHg and more than one of the following: 3 or more active life-threatening bleeding sources and/or any evidence or suspicion of coagulopathy [12]. The nonoperative management of splenic lesions is, to date, the method of choice in the hemodynamically stable patient, even if, in our case, the unstable patient has been rapidly stabilized. Currently about 70% of splenic lesions are treated non-operatively. This diffusion is based on the rationale of avoiding unnecessary surgical interventions and the risks associated with them and maintaining an organ that has, especially in young ages, an important immune function and whose removal is not risk-free (13).

Non-operative management of splenic lesions can be divided in observation or splenic artery embolization. Splenic artery embolization can increase the success rate of non-operative management by stopping ongoing bleeding and by preventing delayed rupture of the spleen [14,15, 16). These studies advocate the use of angiography and embolization in the presence of the following CT findings: contrast extravasation, pseudoaneurysm or arteriovenous fistula, large haemoperitoneum (our case), and a high grade of injury. Minimal haemopetironeum was definited as intraabdominal blood located only in the perisplenic recess, significant haemoperitoneum was definited as intra-abdominal blood located in four peritoneal recess. In our case, hemodynamically unstable patients were responding to fluid resuscitation and treated with proximal embolization of splenic artery. Diagnostic series of the splenic artery were obtained using a 5Fr catheter and the selective catheterization of splenic artery branches coaxial microcatheters and microguidewires were required.

## **Conclusions**

Non operative management of patients with atraumatic splenic rupture is associated with a reduction in morbidity and mortality in critically ill patients. In this way, by implementing interventional radiology within a customizer algorithm will bring about damage control interventional radiology as an effective alternative or partner to surgery.

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collection, analysis and interpretation of data, writing of the report and the decision to submit the article for publication.

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