Spontaneous Splenic Infarcts and Rupture Presenting as Massive Haemoperitoneum

Gezahen N. Ayane, DM¹, Cox Megan, DM², Jemal Z. Shifa, DM³, Kadimo Khutsafalo⁴,

Abstract

Spontaneous rupture of a normal spleen without a history of an evident blunt abdominal trauma is a rare clinical entity and can occur in both histologically normal and diseased spleen. Nontraumatic spontaneous splenic rupture (NSSR) has a high morbidity, as there is often little or no clinical history to suggest its presence, and it is vital that physicians consider this diagnosis when evaluating patients with abdominal pain and hypotension. We present a case of 32–years–old male who presented to Accident and Emergency Department with history of abdominal pain, vomiting and diarrhea, there was no history of trauma. Examination revealed initial hypotension, epigastric abdominal tenderness, and no palpable masses. An-abdominal ultrasound revealed massive free fluid and splenic hematoma; an emergency splenectomy was performed successfully. The patient recovered uneventfully and was discharged from the hospital. This case report has taught as that spontaneous rupture of a normal spleen can occur in a patient with negative results of malaria peripheral blood smear and antigen for P. Vivax and P. falciparum. Physicians should consider the diagnosis of spontaneous spleen rupture in a patient presenting with abdominal pain and hypotension even without a history of trauma; because an early intervention can be life-saving.

Keywords: Spontaneous, spleen, disease, hypotension, ultrasound, splenectomy.

¹ Gazahen N. Ayane, University of Botswana, Faculty of Medicine, Private Bag 00713, Gaborone, Botswana Email: Medpeter2003@yahoo.fr.

² University of Botswana, Faculty of Medicine,

³ University of Botswana, Faculty of Medicine,

⁴ University of Botswana, Library

Introduction

Rupture of the spleen is potentially life threatening complication usually occurring after abdominal blunt trauma. Unlike traumatic splenic rupture, non-traumatic splenic rupture is extremely rare and may occur in 0.1-0.5% of patients with no associated trauma(Agarwal, Sonkar, Kushwaha, & Gaurav, 2013; Gedik, 2008). The first case was reported by Rokitansky in 1861(Agarwal et al., 2013) and Atkinson in 1874(Amonkar & Kumar, 2009; Gazel, Açıkgöz, Kasap, Yiğman, & Güneş, 2015; Wehbe, Raffi, & Osborne, 2008)and Knoblich distinguished the non-traumatic rupture of a pathological spleen from the extremely rare case of non-traumatic splenic rupture of an unknown origin(Kim, Lee, Park, Lee, & Chang, 2005). Spontaneous rupture occurs most commonly as a result of pre-existing pathology of the spleen, such as splenic artery aneurysm, thalassemia, malignancy or infectious mononucleosis(Amonkar & Kumar, 2009; Fernandes, dos Santos, Costa, & Barata, 2013; Gedik, 2008; Kim et al., 2005; Randriamarolahy et al., 2010). It rarely occurs in the setting of a normal spleen. We report a case of spontaneous splenic infarction and rupture that occurred in patient previously infected with malaria, but clinically asymptomatic and serum-negative for malaria, along with an appropriate literature review.

Case report details

A 32-years-old HIV-negative male with previous medical history of Malaria (episodes in 1999; 2008; and 2011), and family medical history of Diabetes Mellitus, presented with a one day history of abdominal pain, nausea, vomiting and diarrhea into the accident and emergency department. His vital signs were normal [(BP88/33 mmHg); pulse (86 beat/minute); (RR18 per/Minute)] and his abdomen was soft, slightly distended with tenderness at epigastrium, no guarding. There was no hepato-splenomegaly; a digital rectal examination was also found normal. A fast ultrasound showed a small amount of intra-abdominal free fluid; but chest and abdominal erect X-rays were normal. His blood laboratory investigations showed the results in Table 1.

Features	Patient	References	Conclusions
Assessed	Findings	Values	
Hb (Hemoglobin)	9.6 gr/dl	12-15gr/dl	Low
MCV (Mean Corpuscular Volume)	85.3	83-99fl	Normal
WBC (White Blood Cells)	10.4	4.5-10x10 ⁹ /L	Normal
Plet (Platelets)	69	$150-400.10^3$	Low
U (Urea)	12.0	2.0-7.0mol/L	High
Cr (Creatinine)	313	53-132mol/L	High
RBS (Random Blood Sugar)	16mmol/L	4.4-7.8mmol/L	High

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Furthermore, other blood investigations such as electrolytes and, liver function tests were normal as well as hematology coagulation tests. Thick and thin peripheral blood smear did not show any malaria parasites. There were no abnormal cell in peripheral blood smear. Antigen test for p. vivax and P. falciparum species were negative.

This patient was reviewed by an internal medicine specialist and admitted to the medical ward with "new diagnosis" of Diabetes Mellitus with acute renal failure and ascites. One day after his admission he still complained of abdominal pain, and abdominal distension, and his vitals were within normal ranges (BP-107/53); Pulse (84/ Minute) but his Hb had dropped to 6.7gram/dl. A repeat of abdominal ultrasound revealed (massive free fluid in the abdominal cavity, and a perisplenic hematoma). The patient was taken for emergency laparotomy, 3200 cc of blood were found in the abdominal cavity, and a ruptured spleen was observed while the rest intra-abdominal organs were unaffected. Splenectomy and abdominal lavage were performed, and the patient was admitted to ICU for close observation and transfusion of 5 units of whole blood and 2 units of fresh frozen plasma. After 24 hours, the patient was transferred into a surgical ward where he successfully recovered and was discharged without any complication after two days. The post splenectomy pathology report stated that the spleen weighted 764gm and measured 19x16x14 cm. It was ulcerated and hemorrhagic. The spleen showed shaped wedges infracts in addition to rupture. The etiology of these infracts was not established and the final diagnosis was confirmed as "splenic infracts with rupture". There was no evidence of active malaria infestation because of the negative rapid and smear tests as well as the fact that the spleen histopathology report was not compatible with a malaria symptomatology.

Discussion

Trauma is the most common cause of splenic rupture, while non-traumatic splenic rupture (NSR) is a rare condition. Non-traumatic rupture of this very vascular organ is described as either "pathological" rupture or" spontaneous" rupture. Recognized causes include infections, haematological, malignant, metabolic, infiltrative and local disorders as reported in Table 2.

Infectious causes	Malaria; infectious mononucleosis; hepatitis; bacterial endocarditis;		
	brucellosis; syphilis		
Haematological causes	Anticoagulant therapy; leukemia; lymphoma; myeloma; hemophilia		
Metabolic causes	Amyloidosis; sarcoidosis; Gaucher's disease		
Local cause	Splenic vein thrombosis; splenic artery aneurysm; Pancreatitis		
Other causes	Metastases; prolonged vomiting; repeated coughing; pregnancy;		
	Crohn's disease; after colonoscopy; Electroconvulsive Therapy		

Source: Adapted from (Amonkar & Kumar, 2009)

In addition, its well documented that malaria is the most common cause of pathological rupture of spleen in the tropical countries where life-threatening complications occur in up to an estimated 2% of cases (Agarwal et al., 2013; Gedik, 2008). Most cases of pathological ruptures of the spleen occur during acute malaria seasons.

The following mechanisms are implicated: An increased intrasplenic hyperplasia and engorgement, followed by a compression by the abdominal musculature during physiological activities such as sneezing, coughing and defecation, an abnormally mobile spleen undergoing

intermittent torsion, spasms of the splenic vein as well as vascular occlusion caused by reticular endothelial hyperplasia which may result in thrombosis and infraction (Agarwal et al., 2013; Amonkar & Kumar, 2009; Gedik, 2008; Robert A. Badura, Olivia Oliveira, 2001). A well-published paper by Orloff and Peskin in 1958 described four criteria required to diagnose non-pathological spontaneous splenic rupture (Fernandes et al., 2013; Gazel et al., 2015; Hadary, Dashkovsky, Rapaport, & Cozakov, 2008; Kim et al., 2005; Randriamarolahy et al., 2010). A fifth criterion was added in 1991 by Crate and Payne. These criteria are listed below:

Orloff and Peskin's criteria for spontaneous splenic rupture ;(Crate and Payne's fifth criteria also included)

- 1. No history of trauma or unusual effort, either prior to or on retrospect questioning after operation,
- 2. No evidence of disease that could involve the spleen
- 3. There should be no evidence of adhesion or scarring of the spleen to suggest trauma or previous rupture
- 4. Other than rupture and hemorrhage, the spleen should be normal on gross and histological examination.
- 5. There should be no rise in viral antibody titers in either that acute or convalescent period to suggest viral infection with the types associated with splenic involvement.

In this case all the above criteria's were met. Common symptoms and signs of splenic rupture are left upper quadrant abdominal pain radiating to the left shoulder (Kehr's sign) and dullness to percussion in the left flank and shifting dullness to percussion in the right flank (Balance's sign)(Robert A. Badura, Olivia Oliveira, 2001). This abdominal pain can become generalized, with distension, tenderness and rigidity in later stages, and may be accompanied by pallor, tachycardia, hypotension and oliguria. Eventually, more than half of patients will suffer hemorrhagic shock if the condition is left untreated (Agarwal et al., 2013; Gedik, 2008).

The diagnosis of splenic rupture is based on clinical symptoms and confirmatory diagnostic tests. Some authors have argued that paracentesis is the most effective diagnostic procedure (Aydin, Şenocak, Bilge, Kaymak, & Kilbaş, 2015; Gedik, 2008; Wang, Tu, Li, Luo, & Norwitz, 2011). Paracentesis is not only useful in non-traumatic splenic rupture, but also in patients in whom we think there is intraperitoneal hemorrhage and who are unstable. It is noted that, although a negative result of paracentesis does not prove that there is no hemorrhage, a positive result indicates the possibility of intraperitoneal hemorrhage. Other diagnostic means include ultrasound and computer tomography (CT). Abdominal ultrasound is also a non-invasive and quick way to obtain a diagnosis of intraperitoneal fluid (Gazel et al., 2015; Gedik, 2008; Kim et al., 2005). Abdominal computer tomography scan can clearly show the grade of splenic damage severity and intraperitoneal free fluid (Aydin et al., 2015; de Kubber, Kroft, & de Groot, 2013; Gazel et al., 2015; Gedik, 2008; Kim et al., 2008; Kim et al., 2005).

The differential diagnosis of non-traumatic hemoperitoneum includes unreported trauma, gynecological pathologies, ruptured aortic aneurysm, spontaneous rupture of hepatic hemangioma and hemorrhagic pancreatitis. The case fatality rate is relatively low when an underlying etiology is absent but can be as high as 12% when caused by an underlying disease (de Kubber et al., 2013)⁻

The management of spontaneous or pathological splenic hemorrhagic has been a subject of much debate. There is no doubt that surgical intervention based on splenectomy is a necessity in patients

with serious hemoperitoneum and refractory hypovolemic shock. The emergency splenectomy can be performed via laparoscopic or open "laparotomy". Laparoscopic splenectomy is the preferred surgical approach for benign hematologic disorders when elective removal of the spleen is indicated (Chau, Chan, & Chan, 2008). The main controversy resides in the selection of patients who would benefit from a conservative approach. This is when, the choice between spleenpreserving managements or other surgical methods should be dictated by the degree of rupture. Spleen-preserving should be preferred whenever it is feasible; hence, a less invasive approach such as splenic artery embolization can be performed (Gazel et al., 2015; Mohammed, Majid, & Villatoro, 2016). Finally it should be noted that post-splenectomy administration of polyvalent pneumococcal vaccines in adulthood is debated. This was not done in this case.

Lessons learned

- ✓ Non-traumatic splenic rupture is a rare clinical entity that needs a high index of suspicion for diagnosis;
- ✓ The absence of a history of trauma can make it difficult to reach a diagnosis, which causes delay in treatment ;
- ✓ Rapid diagnosis, aggressive resuscitation and surgical intervention can lead to a successful outcome in patients with spontaneous splenic rupture;
- ✓ Ultrasound is a fast, non-invasive diagnostic tool which quickly assists in the diagnosing of splenic rupture.

Conclusion

This was a case report of spontaneous rupture of a normal spleen in a patient with negative results from malaria peripheral blood smear and antigen for *P. Vivax* and *P. falciparum*. Physicians should consider the diagnosis of spontaneous spleen rupture in patients presenting with abdominal pain and hypotension even without a history of trauma; because an early intervention can be life-saving.

Conflict of Interests: The author declare that they have no competing interests.

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