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*Ann. Ital. Chir.*  
Published online (EP) 23 December 2015  
pii: S2239253X1502438X  
[www.annitalchir.com](http://www.annitalchir.com)

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## Atraumatic splenic rupture: an atypical presentation in a previously healthy patient

*Atraumatic splenic rupture represents a life-threatening abdominal event that requires immediate diagnosis and prompt surgical treatment to ensure the survival of the patient. Atraumatic rupture is relatively uncommon and can occur either in pathological spleens or, more rarely, in normal ones. It has a high morbidity, frequently with few and non specific signs suggesting its presence, can be associated to other pathologies incidentally discovered by imaging. We present a case, successfully treated, of a 51-year-old man, previously healthy, that referred to our hospital for arterial hypertension and abdominal pain; the patient, for an idiopathic splenic rupture with haemoperitoneum, underwent an open splenectomy whose histology examination showed a normal spleen.*

KEY WORDS: Atraumatic splenic rupture, Normal spleen, Spontaneous haemoperitoneum

### Introduction

Blunt thoraco-abdominal trauma is the most frequent cause of splenic rupture (about 90% of cases) and represents a life-threatening condition if late diagnosed and treated<sup>1,2</sup>. Nevertheless many other systemic disorders are likely to determine an atraumatic splenic rupture

(ASR) in pathological spleens<sup>3</sup>. More rarely than the latter are the spontaneous ruptures that occur in healthy spleens; they represent a distinct clinico-pathological entity and are classified as idiopathic<sup>2,3</sup>. Patients may show a plethora of clinical presentations mimicking other medical emergencies (i.e. myocardial infarction, angina pectoris, pulmonary embolism, acute appendicitis, ectopic pregnancy, acute sigmoid diverticulitis and visceral perforation) yet the abdominal pain, localised in the left-side upper quadrant, along with signs of acute bleeding or haemorrhagic shock are the most frequent clinical findings<sup>2,4</sup>. Furthermore, ASR is usually diagnosed late, due to the absence of any history of trauma, and this delay may affect significantly the patient outcome<sup>2,3</sup>. We report the case of a man, previously in healthy condition, who presented for abnormal blood pressure values

*Pervenuto in Redazione Giugno 2015. Accettato per la pubblicazione Settembre 2015.*

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and a drug-resistant epigastric pain; after he developed a haemoperitoneum due to a spontaneous splenic rupture and underwent a successful open splenectomy.

### Case Report

A 51-year-old man was admitted for an isolated epigastric pain associated to palpitations and slightly increased values of blood pressure (155/95 mmHg). He denied history of abdominal trauma, previous major surgery, other pathologies and drug treatments. At physical examination the abdomen was mildly painful only in epigastric region, distended for meteorism, without spleen size enlargement and signs of peritoneal irritation. Laboratory routine tests were all within the normal range (platelet count 187,000/ $\mu$ l and white blood cells count 5,900/ $\mu$ l), and his initial haemoglobin (Hb) value was 14.7 g/dL. Furthermore, electrocardiogram and thorax x-ray were unremarkable. After pharmacological treatment with clonidine and ranitidine, his clinical status improved and blood pressure normalised. Notwithstanding, for the suspicion of a latent coronary syndrome, patient was further observed and monitored with blood and instrumental tests. Five hours later, his clinical condition suddenly deteriorated, with flare of pain, tachycardia and hypotension. His Hb-level shrank to 8.1 g/dL, without any evidence of external bleeding. A contrast enhanced computed tomography (CT) showed a wide subcapsular and intraparenchymal splenic haematoma with an abundant haemoperitoneum (Fig. 1). Radiologist hypothesize a spontaneous rupture of an unknown benign vascular tumour (i.e. haemangioma). Patient underwent an open splenectomy with aspiration of, approximately, 2000 ml of blood from the peritoneal cavity. Both, serological,



Fig. 1: Contrast-enhanced computed tomography (CT), axial plane: the white arrow points at splenic rupture with haemoperitoneum.

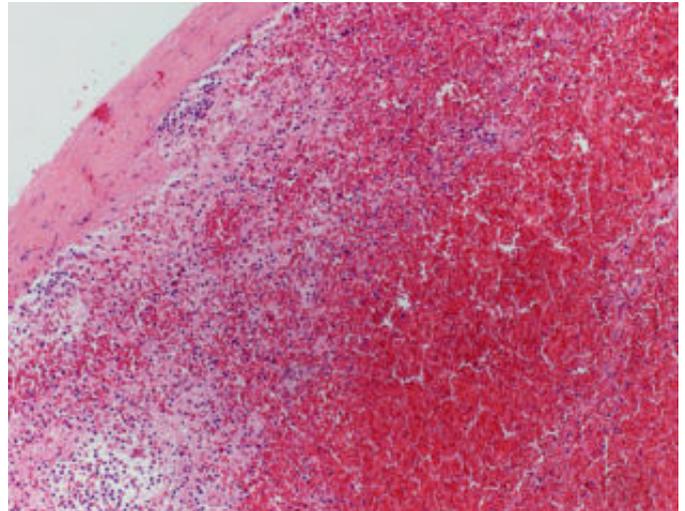


Fig. 2: Subcapsular haematoma of the spleen (haematoxylin-eosin stain, original magnification x 180).

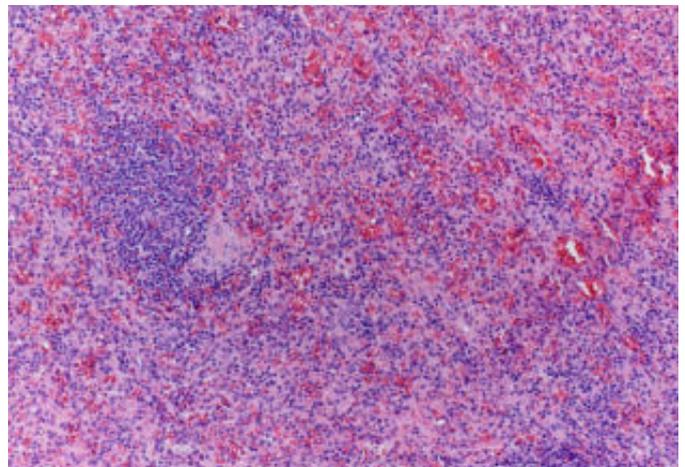


Fig. 3: Normal splenic tissue with blood congestion (original magnification x 200).

virological and haematological tests were also negative. Gross examination of the specimen (measuring 12x11x6 cm and weighing 320 g) showed a fractured spleen with a deep (3.5 cm) and large (6 cm) wound. Histology revealed a normal splenic tissue without underlying any atypia (Figs. 2, 3) and any evidence of benign vascular tumour, thus allowed the diagnosis of idiopathic rupture. His clinical course was uneventful and the patient was discharged in 8<sup>th</sup> post-operative day, with appropriate post-splenectomy antibiotic prophylaxis. Six months later, he was well and free from significant complaints.

## Discussion

The atraumatic splenic rupture is usually due to a large group of diseases, accounting approximately 10% of all splenic ruptures<sup>1,5,6</sup>. ASR, had a male/female ratio of 2:1 and may be divided into two categories: *pathological* concerning abnormal spleens and *idiopathic* occurring in healthy spleens<sup>3,6</sup>. Pathological subtype recognizes the following multiple aetiological causes: neoplastic (i.e. lymphoma, leukaemia, metastases, benign or malignant spleen tumours), infectious (i.e. malaria, infectious mononucleosis), metabolic (amyloidosis, Gaucher's disease), inflammatory non-infectious (i.e. rupture of splenic cyst, infarction of the spleen, splenomegaly and hypersplenism), and drug/treatment-related (i.e. abuse of anti-coagulants and thrombolytics, endoscopic procedures)<sup>1,3,5</sup>. The idiopathic subtype, widely described as rare, had a reported prevalence of about 7% of spontaneous splenic rupture<sup>1-3</sup>. The four criteria to assess the diagnosis of spontaneous splenic rupture were formulated more than fifty years ago and currently are still considered as the gold standard: 1) on thorough questioning either before or after the operation there should be no history of trauma or unusual effort which conceivably could injure the spleen; 2) there should be no evidence of disease in organs other than the spleen, which is known to affect the spleen adversely; 3) there should be no evidence of perisplenic adhesions or scarring of the spleen, which suggest that it has been traumatized or had ruptured previously; 4) other than the findings of haemorrhage and rupture, the spleen should be normal on macroscopic as well as microscopic examination<sup>7</sup>. A fifth criterion was added later: 5) full virological studies of acute phase and convalescent sera should show no significant rise in antibody titres suggesting recent viral infection of types known to be associated with splenic involvement<sup>8</sup>.

The timely ASR recognition has an utmost relevance in order to well-manage patients with predisposing conditions and reduce mortality rate, that may reach 13% in worldwide literature<sup>9,10</sup>. The major risk factors increasing the mortality rate are: enlarged spleen size, neoplastic diseases and age over forty years<sup>3,11</sup>. The spleen rupture can occur in the absence of major trauma or any previously diagnosed splenic disease. Even minor events, such as coughing, vomiting, or sneezing may be cause of pathological spleen ruptures<sup>12</sup>. The typical clinical presentation includes a sudden-onset upper abdominal pain, worse in the left upper quadrant and also referred on the left shoulder (Kehr's sign), abdominal rigidity and hemodynamic instability<sup>1,3,5</sup>. Less frequently is possible to recognize a tender mass in the splenic region (Balance's sign) and non-specific symptoms as nausea, dizziness or syncope may also be associated<sup>1,12</sup>. An atypical presentation or the absence of these signs are likely to lead to the risk of misdiagnosis. Laboratory investigations may show a normal or low Hb-level. Ultrasounds are per-

formed routinely and may be useful, notwithstanding CT scan represents the main diagnosis tool to detect splenic lesions and possible causative pathological processes<sup>4</sup>. Histological features of idiopathic spleen rupture are similar to those found in traumatic ones: subcapsular or parenchymal haematoma, pedicle laceration, and perisplenic haematomas<sup>1,3,12</sup>. Patients with splenic rupture are more frequently treated with total splenectomy, realised by open approach or laparoscopy, in order to remove the bleeding source and make a diagnosis of any underlying disease, despite the correct management is still debated<sup>1,3,10,13,14</sup>. A conservative procedure or spleen-preserving may be carried out only in patients with known underlying splenic cause, whereas the non-operative management (that requires bed staying and fluid resuscitation) can be performed in patients with haemodynamic stability<sup>1,3,5,6,15</sup>.

## Conclusions

ASR represents a rare and potentially life-threatening condition, that should be suspected and considered in the differential diagnosis of the acute upper abdominal pain. The diagnosis may be delayed and difficult to make, especially in patients with atypical features.

Our clinical case highlights that patients with splenic rupture may present unusual symptoms; in such cases a possible early recognition associate to a correct surgical management can be life-saving.

## Acknowledgement

For the accurate revision of the manuscript, We wish to thank Mr Sam Palella, a native speaker, with an extensive experience on scientific papers; furthermore we also thank Dr. Francesco Armaleo who contributed to the data collection and to the clinical case management.

## Riassunto

La rottura splenica non traumatica rappresenta un evento addominale potenzialmente fatale che richiede immediata diagnosi e rapido trattamento chirurgico per garantire la sopravvivenza del paziente. La rottura non traumatica è relativamente non comune e può verificarsi sia in milze patologiche che, più raramente, in milze normali. Essa presenta un'elevata morbilità, frequentemente con pochi ed aspecifici segni che possano suggerire la diagnosi, e può essere associata ad altre patologie incidentalmente scoperte con l'imaging radiologico. Noi presentiamo un caso, trattato con successo, di un uomo di 51 anni, apparentemente in buona salute, visitato presso il nostro ospedale per rialzo pressorio e dolore addominale; il paziente, a causa di una rottura splenica idio-

patica con emoperitoneo, è stato sottoposto a splenectomia laparotomica e l'esame istologico ha mostrato un parenchima splenico normale.

## References

1. Renzulli P, Hostettler A, Schoepfer AM, Gloor B, Candinas D: *Systematic review of atraumatic splenic rupture*. Br J Surg, 2009; 96(10):1114-21.
2. Debnath D, Valerio D: *Atraumatic rupture of the spleen in adults*. J R Coll Surg Edinb, 2002; 47(1):437-45.
3. Aubrey-Bassler FK, Sowers N: *613 cases of splenic rupture without risk factors or previously diagnosed disease: A systematic review*. BMC Emerg Med, 2012; 12:11.
4. Amonkar SJ, Kumar EN: *Spontaneous rupture of the spleen: Three case reports and causative processes for the radiologist to consider*. Br J Radiol, 2009; 82(978):e1111-3.
5. Adachi K, Arima D, Hosaka A, Kiriu T, Sakashita K, Kojima A, Kozai Y: *A non-traumatic splenic rupture leads to diagnosis of underlying abnormality*. Lancet, 2014; 384(9956):1820.
6. Vidarsdottir H, Bottiger B, Palsson B: *Spontaneous splenic rupture and multiple lung embolisms due to cytomegalovirus infection: A case report and review of the literature*. Int J Infect Dis, 2014; 21:13-4.
7. Orloff MJ, Peskin GW: *Spontaneous rupture of the normal spleen; a surgical enigma*. Int Abstr Surg, 1958; 106(1):1-11.
8. Crate ID, Payne MJ: *Is the diagnosis of spontaneous rupture of a normal spleen valid?* J R Army Med Corps, 1991; 137(1):50-1.
9. Carlin F, Walker AB, Pappachan JM: *Spontaneous splenic rupture in an intravenous drug abuser*. Am J Med, 2014; 127(3):e7-8.
10. Roche M, Maloku F, Abdel-Aziz TE: *An unusual diagnosis of splenic rupture*. BMJ Case Rep, 2014; 2014.
11. Abbott RM, Levy AD, Aguilera NS, Gorospe L, Thompson WM: *From the archives of the AFIP: primary vascular neoplasms of the spleen: radiologic-pathologic correlation*. Radiographics, 2004; 24(4):1137-63.
12. Toubia NT, Tawk MM, Potts RM, Kinasewitz GT: *Cough and spontaneous rupture of a normal spleen*. Chest, 2005; 128(3):1884-886.
13. Grossi U, Crucitti A, D'Amato G, Mazzari A, Tomaiuolo PM, Cavicchioni C, Bellantone R: *Laparoscopic splenectomy for atraumatic splenic rupture*. Int Surg, 2011; 96(1):87-9.
14. Bracale U, Merola G, Lazzara F, Spera E, Pignata G: *Spleen rupture: An unusual postoperative complication after laparoscopic cholecystectomy*. Ann Ital Chir, 2013; 84(ePub).
15. Lin WC, Chen YF, Lin CH, Tzeng YH, Chiang HJ, Ho YJ, Shen WC, Chen JH: *Emergent transcatheter arterial embolization in hemodynamically unstable patients with blunt splenic injury*. Acad Radiol, 2008; 15(2):201-8.