Idiopathic Atraumatic Splenic Rupture, an underestimated cause of acute abdomen

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Abstract

Atraumatic splenic rupture is an uncommon but important clinical entity. We report a case of young healthy male who had pain abdomen secondary to atraumatic rupture of spleen. On thorough workup, no conditions affecting the spleen was found and the patient was suspected to have idiopathic rupture.

Introduction

Rupture of spleen is mostly caused by trauma. Infections, neoplasms and coagulopathies represent three major aetiologic groups for such rupture. However, in rare cases, this can occur without any predisposing factors which is then called idiopathic atraumatic splenic rupture.

Atraumatic splenic rupture is rare but life-threatening event, typically not at the top of a clinician's differential for acute abdominal pain, in the absence of trauma. Diagnosing atraumatic splenic rupture can be challenging and it is often missed.

Case presentation

We encountered a previously healthy 20-year-old male, who was brought to the emergency department for unrelenting left sided abdominal pain which started suddenly 3 days earlier. The pain occasionally radiated to the tip of left shoulder. He had few episodes of non-bilious vomiting. He denied any history of recent trauma or trivial injury to the abdomen. He was not on any anticoagulants. He is a non-smoker and does not drink alcohol. He had no personal or family history of leukemia, lymphoma, autoimmune disease or coagulopathies.

On physical examination, he was alert and oriented. He was afebrile with heart rate of 106 beats per minute and blood pressure of 106/72 mm of Hg. There was no lymphadenopathy. The abdomen was tender on deep palpation, however, no rigidity or guarding or peritoneal signs were elicited. Review of other systems was normal.

His hemoglobin was 10.4 gm %, white blood cells count of 5340 cells/mm^3 and platelets of 1,31,000/mm^3. He had normal bleeding and coagulation profile. Blood smear showed no atypical lymphocytes and the differential was otherwise normal. The rest of his routine biochemical investigations including serum lipase and amylase were unremarkable. He had negative blood cultures and negative serologies for Viral hepatitis, Human Immunodeficiency Virus, Enteric fever, Leishmaniasis and Syphilis. Transthoracic echocardiogram was negative for signs of bacterial endocarditis. Abdominal ultrasonography revealed free fluid in peritoneum. CT with intravenous contrast revealed hyperdense (85 HU) areas in peri splenic region suggestive of hematoma along with hyperdense free fluid collection in peritoneal cavity (Figure 1).

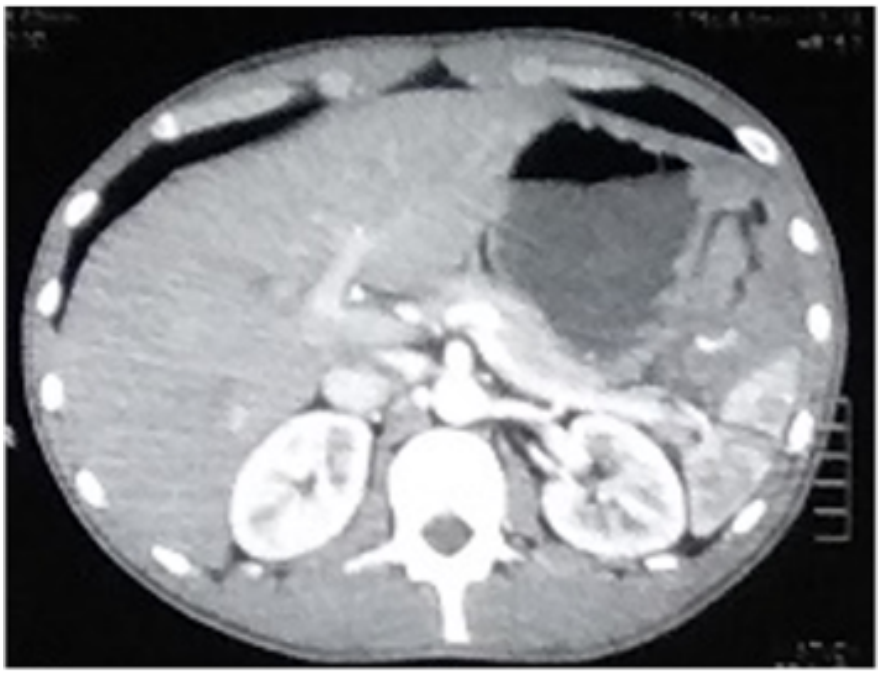

Figure 1: Contrast Enhanced CT scan showing hematoma in perisplenic region

He was managed conservatively with bed rest and other supportive measures. Over the next 5 days, hemoglobin was constant and the patient remained hemodynamically stable. The pain abdomen gradually subsided. The patient did not require blood transfusion or any intervention and was discharged after 5 days. On two-years follow up, the patient is comfortable and has no similar episode of pain abdomen.

Discussion

Splenic rupture is potentially uncommon but lethal cause of acute abdomen with disastrous consequences. The causes of splenic rupture can be generally divided into two categories - traumatic and atraumatic. Atraumatic splenic rupture is termed "pathological" if it occurs in a previously diseased spleen. Idiopathic atraumatic rupture occasionally occurs in a normal appearing spleen without predisposing factors.

The first cases of spontaneous splenic rupture were reported by Rokitansky in 1861 and Atkinson in 1874. The incidence of true atraumatic rupture varies from <1% to 7% with a mortality rate of approximately 12.2%[1]. Recently, Jian in his retrospective review of 251 cases of splenic rupture has shown an incidence of 3.2%[2].

Over 90 % of cases of splenic rupture are pathologic which develops in diseased spleen from wide range of disorders listed in Table 1[3]. A small number of cases, (7% of all atraumatic ruptures), "Idiopathic" rupture may occur in a normal spleen[4]. The exact mechanism of idiopathic atraumatic splenic rupture is poorly understood however two hypothesized mechanisms include:

Hyperplasia of intrasplenic cellular or reticuloendothelial cells leading to parenchymal engorgement and vascular occlusion and

Compression by the abdominal musculature during physiological activities such as sneezing, coughing or defecation[3].

Table 1: Causes of atraumatic splenic rupture

Causes of atraumatic splenic rupture

Abnormal coagulation	Therapeutic anticoagulation -Heparin, Warfarin -Systemic Tissue Plasminogen (tPA) thrombolysis Immune thrombocytopenic purpura, Platelet deficiencies, Uremia
Infections	Malaria, Epstein Barr Virus, Human Immunodeficiency Virus, Cytomegalovirus, Endocarditis, Enteric fever, Babesiosis, Dengue, Syphilis
Hematological malignancies	Acute and chronic myelogenous leukemia, Acute lymphoblastic leukemia Waldenstrom's disease
Solid malignancies	Hodgkin's and Non-Hodgkin's disease, Splenic metastasis, Splenic angiosarcoma
Noninfectious inflammatory disorders	Systemic lupus erythematosus, Polyarteritis nodosa
Miscellaneous	Pregnancy, Acute pancreatitis, Vascular Ehlers-Danlos syndrome, Amyloidosis, Ruptured benign splenic lesions (cyst, infarction, hamartoma, hemangioma)

The diagnosis of idiopathic splenic rupture can be made with Orloff and Peskin criteria[5], when the following four criteria are met:

1. Absence of any history of trauma;
2. Absence of any pre-existing splenic disease
3. Absence of adhesions or scarring in the spleen; and
4. Presence of grossly normal spleen, macroscopically and histologically

Fifth criteria was added by Crate and Payne

Full virological studies of acute phase and convalescent sera show no significant rise in viral antibody titers[6].

Idiopathic atraumatic rupture can occur in any group varying from teenagers and young people to the elderly. Symptom manifestation may vary in patients with splenic rupture. The common presentations include upper or left sided abdominal pain, tenderness, malaise, nausea, vomiting, hypotension, tachycardia, followed at a later stage by generalized abdominal tenderness, peritonism and progressive hemodynamic shock. In 20% of the cases, a sharp radiating pain to the left shoulder (Kehr's sign) was observed[7]. The consistent history is negative for recent trauma or surgery, known disease affecting the spleen, coagulopathies, history of taking anti-coagulants and systemic infection. So, the clinicians should have a high degree of suspicion regarding idiopathic atraumatic rupture on the background of presenting complains, clinical findings and absence of trauma and other causes of splenic rupture.

Management for atraumatic splenic rupture is determined by hemodynamic stability, amount of blood products required, degree of hemoperitoneum and the extent of the splenic injury as described by American Association for the Surgery of Trauma (AAST) scale[8]. Due to risk of post splenectomy infections, there is an increasing trend towards non-operative management Nonetheless, further consideration must be given to the underlying pathology in these cases. Patients with malignant etiology generally require total splenectomy. Patients with non-malignant etiology are primarily managed conservatively which includes bed rest, intravenous fluids and blood transfusions. Strict clinical, laboratory and imaging monitoring is required during non-operative management. Interventional treatment with arterial embolization may also be a non-surgical option in clinically stable patients. Surgery is indicated for clinically unstable patients. If surgery is to be done, splenorrhaphy, partial or total splenectomy can be performed depending on the extent of splenic injury.

Patients should be advised to avoid high impact sports between 1 to 6 months post rupture depending on the degree of injury.

Conclusions

Idiopathic splenic rupture is a very rare, lethal but potentially treatable condition. Emergency physicians should be well aware that splenic rupture with or without hemoperitoneum may occur in absence of trauma and of previously diagnosed diseases involving spleen. It requires a high index of clinical suspicion for immediate diagnosis, appropriate resuscitation and intervention.

Data Availability

Data are available on request from Shankar Adhikari, MS (snr.adhikari05@gmail.com)

Consent

Written consent for the publication of patients' detail was obtained.

Conflicts of Interest

The authors declare no conflicts of interest.

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