Spontaneous Splenic Rupture as the First Presentation of Splenic Marginal Zone Lymphoma: A Case Report

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The most common cause of splenic rupture is trauma. Spontaneous splenic rupture as the first presenting feature of hematologic malignancy, it is a rare phenomenon, is often rapid clinical worsening and considered surgical emergencies. Herein, we reported a case of spontaneous splenic rupture presenting as the first manifestation of splenic marginal zone lymphoma. This patient is discharged without any morbidity.

Key words: spontaneous splenic rupture, splenomegaly, shock, lymphoma

Introduction

Spontaneous rupture of a spleen owing to hematologic malignancy is a rare but life threatening event. It may occur unexpectedly with no previous history of hematologic malignancy. Herein, we reported a case of spontaneous splenic rupture presenting as the first manifestation of splenic marginal zone lymphoma.

Case Report

A 46-year-old previously healthy woman presented to the emergency department with a near-syncope episode and a 7-days history of abdominal pain. On arrival, she had clinical shock with an unmeasurable blood pressure and heart rate 144 beat per minute (sinus tachycardia). She was pale looking, diaphoretic drowsy with a Glasgow Coma Score of E2V2M4. Her abdomen was distended with diffuse tenderness without muscle guarding. Fluid resuscitation was started and a portable abdominal ultrasonography revealed intraperitoneal fluid. Laboratory analyses showed white blood cells (WBC) to be 118100/μL, hemoglobin 7.3 g/dL, and platelets 154000/μL. The WBC differential count showed 3% neutrophils, 8% blast cells, 82% lymphocytes and 7% atypical lymphocytes. Her blood pressure 92/58 mmHg was measured after fluid resuscitation. A computerized tomography scan demonstrated rupture of the spleen and hemoperitoneum (Fig. 1). She underwent an emergency laparotomy within 3 hours of presentation, which showed 4 L of blood in the peritoneal cavity and an enlarged spleen of 1060g (Fig. 2), ruptured with active bleeding. Splenectomy was successfully performed. Pathology confirmed splenic marginal zone lymphoma with high-grade transformation. The postoperative course was without complications and she was discharged smoothly on the tenth postoperative day.
Discussion

The most common cause of splenic rupture is trauma. Spontaneous splenic rupture is a rare phenomenon. It commonly occurs in splenomegaly caused by a pathologic spleen, usually as a result of infection, malignancy, or metabolic disorders.

Spontaneous splenic rupture as the first presenting feature of hematologic malignancy, described only as case report before, is often rapid clinical worsening and considered surgical emergencies \(^{(1-3)}\).

Within the A.A.N. Giagounidis series of 136 cases of pathologic splenic rupture, the acute leukemias were commonest (34%), followed by non-
Hodgkin’s lymphoma (34%) and chronic myeloid leukemia (18%)⁴. The overall ratio of male-to-female is roughly 3:1.

The actual mechanism of spontaneous spleen rupture is not yet fully established, although splenomegaly plays a significant role. There are three pathophysiological hypotheses: infiltration of the spleen by the malignant cells with changes in structure, splenic infarcts leading to areas of reduced resistance, and coagulation disorders inducing intrasplenic and subcapsular bleeding⁴. Few cases of this catastrophic event without splenomegaly (Spleen weight >200 g) have also been reported before.

A combination of abdominal pain and hypovolemic shock features for this acute abdomen. Kehr’s sign, acute referred pain in the tip of left shoulder due to diaphragmatic irritation, is considered a classical symptom. However, it presents only in approximately 17 to 19 percent of patients with spontaneous splenic rupture⁴. The differential diagnosis of spontaneous splenic rupture is extensive. Perforated bowel, perforated gastric ulcer, pancreatitis, aortic aneurism, pulmonary embolus, and acute coronary syndrome all must be considered. Ultrasound and computerized tomography scan scans of the abdomen are both useful to establish the diagnosis. Recent experience favors computerized tomography scan as the initial study of choice, because it can rule out other differential diagnoses and make diagnosis fast and accurate in the emergency department setting⁵.

The aim of this paper is to draw attention to consider spontaneous rupture of the spleen as a dramatic but important abdominal emergency. Early and correct diagnosis and immediate surgical intervention will ensure the patient’s survival.

References

脾臟邊緣的淋巴瘤以自發性脾臟破裂為
初始表現個案報告

嚴勤凱 郭炳讓 林宏榮

外傷是最常造成脾臟破裂的原因，自發性脾臟破裂發生於急性腫瘤雖然少見，但一旦發生卻是極具致命性，在此我們報告一位脾臟邊緣的淋巴瘤以自發性脾臟破裂為初始表現的個案，這種病人必須正確早期診斷，盡快開刀治療才有活命的機會。

關鍵詞：自發性脾臟破裂，脾臟腫大，休克，淋巴瘤