Characteristics of Patients With Spontaneous Splenic Rupture

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In the present study, we aim to share our clinical experience in patients with spontaneous splenic rupture. Splenic rupture without trauma is known as spontaneous splenic rupture. The major problems in the management of spontaneous splenic rupture are missed or delayed diagnosis due to the lack of trauma in most cases. The records of all patients, who were admitted to Cerrahpaşa Medical Faculty, Istanbul University, were retrospectively reviewed from January 2000 to March 2013. Twelve patients were admitted to the emergency department and they were diagnosed with spontaneous splenic rupture. The mean age was 47.6 years. All patients had complaints of abdominal pain. The mean hematocrit value was 22%. Radiologic assessment revealed hemoperitoneum and/or subcapsular hematoma in 8 patients while splenic abscess was diagnosed in 2 patients. Eleven patients underwent splenectomy whereas one was managed conservatively. The most common cause of spontaneous splenic rupture was determined to be use of anticoagulants. Etiology was considered to be idiopathic in 1 patient. Two patients died in the postoperative period. Although rare, spontaneous splenic rupture must be suspected in emergency patients who have used especially anticoagulants and antiaggregants and who have had no recent history of trauma. One of the important causes of mortality is missed or delayed diagnosis.

Key words: Spontaneous – Atraumatic – Splenic rupture – Drug use – Mortality

Although splenic rupture often occurs in association with the trauma, it may appear due to causes other than trauma, and has a fatal course.

Ruptures occurring without trauma are often called spontaneous or atraumatic splenic rupture in the literature.^{1,2} They are extremely rare and the fre-

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quency remains unclear. A study reported the plausible incidence to be 0.1–0.5%.³ It usually develops due to a pathologic cause such as malignancy or infection. It may develop less frequently in a normal spleen without being associated with any pathology and refers to idiopathic splenic rupture.⁴ It is an important problem because it delays the definitive diagnosis in patients without the history of trauma, resulting in disastrous consequences.

The data are poor on the characteristics of the patients, etiology, and incidence of the disease. This study aims to identify the characteristics, etiology, and the diagnostic and therapeutic processes of patients with splenic rupture occurring in the absence of trauma and to highlight our approach to less commonly encountered cases.

Patients and Methods

Study authors evaluated retrospectively the medical recordings of all patients who presented to the emergency department Cerrahpaşa Medical Faculty at Istanbul University between January 2000 and March 2013.

The local clinical research ethics committee approved the study.

Admission notes, operative reports, and pathology results of all patients treated for splenic pathology were examined. Patients who were not included in the study were those who had splenic pathology without traumatic, iatrogenic, or splenic rupture. Rupture of the spleen was regarded as spontaneous splenic rupture (SSR) in a patient who had received low-molecular-weight anticoagulant therapy due to development of pulmonary embolism following elective laparoscopic cholecystectomy 1 week before presenting to emergency department. The patient was included in the study due to uneventful recovery from laparoscopic cholecystectomy, no evidence of iatrogenic injury at the histopathologic examination, and history of anticoagulant use (Patient 7 in Table 1).

The complaints at presentation, clinical findings, medical history, and laboratory and radiologic findings of all patients were documented. Histopathologic findings of the specimens were evaluated in patients who had undergone splenectomy.

Splenectomy materials >200 g or >110 *70 *50 mm in size were considered as splenomegaly.^{5,6}

Results

The medical records of emergency department of our hospital were examined. Twelve patients with the diagnosis of SSR (spontaneous splenic rupture; mean age, 47.66 years; range, 25–79 years; male / female ratio, 1/2) were evaluated. Demographic characteristics of the patients are given in Table 1.

The complaints of all patients were predominantly abdominal pain. Seven patients were admitted to our hospital within 24 hours after onset of complaints. Time of admission of the other 5 patients varied from 3 to 20 days.

When medical history of the patients was analyzed, it was found that 2 patients were diagnosed with and followed-up for lymphoma, 1 with amyloidosis, 1 patient with chronic hepatitis, and 1 with factor XIII deficiency. Four patients had the history of antiaggregant-anticoagulant drug use. One of these patients (Patient 8) had spontaneous rupture of the liver and spleen. One patient (Patient 10) presenting with acute abdomen to the emergency room underwent laparotomy who had chronic hepatitis induced by hepatitis B. At surgical exploration, the spleen was noted to be enlarged; both abdominal and retroperitoneal hemorrhage were present and the source was identified the splenic vein. The medical history of 3 patients (Patients 2, 5, 9) was unremarkable. The mean hematocrit value of 12 patients was 22.2% (range, 17–36%).

Ten patients underwent abdominal ultrasonography and / or computed tomography; eight had hemoperitoneum and / or subcapsular hematoma of the spleen and the findings of 2 patients were suggestive of splenic abscess (Fig. 1). Two patients (Patients 4 and 10) who had not gone radiologic assessment underwent operation on immediately because their general condition was poor and they had acute abdominal condition.

One case was followed-up conservatively and the other 11 cases underwent total splenectomy. All patients but 1 who underwent surgery were operated on within 24 hours after admission. The patient (Patient 2) whose operation was delayed had radiologic findings suggestive of a splenic abscess, and imaging-guided percutaneous drainage was applied; however, when aspiration material proved to contain blood, the actual diagnosis was considered to be splenic rupture.

The patient was a 68-year-old female (Patient 11) who was followed-up without surgery. She presented with a 24 hour history of abdominal pain and use of acetyl salicylic acid due to cardiac problems.

Table 1 Demographic and clinical characteristics of the patients

Patient no	Age (years)	Sex	Medical history	Duration of the symptom	Hct (%)	Diagnostic method	Treatment	Splenomegali	Histo-pathology
1	53	M	Warfarin	3 days	18,8	CT	splenectomy	-	nonspecific
2	73	M	No	<24 hours	26,3	CT	splenectomy	+	nonspecific
3	60	F	Amyloidosis	<24 hours	17	US+CT	splenectomy	+	Amyloid deposits
4	33	F	Hodgkin's Lymphoma	<24 hours	17	Clinal signs	splenectomy	+	Diffuse large B cell lymphoma
5	33	M	No	3 days	21,1	US+CT	splenectomy	+	Hairy cell leukaemia
6	25	M	Factor 13 deficiency	<24 hours	17,6	US+CT	splenectomy	+	nonspecific
7	56	M	LMWH ^a	<24 hours	24,9	CT	splenectomy	+	nonspecific
8	35	M	Warfarin	20 days	22	US+CT	splenectomy	+	nonspecific
9	31	F	No	<24 hours	36	CT	splenectomy	-	Simple cyst
10	26	M	Chronic hepatitis	4 days	20	parasyntesis	splenectomy	+	nonspecific
11	68	F	ASA ^b	<24 hours	21	US+CT	conservative	+	-
12	79	M	Mantle cell Lymphoma	16 days	25	US+CT	splenectomy	+	Mantle cell lymphoma

^aLMWH, low molecular weight heparin.

^bASA, acetyl salicylic acid.

Hematocrit value was 21% and abdominal imaging showed splenomegaly, posterior subcapsular hematoma of the spleen, and accumulation of fluid in the pelvis. Abdominal findings and laboratory improved after fluid and red blood cell replacement.

Five patients had specific findings at the histopathologic examination of splenectomy specimens. Two (Patients 5 and 9) presented with spontaneous rupture of the spleen, 1 of whom had a diagnosis of leukemia and the other had epithelial simple cyst. Six patients had nonspecific findings at histopathologic examination. All cases but 2 (Patients 1 and 9) had splenomegaly.

In our study, 2 patients (Patients 10 and 12) experienced mortality, both of whom were examined due to abdominal pain at outside centers, therefore, we could establish the diagnosis on the fourth and 16th days after presentation, respectively. Splenectomy was expeditiously performed, howev-

er, they succumbed at 4 and 6 postoperative days, respectively.

Discussion

The result of our study shows that, contrary to popular belief, SSR induced by drug use appears to be quite common and delay in diagnosis is a major cause of mortality.

Our identifying a total of 12 cases in the last 13 years has proved that SSR is an infrequently encountered condition also in our hospital, which is one of the leading medical faculties in Turkey that serves a large number of patients. Although it has been reported that the frequency of rupture of the spleen is likely to be 0.1% to 0.5% in the literature,³ the true incidence remains to be unclarified.

When the etiologies of SSRs are examined, neoplastic diseases, infectious diseases, and noninfectious inflammatory diseases rank high on the list.^{1,4} Among the neoplastic diseases, hematologic malignancies account for the largest group.^{1,7} While our 3 patients had SSR associated with hematologic malignancy (25%), there was no rupture secondary to malignancy other than a hematologic one. In our study, there was no infectious disease determined. The fact that we did not identify the etiologic factor that was the focus^{8,9} of a variety of studies could be explained by regional differences.

The most common etiologic cause was the history of anticoagulant-antiaggregant use in our 4 patients (33%). One of these patients had spontaneous rupture of the liver and spleen associated with warfarin use.¹⁰ Ruptures caused by drug use have

Fig. 1 Abdominal computed tomography: axial images that reveal subcapsular hematoma (Patient 2).

been reported 9.1% in the literature.¹ It could be thought that increase in the rates of our study might be attributed to the limited number of our patients.

We had 2 patients (16%) with the history of amyloidosis (Patient 3) and of chronic hepatitis (Patient 10), respectively. We evaluated the patients in noninfectious inflammatory diseases. Cases with splenic rupture associated with chronic hepatitis have been very infrequently reported in the literature.¹¹ Our case was observed to experience both intraperitoneal and retroperitoneal hemorrhage intraoperatively, similar to the one reported from Japan. The source of bleeding was noted to be the splenic vein in this patient with splenomegaly.

A primary epithelial splenic cyst is a rare condition and is often asymptomatic. It may sometimes present itself with such complications as infection or rupture. There are a very limited numbers of cases with SSR associated with epithelial cyst of the spleen in the literature.¹² We established development of SSR associated with simple epithelial cyst of the spleen in 1 of our cases (Patient 9).

It has been reported that no histologic changes or etiologic factors have been detected in atraumatic-idiopathic splenic rupture, and its incidence varies from 5% to 7%.^{1,8} We did not identify any factor that could account for the condition in 1 patient with splenomegaly, and this case was considered to be idiopathic (Patient 2). The prevalence of idiopathic splenic rupture is 8.3% in our study. In contrast to 2 separate studies including a small number of patients,^{4,13} which reported higher incidence of idiopathic spontaneous splenic rupture (4/7 and 3/10, respectively), the results of our study also comprising a small number of patients are consistent with the reviews evaluating many more cases.

All patients but 1 underwent total splenectomy (91%). In a study by Renzulli, the rate was reported at 84.1%.¹ Although there have been studies advocating nonoperative treatment, the rate of operation was 86% in the study.¹⁴ Although there has been no guideline for the treatment of SSR, and total splenectomy is recommended for SSR associated especially with neoplasms, organ-preservation surgery or conservative treatment is recommended for ruptures except for neoplasms.¹ It has been cited that conservative treatment is mostly performed for SSR induced by infectious diseases (approximately 30%) and that the success rate is about 80%.¹ As all of our patients had rupture of the spleen due to causes other than

infection we had a lower rate of conservative treatment.

At the histopathologic examination of the specimens from splenectomy, while 6 patients had no specific findings, 4 patients had a history of drug use, 1 patient had F13 deficiency, and 1 patient had chronic liver disease in their medical history. Therefore, the cause of rupture was ascribed to these conditions.

Two of our patients died (16.6%), the reasons for both deaths were considered to be delay in diagnosis and deterioration of general health condition in association with comorbid diseases (Patients 10 and 12). The mortality rate for SSR has been identified at 12.2% and mortality has been reported to increase significantly especially together with the presence of an underlying neoplastic disease, age (> 40 years) and splenomegaly in the literature.¹ Our 2 patients who died were 26 and 79 years old respectively, both of whom had splenomegaly and the second one had also hematologic malignancy. Of the 3 criteria that are ascribed to mortality only 1 was present in the first patient and the second patient had all 3 criteria, both of whose diagnoses were delayed. Therefore, the reason for mortality, especially the first patient, was thought to be delay in diagnosis.

Conclusions

As a result, despite remaining clinically vague, the diagnosis of spontaneous splenic rupture should be kept in mind in patients without a history of trauma but with predisposing factors such as the history of especially antiaggregant-anticoagulant drug use, and hematologic malignancy. One of the important causes of mortality is delay in diagnosis.

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