Spontaneous rupture of the spleen due to factor XIII deficiency is a rare cause of abdominal pain with disastrous consequences and can be easily confused with other abdominal pathologies until a strong possibility is entertained. The patients usually present in emergency with acute abdomen and diagnosis is difficult. The diagnosis is usually made on high degree of clinical suspicion and falling hematocrit and failure of conservative management or by imaging. The radiologist need to be aware of this potentially fatal complication since early intervention is life saving. CT imaging can also help in conservative management in non critical patients.

Key words: spontaneous rupture of spleen • Computed Tomography • Factor XIII deficiency

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Background
Spontaneous rupture of spleen is a rare but potentially lethal cause of abdominal pain with disastrous consequences. Splenic rupture usually is associated with trauma (blunt or penetrating injuries) and in these conditions the role of imaging is to define the presence and extent of splenic injuries. Spontaneous rupture of spleen, however, can be easily confused with other abdominal pathologies until a strong possibility is entertained especially when a predisposing cause like hematological and neoplastic conditions, tropical endemic disease or other causes of spontaneous rupture of the spleen is present (Table 1). We here report two cases of Factor XIII deficiency that presented with atypical histories and on evaluation revealed spontaneous rupture of normal sized spleen.

Case Report

Case 1

18-year-old male, known case of Factor XIII deficiency on treatment presented in Emergency Department with sudden abdominal pain of one day duration after straining in toilet. He had shortness of breath and palpitations with mild chest pain on left side.

On examination, patient looked tired and pale but was conscious, and well oriented.

On clinical examination, the patient had tachycardia (pulse rate 106/min), Respiratory Rate 43/min. Blood Pressure was 120/66 mmHg on admission he had tenderness in left lumbar region. No obvious ecchymosis or petechiae were noted.

The patient’s investigation revealed Hb – 9.2 gm%, RBC – 5×10^12/ul, WBC – 17.3×10^9/ul with predominant polymorphs (85%) and hematocrit 29.6. The patient was kept under observation with symptomatic treatment but he did not show any relief.

Repeat hemogram at 6 hours showed that Hb had fallen 6.5 g/dl with hematocrit of 20.1 and WBC count 14.6×10^9. The INR was 1.1, PT 13.0 (control range 10.3–12.4) and PTT 30.7. Emergency CT of abdomen was requested.

CT with intravenous contrast revealed evidence of hemothorax and approximately 3.7 cm triangular, non enhancing area in the upper part of spleen extending up to the lateral margin. The splenic size was normal. In view of the clinical background, the diagnosis of spontaneous rupture of the spleen was considered.

The patient was managed conservatively with blood transfusion and Factor XIII cryoprecipitate. The patient stabilized and repeat CT was performed which showed no evidence of arterial blush and no increase in hemothorax.
So the decision to carry on with conservative treatment was taken and the patient was discharged one week later on follow up advice.

Case 2

17-year-old male, known case of Factor XIII deficiency presented to Emergency department with history of sudden abdominal pain for last two days and dizziness and lightheadedness.

O/E patient looked pale, conscious, oriented. The patient revealed mild tachypnea (40/min), pulse was 110/min with BP 180/60 mmHg. The rest of the clinical examination was unremarkable. On admission, his blood profile revealed hemoglobin of 6gm/dl. Reviewing the previous records showed that his hemoglobin had fallen from 13 gm – 6 gm/dl in three days (Patient had visited hematology clinic three days ago).

Subsequently the patient was referred for urgent ultrasound which revealed free fluid in abdomen with a focal hypoechoic area in spleen and left pleural effusion. The differential diagnosis of splenic injury or ruptured abscess was considered and patient was referred for CT for further evaluation.

CT revealed large free fluid accumulation in abdomen with approximately 4cm sized ill defined hypodensity involving the spleen suggesting grade III injury. The diagnosis of spontaneous splenic rupture was suggested.

The patient was treated with blood transfusion, fresh frozen plasma, cryoprecipitate and splenectomy. Postoperative treatment was continued with fresh frozen plasma, cryoprecipitate and was discharged after one week.

Discussion

Spontaneous rupture of spleen requires a high degree of clinical suspicion for immediate diagnosis, appropriate resuscitation and intervention.

Orloff and Peskin[1] suggested following criteria for spontaneous splenic rupture:

a. No history of trauma prior to operation or retrospectively after operation.

b. No evidence of disease that can affect spleen.

c. No evidence of perisplenic adhesions or scarring of spleen.

d. Spleen is normal on gross and histological examination.

e. Crate and Payne added fifth criterion “full viral studies of acute phase and convalescent sera show no significant rise in viral antibody titers [2].

The first cases of spontaneous splenic rupture were reported by Rokitansky in 1861 [3] and Atkinson in 1874 [4]. Weidman in 1927 first used the term spontaneous rupture of spleen – without apparent trauma following insignificant events, including muscular exertion [5,6]. Knoblich in

Table 1. Causes of Spontaneous splenic rupture.

| Infections           | Infectious mononucleosis |
|----------------------|--------------------------|
|                      | Malaria                  |
|                      | Kalazar                  |
|                      | Viral Hepatitis          |
|                      | Subacute bacterial endocarditis |
|                      | Typhoid fever            |
|                      | Tuberculosis             |
| Hematologic and neoplastic causes | Hemophilia            |
|                      | Anticoagulation          |
|                      | Hemolytic anemia         |
|                      | Myeloid metaplasia/fibrosis |
|                      | Lymphoma and leukemia    |
|                      | Multiple myeloma         |
| GI causes            | Crohn’s disease          |
|                      | Pancreatitis             |
| Infiltrative causes  | Amyloidosis              |
|                      | Felty’s syndrome         |
|                      | Sarcoidosis              |

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The etiology of spontaneous rupture of the normal spleen is not known. There are many different speculations regarding the cause of this rare clinical entity, but most of these theories lack strong evidence to support them. These theories include [1,4]:

1. Localized involvement of the spleen with a pathologic process, which upon rupture all evidence of pathologic changes are destroyed.

2. Reflex spasm of splenic vein causing acute splenic congestion.

3. Portal venous congestion with chronic splenic congestion.

4. Abnormally mobile spleen that undergoes recurrent torsions and the resultant congestion leads to rupture.

5. Rupture of a degenerative or aneurysmal splenic artery.

6. Forgotten or unnoticed trauma.

7. Sudden increase in abdominal pressure leads to rupture (i.e., a heavy meal, defecation, lifting, sexual intercourse).

We report two cases of splenic rupture due to Factor XIII deficiency.

Factor XIII deficiency is a very rare autosomal bleeding disorder with frequency 1:2,000,000 in general population. Factor XIII is the last enzyme to be activated in blood coagulation pathway and functions to cross link and alpha and beta fibrin chains resulting in stronger clots. Only 20% of patients experience bleeding in CNS, mouth and muscle or from lacerations and this occur usually without trauma [7].

We could only find two reports in English literature of Factor XIII deficiency who suffered spontaneous splenic rupture [7,21].

Although definite diagnosis can be made only on laparotomy, imaging can help in immediate diagnosis of this condition. Radiologists need to be aware of this possibility especially when confronted with background of hematological diseases. Ultrasound though can raise the suspicion of this entity but can be confused with other diagnostic possibilities as occurred in our case. CT is a rapid diagnostic tool for reliable evaluation of the spleen and follow up as one case was conservatively managed.

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