CASE REPORT

Spontaneous splenic rupture in a patient with congenital factor XIII deficiency: A case report and literature review

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ABSTRACT

Spontaneous (atraumatic) splenic rupture is a rare but potentially life-threatening clinical condition. Most reported cases of spontaneous splenic rupture are associated with splenomegaly due to infectious, neoplastic, or inflammatory diseases. Spontaneous rupture in the absence of these conditions is extremely rare. There is often an underlying trauma history. Factor XIII deficiency, a rare autosomal recessive bleeding disorder, is associated with abnormal fibrin stabilization despite normal routine coagulation parameters. Therefore, it can also lead to defects in wound healing. For the same reason, spontaneous splenic rupture has rarely been reported in patients with Factor XIII deficiency. We report a 32-year-old male with congenital Factor XIII deficiency who presented to the emergency room with hypovolemic shock following sudden-onset abdominal pain. The case was complicated by spontaneous splenic rupture and severe intra-abdominal bleeding. Contrast-enhanced abdominal computed tomography revealed a large perisplenic hematoma. Despite intensive blood transfusion and replacement therapy with fresh frozen plasma and cryoprecipitate, the patient required surgical intervention, and a splenectomy was performed. Histopathological examination revealed no underlying splenic pathology. Spontaneous splenic rupture is extremely rare in patients with factor XIII deficiency, but it should be considered in the differential diagnosis in this patient group presenting with an acute abdomen. Suspicion, early recognition, and supportive care with appropriate blood products and factors, along with prompt surgical intervention, are crucial to prevent mortality.

Keywords: Atraumatic rupture; Factor XIII deficiency; hemorrhage; splenectomy; spontaneous splenic rupture.

INTRODUCTION

The spleen is an intraperitoneal organ located in the upper left quadrant of the abdomen. It is the largest lymphoid organ in the human body and plays essential roles in filtering aged erythrocytes, blood storage, immune surveillance, and lymphocyte production. Splenic rupture most commonly occurs secondary to blunt or penetrating abdominal trauma. Spontaneous (atraumatic) splenic rupture, first described by Orloff and Peskin[1] in 1958, is a rare and potentially fatal condition, often associated with infections, hematologic malignancies, or inflammatory disorders.[1] However, spontaneous rupture in the absence of these risk factors is extremely uncommon and may occur due to congenital or acquired coagulation disorders.

Factor XIII (fibrin-stabilizing factor) is a transglutaminase enzyme involved in the final stage of the coagulation cascade, catalyzing the cross-linking of fibrin monomers to stabilize the clot.[2] Factor XIII deficiency, which may be congenital or acquired, results in defective clot stabilization despite normal standard coagulation parameters. Clinical manifestations include delayed wound healing, recurrent pregnancy loss, and life-threatening bleeding episodes, including intracranial hemorrhage.[3] The global prevalence of congenital Factor XIII deficiency is estimated at 1/3–5 million individuals.[4]

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Herein, we present a rare case of spontaneous splenic rupture in a patient with congenital Factor XIII deficiency, review the diagnostic challenges, and discuss management strategies.

CASE REPORT

A 32-year-old male with a known diagnosis of congenital Factor XIII deficiency presented to the emergency department with sudden-onset abdominal pain and nausea. He reported a similar episode 10 years prior and had last received fresh frozen plasma (FFP) replacement therapy 6 months earlier. He also has an epileptic disorder which is controlled with levetiracetam.

Clinical Examination

On admission, the patient was pale, diaphoretic, and in distress. Physical examination revealed marked tenderness and guarding in the left upper quadrant, progressive abdominal distension, tachycardia (105 bpm), and hypotension (80/45 mmHg).

Laboratory Findings

Initial tests revealed hemoglobin and platelet counts within normal ranges and normal coagulation parameters. White blood cell count was 14.37×103/μL, and C-reactive protein was 12.7 mg/L. His latest factor XIII activity is 10%.

Imaging

Contrast-enhanced abdominal computed tomography (CT) demonstrated irregular splenic margins, perisplenic fluid collection, and a heterogeneous hyperdense lesion (140×110×100 mm), consistent with a large hematoma extending into the mid-abdominal region. The spleen's size and position were normal. Radiological findings are shown in Figure 1.

Management

The patient was admitted to the intensive care unit for hemodynamic monitoring. Preoperatively, the patient received 3 units of packed red blood cells, 2 units of FFP, and 4 units of cryoprecipitate. Despite aggressive fluid and blood component therapy, hemoglobin decreased to 8.7 g/dL, hematocrit to 24.9%, and hypotension persisted. Following stabilization with packed red blood cells, an emergency laparotomy was performed. Intraoperative findings included approximately 2 liters of hemoperitoneum and a laceration at the lower pole of the spleen. Splenectomy was performed. Intraoperatively and postoperatively, an additional 6 units of packed red blood cells, 3 units of FFP, and 4 units of cryoprecipitate were administered.

Histopathology

Microscopic and macroscopic examinations revealed no underlying pathology except for fibrohyalinized tissue and calcifications in the splenic hilum.

Outcome

The post-operative course was uneventful. The patient received appropriate immunizations within 2 weeks and was discharged in stable condition.

DISCUSSION

The concept of spontaneous splenic rupture was first proposed by Orloff and Peskin (1958), who defined four diagnostic criteria: absence of trauma, absence of perisplenic adhesions, no underlying splenic pathology, and normal histology of the spleen.[1] Crate and Payne (1991) later added a fifth criterion – exclusion of viral etiologies.[5] Our patient met all modified criteria for true spontaneous rupture.

Although spontaneous splenic rupture has been reported in association with hematological malignancies, infections, and inflammatory disorders, its occurrence in congenital Factor XIII deficiency is exceedingly rare. Factor XIII deficiency is characterized by impaired fibrin stabilization, leading to de-

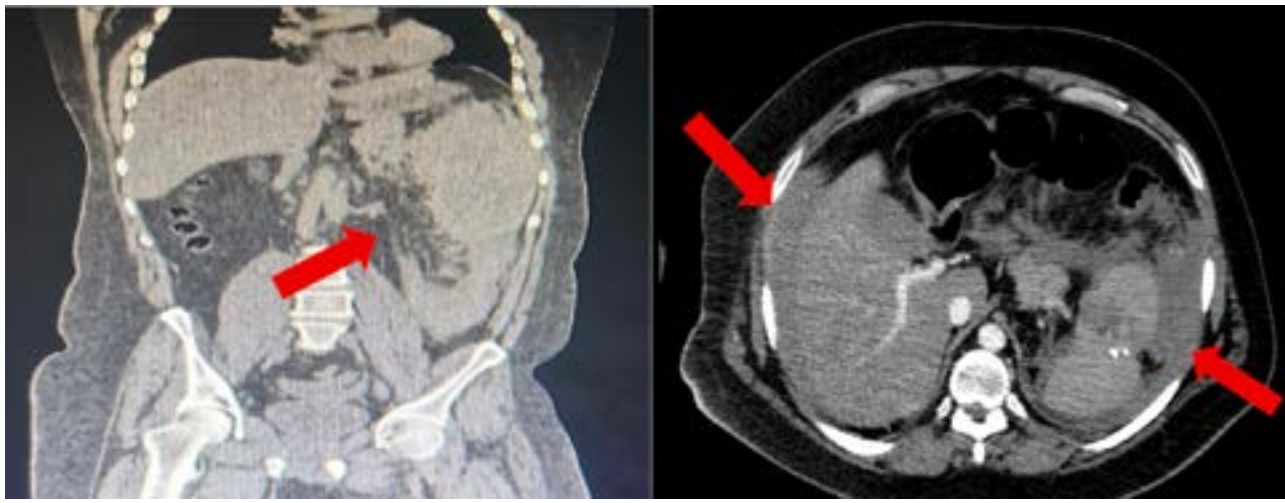

Figure 1. Abdominal computed tomography (CT) of the patient demonstrating free fluid consistent with hemorrhage in the perihepatic and perisplenic regions. Active bleeding from the inferior pole of the spleen due to splenic rupture is indicated by red arrows in both CT slices.

fective clot formation despite normal prothrombin time, activated partial thromboplastin time, and thrombin time. Clinical manifestations range from umbilical stump bleeding in neonates to intracranial hemorrhage in adults.[6,7]

In our case, the absence of splenomegaly and histological abnormalities highlights the role of coagulation instability as the primary mechanism. This suggests that even minor, clinically inapparent trauma may precipitate catastrophic hemorrhage in these patients. Probably, this was the second spontaneous rupture of the spleen. Ten years ago, he experienced a similar clinical scenario. Fortunately, the first rupture was self-limiting. The patient had intellectual disability, but the underlying cause had not been clarified so far. Retrospectively, cranial CT scans were reviewed and revealed multiple cerebromalacic areas. Our hypothesis is that these radiological findings are related to old intracranial hemorrhages, which may also be the origin of the epileptic foci. There was no family history of epilepsy; the patient was the first known case in his family. All these findings were compatible with Factor XIII deficiency and the expected bleeding complications. Fibrogammine is a widely used replacement therapy. However, we were unable to use this replacement therapy because we did not have access to it in our country. We gave the patient FFP and cryoprecipitate support instead of fibrogammine. Thankfully, he was able to recover from the splenic rupture that caused hypovolemic shock with this support.

Management Considerations

Treatment involves aggressive resuscitation, Factor XIII replacement therapy, and prompt surgical intervention when conservative measures fail. Splenectomy remains the definitive treatment for uncontrolled bleeding.

CONCLUSION

Spontaneous splenic rupture is a rare but life-threatening complication in patients with congenital Factor XIII deficiency. Clinicians should maintain a high index of suspicion for atraumatic splenic rupture in these patients presenting with acute abdomen and hemodynamic instability. Early diagnosis and timely surgical intervention are critical for survival.

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Informed Consent: Written informed consent was obtained from the patient for the publication of clinical details and accompanying images.

Conflict of Interest: None declared.

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OLGU SUNUMU - *ÖZ*

Konjenital faktör XIII eksikliği olgusunda spontan dalak rüptürü: Bir olgu sunumu ve literatür derlemesi

Spontan (atravmatik) dalak rüptürü nadir görülen ancak potansiyel olarak yaşamı tehdit eden bir klinik durumdur. Spontan dalak rüptürü bildirilen olguların çoğu enfeksiyöz, neoplastik veya inflamatuar hastalıklara bağlı splenomegali ile ilişkilidir. Bu durumların yokluğunda spontan rüptür son derece nadirdir. Çoğu kez altta travma öyküsü vardır. Nadir görülen otozomal resesif kanama bozukluğu olan Faktör XIII eksikliği, normal rutin pıhtılaşma parametrelerine rağmen anormal fibrin stabilizasyonu ile ilişkilidir. Bu yüzden yara yeri iyileşmesinde de defektlere yol açabilmektedir. Aynı nedenle de Faktör XIII eksikliği bulunan olgularda nadiren spontan dalak rüptürlerinin olduğu bildirilmiştir. Ani başlangıçlı karın ağrısınının ardından acile hipovolemik şokla başvuran, konjenital Faktör XIII eksikliği bulunan 32 yaşında bir erkek hasta sunulmaktadır. Olgu spontane dalak rüptürü ile ağır batın içi kanama ile komplike olmuştur. Kontrastlı karın bilgisayarlı tomografisinde, büyük bir perisplenik hematom saptanmıştır. Yoğun kan desteği ve taze dondurulmuş plazma, kriyopresipitat ile replasman tedavisine rağmen hastaya cerrahi müdahale gerekmiştir ve splenektomi yapılmıştır. Histopatolojik incelemede altta yatan bir dalak patolojisi saptanmamıştır. Faktör XIII eksikliği olan hastalarda spontan dalak rüptürü oldukça nadirdir, ancak bu hasta grubunda akut karın tablosu ile başvurularında ayırıcı tanıda düşünülmelidir. Mortaliteyi önlemek için şüphe etmek, erken tanımak ve uygun kan ürünü ve faktörlerle desteklemek, hızlı cerrahi müdahale etmek çok önemlidir.

Anahtar sözcükler: Atravmatik rüptür; Faktör XIII eksikliği; kanama; splenektomi; spontan dalak rüptürü.

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