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Spontaneous splenic rupture in a patient with factor XIII deficiency

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Abstract

Congenital factor XIII deficiency is exceedingly rare entity with only one in 3-5 million population suffering from the disorder. Spontaneous splenic rupture in factor XIII deficient patients has been reported in only 6 cases to date. We had a 24 years old boy who was a diagnosed case of factor XIII deficiency. He presented to us in emergency room with complains of abdominal pain and distension. He was tachycardic upon presentation. Initial hemoglobin level turned out to be 7.5 g/dl. CT scan of abdomen with IV contrast revealed ruptured spleen with peri-splenic hematoma. He was initially managed non-operatively with close monitoring of hemodynamics and hemoglobin level. His serum factor XIII levels were kept above 50% by cryoprecipitate transfusions. Splenectomy was performed on second day of admission due to failure of conservative management. Previously reported cases show that all patients were males and of less than 20 years of age. Four out of total seven cases including this one were Pakistani in origin.

Keywords

factor XIII; factor XIII deficiency; spontaneous splenic rupture; splenectomy

Introduction

Spleen is a hematological organ lying below the diaphragm in left upper quadrant of the abdomen. It is protected by lower ribs from external trauma. Rupture or laceration is commonly encountered after blunt trauma to the region [1], and is frequently associated with lower ribs fractures on the left side. Enlarged spleen in certain hematological disorders or infectious etiologies is particularly at risk of getting injured even by trivial trauma, and at times without any known physical assault [2]. Spontaneous splenic rupture in patients suffering from congenital factor XIII deficiency is reported only in six cases to date.

Factor XIII also known as Fibrin stabilizing factor is involved in cross-linking of fibrin hence stabilization of clot. Congenital deficiency of this factor is transmitted in an autosomal recessive manner and is exceedingly rare [3]. Only one in 3-5 million [4] suffer from this disorder around the world. The prevalence is reported to be twelve times higher in Iran where consanguineous marriages are common [5]. Owing to lack of national level registry of hematological disorders, prevalence in Pakistan is not known; however it is estimated to be on the higher side as consanguinity is quite common [6].

Case Presentation

24 year old boy diagnosed case of factor XIII deficiency since childhood, presented to emergency room of our hospital with complains of abdominal pain and distension for 2 days. He was referred from another hospital where he underwent transfusion of six units of fresh frozen plasma (FFP). Pain was sudden in onset, mild to moderate in intensity, continuous, not associated with any trauma or unusual physical activity. Abdominal distension was gradually worsening with progressively worsening shortness of breath. Upon presentation, he was tachycardic with heart rate of 120/min while maintaining other vital signs. Abdominal examination revealed generalized tenderness and dullness on percussion.

Hemoglobin (Hb) level upon presentation was 7.5 G/dl. Platelets count and coagulation profile was within normal limits. Factor XIII levels that were checked upon presentation turned out to be 38%. Computerized Tomographic (CT) scan of abdomen and pelvis with and without intravenous contrast was acquired to localize source of bleeding. Images as shown in fig 1 and 2 showed grade III splenic laceration with perisplenic hematoma and hemoperitonium. There was no active contrast extravasation at the time of CT scan. He was admitted to high dependency unit with plan of close observation of hemodynamics and Hb monitoring. Factor XIII levels were checked every 48 hourly and due to non-availability of recombinant factor XIII, cryoprecipitates were transfused to keep levels above 50%.

On second day of admission, patient dropped Hb from 12.1 to 9 g/dl along with increased abdominal girth and tachycardia. Making a working diagnosis of ongoing bleed, patient was taken to operating room for splenectomy. Midline laparotomy was done, about 2.5 liters of altered and clotted blood was evacuated. Ruptured spleen identified and splenectomy performed. Grossly spleen looked normal except the ruptured area. Picture is as shown in Fig 3. Histopathology of the splenic tissue was unremarkable.

Post operatively patient had unremarkable recovery. Immunization was done two weeks after splenectomy. Patient was followed in hematology clinic till six weeks after surgery to keep factor XIII levels above 5% for optimum wound healing.

Discussion

Diagnosis of spontaneous rupture of spleen requires ruling out factors related to splenic laceration. After a comprehensive review of splenic lacerations in 1958 Orloff MJ et al [7] proposed a four points criteria to diagnose spontaneous splenic rupture which was later modified by Crate ID et al [8] in 1991. This modified criteria is as follows:

1. Upon detailed inquiry, either before operation or in retrospect after operation, there should be no history of trauma or of unusual physical activity, which conceivably could injure the spleen.

2. There should be no evidence of disease in organs other than the spleen that is known to affect the spleen adversely and, thereby, could cause the laceration.

3. There should be no perisplenic adhesions or scarring of the spleen, which suggests that it had been traumatized or had ruptured previously.

4. Other than the findings of hemorrhage and rupture, the spleen should be normal on both gross and histologic examination.

5. Full virological studies of the acute phase should show no significant rise in viral antibody titers suggesting recent viral infection of the type associated with splenic involvement.

In our patient none of the above factors were positive. He was already known to have anti hepatitis C virus antibodies positive on screening but any viral hepatitis was ruled out by PCR analysis and liver function tests. We did splenectomy after initial trial of conservative management. Though no guidelines are available for management of spontaneous rupture of spleen, management protocol for splenic laceration after trauma in case of hemodynamic stability recommends initial trial of conservative management and splenectomy is indicated in case of hemodynamic instability [9].

Table 1 shows brief details of previously reported cases. Out of seven cases including ours, five (71%) underwent splenectomy. All reported cases except ours were below 20 years of age and all of them were males. Four patients were of Pakistani origin and in rest of three cases ethnicity was not reported. These findings underscore need for national level registry of hematological disorders.

Role of factor XIII in healing of wound is well established [10]. It is therefore suggested that patients should have adequate levels of factor XIII for at least 6 weeks after any surgical intervention.

Conclusion

High index of suspicion is needed to diagnose spontaneous splenic rupture in known cases of Factor XIII deficiency who present with features of hemorrhagic shock.

National level registry of hematological disorders should be made and maintained to document prevalence of various disorders.

Figures

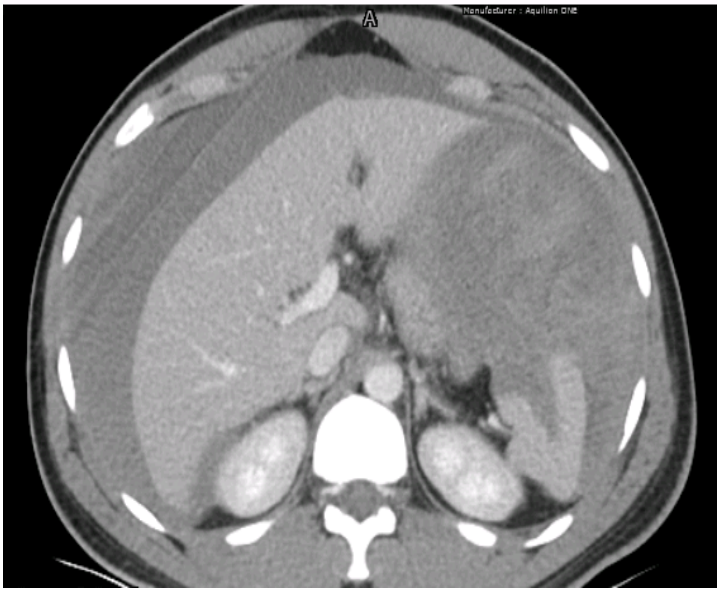

Figure 1: Axil section showing splenic laceration with peri-splenic hematoma and hemoperitonium

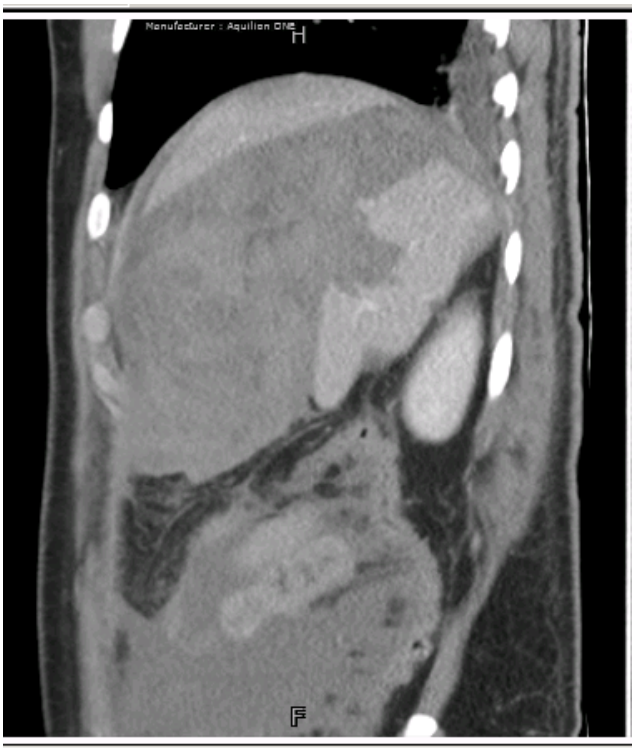

Figure 2: Saggital section showing splenic laceration with peri-splenic hematoma and hemoperitonium

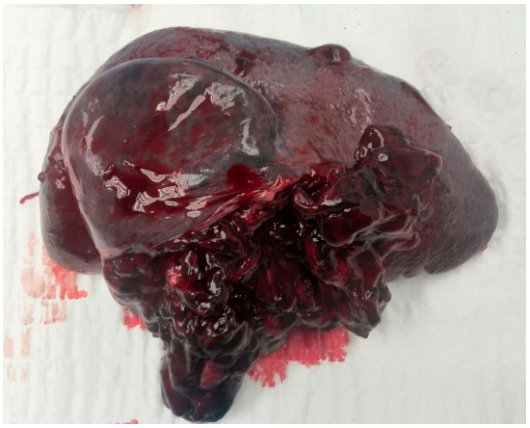

Figure 3: Rupture Spleen

Table

Table 1: Summery of Previous Reports

Author	Year	Number of cases	Age	Gender	Ethnicity	Management
Shariff AH et al [11]	2014	1	17	Male	Pakistani	Splenectomy (Previous episode of spontaneous rupture 8 years back was managed non-operatively)
Bhan P et al [12]	2010	2	18	Male	Not Reported	Conservative
			17	Male	Not Reported	Splenectomy
Khalife H et al [13]	2006	1	7	Male	Not Reported	Splenectomy
Rasul KI et al [14]	2009	2	19	Male	Pakistani	Conservative
			17	Male	Pakistani	Splenectomy

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