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Case Report

Great Role of Abdominal U/S in Management of Splenic Sequestration Crisis

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Abstract

Acute splenic sequestration crisis (ASSC) is a rare complication in adults with sickle cell disease that is diagnosed clinically by means of sudden splenic enlargement and a rapid fall in hematocrit.

The spectrum of severity in this syndrome is wide, ranging from mild splenomegaly to massive enlargement, circulatory collapse, and even death. The diagnosis is usually clinical, based on the enlargement of the spleen with a drop in hemoglobin level by > 2 g/dl.

Unfortunately, this drop of HB and splenomegaly can happen with hemolytic crisis which is common occurrence in SCD, moreover we can see the picture of hemolysis in the sequestration crisis.

Although it is rare that imaging studies are ordered in sequestration crisis, however, in the patient who presents to the emergency department with non-specific findings of an acute abdomen, it is important to recognize the appearance of sequestration on imaging studies.

Abdominal U/S is accessible, noninvasive, safe, cheap, and easily available in ICU nowadays. We use whole body ultrasound in our ICU in management of critically ill patients since long time, recently we admitted a case of SCD with possible severe sequestration crisis as evidenced by marked drop of HB and splenomegaly.

Bed side abdominal U/S was done in ER and revealed marked splenomegaly.

But splenomegaly alone can occur in other SCD crisis and in vast majority of diseases, so we examined the splenic arterial circulation by color and spectral Doppler.

Amazingly we found a unique Doppler flow which is going with sequestration.

Normally splenic circulation is low resistance circulation with a diastolic flow normally above the baseline and appear red in color Doppler because the flow is towards the probe.

In our patient during the sequestration there was blockage of blood flow through the splenic venules which lead to increase resistance to diastolic flow, so we found loss as well as reversed diastolic flow in the splenic artery branches which appeared below baseline in spectral Doppler and appear blue color on color Doppler.

Interestingly, when the patient received blood and the HB S dropped the splenic blood flow had improved in some splenic areas and on repeating splenic spectral Doppler, the diastolic flow became above the baseline denoting forward flow.

So, in our case there was an amazing correlation between sequestration and splenic artery flow examined with Doppler.

In our case, we found also a very important finding in abdominal U/S, we found blood inside the abdomen by positive FAST study and hematocrit sign of intraabdominal fluid, which proved to be upper splenic pool rupture during surgery.

The findings of abdominal U/S were correlated with the abdominal CT with contrast which revealed large heterogeneous spleen with hypodense areas.

At the end we believe that abdominal U/S including Duplex study of splenic vessels should be done in every suspected severe case of sequestration crisis in SCD.

Keywords: *Acute splenic sequestration crisis (ASSC); Abdominal U/S*

Introduction

Normally, flexible and smooth red blood cells pass swiftly through the spleen. The spleen receives about 3 to 5% of the total blood volume per minute.

In most older children and adults with HbSS disease, repeated sickling in the spleen and local infarction (tissue death) eventually results in scarring, fibrosis, and a non-functional spleen (called functional asplenia or auto splenectomy).

Normally splenic artery flow is a low resistance flow with a diastolic flow appear above baseline in spectral Doppler with resistivity index less than 0.7.

During splenic sequestration there will be trapping of sickled RBCs inside the splenic venules with marked increase in splenic size.

We see a large number of SCD patients with different crisis here in Saudi Arabia Eastern province where the prevalence of SC Trait 2 - 27%, and up to 1.4% had SCD in some areas [1].

We use abdominal U/S in our ICU to assess the spleen in case of suspected sequestration to look for splenic size as well as color and spectral Doppler of the splenic vein and artery.

We find a case of splenic sequestration and followed up his spleen with abdominal U/S daily while in ICU.

In the first day the spleen was markedly enlarged and homogenous, splenic artery color Doppler revealed reversed diastolic flow which was confirmed with spectral Doppler with diastolic flow below baseline.

In second day, splenic infarction started which appeared clearly with heterogeneous appearance and scattered hypo echoic areas in U/S, splenic arterial flow was almost absent.

In the coming days the splenic infarctions appeared very clearly by increasing heterogeneity and appearance of echogenic scar areas in U/S.

This was confirmed by CT abdomen with contrast.

Moreover, on admission in the first day abdominal U/S revealed free fluid with hematocrit sign which proved to be blood by CT abdomen and drainage which was due to splenic rupture appear after splenectomy.

Case Presentation

A 21-year-old male patient K/C of SCD came to our hospital after staying in another private hospital for 2 days because of vasoocclusive crisis, he received opioid analgesia and IVF.

Patient came to us complaining of symptoms of anemia [malaise, weakness, dizziness, and palpitation] and severe generalized body ache.

General Exam

Temp 37.5, HR 145/min sinus rhythm, BP 110/60, RR 40/min, 0_2 sat 99% on 3litrs nasal cannula.

Chest: Clear.

Heart: sinus tachycardia.

Abdomen: distended with splenomegaly.

CBC on presentation: HB 3.7gm/dl, Plat 15000/cmm, WBCS 5400/cmm, RI 0.53.

Serum LDH 2196 IU/L, Lactate 4.43 mmol/l, PT 16.1, PTT 26.2, Liver and renal function: normal.

Abdominal U/S on presentation

Spleen: Homogenous texture, markedly enlarged, Color Doppler splenic artery revealed reversed flow in diastole which is confirmed with spectral Doppler.

Moderate free fluid in the abdomen with haematocrit sign denoting bleeding.

CT in the first day revealed homogenous large spleen and intraperitoneal fluid.

CT in day 3 revealed splenic infarction.

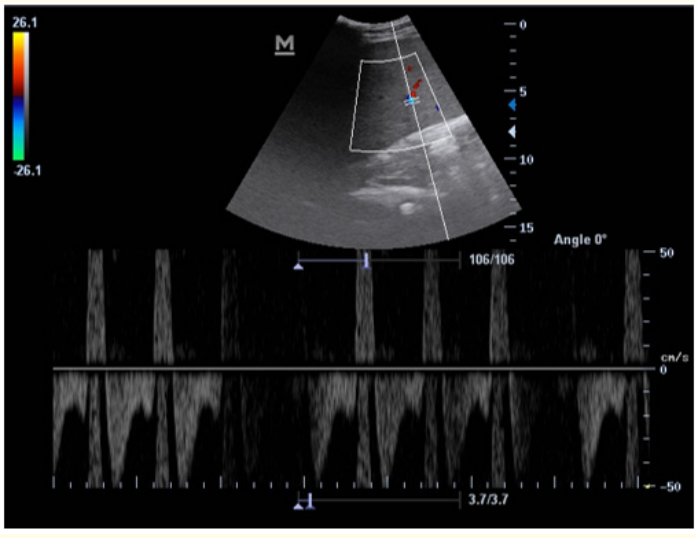

Figure 1: *Spectral Doppler of splenic artery branch reveal.*

Figure 2: *Reversed diastolic flow denoting sequestration.*

Figure 3: *Splenic infarction.*

Patient was stabilized with blood and platelets transfusion, with drop of HBS down to 6%.

Repeated Splenic artery spectral Doppler revealed forward diastolic flow in some splenic areas denoting improved flow with blood transfusion.

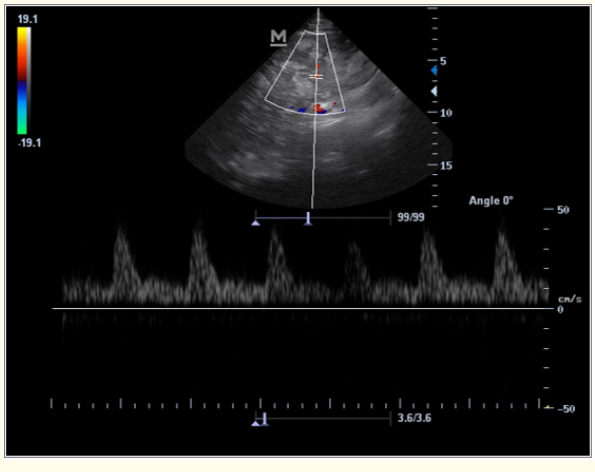

Figure 4: *Forward diastolic flow above baseline in spectral Doppler of splenic artery.*

After stabilization, patient has undergone splenectomy.

Histopathology revealed:

1. Extensive necrosis.
2. Viable splenic tissue [congestion and fibrosis]
3. 2 reactive lymph node.

Patient was discharged from the hospital in good shape and is following with the surgical team.

Discussion and Conclusion

Sickle cell disease (SCD) is a genetic disorder in which there is an alteration in the normal globin chain. It is characterized by red blood cells (RBCs) with abnormal hemoglobin resulting in rigid sickling of the cell, leading to vascular occlusion, and ischemia in multiple organs [2].

A common splenic complication is the sequestration syndrome, which is believed to be the cause of splenomegaly in SCD patients. It consists of a rapid pooling of blood within the spleen; the blood pooling may be larger than expected for a child or adult with sickle cell anemia, resulting in intravascular volume depletion which may progress rapidly to cardiovascular collapse and death [3].

Other diseases that may cause splenomegaly include infectious or granulomatous disease, malignancy, congestive conditions, and other hematologic diseases [4].

So, the question here how to differentiate between splenomegaly due to sequestration or other causes.

It is the role of Doppler [color, spectral] study of splenic artery and its branches, we found in this case study a good correlation between the loss as well as the reversed diastolic flow in splenic arterial circulation and the sequestration crisis, moreover the diastolic flow improved after blood transfusion and drop of HB S.

Abdominal U/S also revealed in this case the presence of intraabdominal bleeding which was due to a rare complication of splenic rupture and can exacerbate the anemia of sequestration.

All previous sonographic studies of spleen in splenic sequestration revealed hypoechoic areas with heterogeneous spleen [5,6].

Ultrasound is a simple, affordable, and easily accessible imaging modality that plays an important role in early detection of the signs of splenic sequestration both by 2D appearance of splenomegaly and heterogeneous echotexture and Doppler study of splenic artery for signs of splenic death like brain death [absent as well as reversed diastolic flow in this low resistance circulation of the vital organs.

Future Recommendation

We need large study with large number of SCD patients with splenic crisis and to follow them by abdominal U/S including Doppler study to clearly know what is going on in this fatal sequestration crisis.

Bibliography

1. Lehmann H., *et al*. "Distribution of sickle-cell hemoglobin in Saudi Arabia". *Nature* 198 (1963): 492-493.
2. Ma'aji SM., *et al*. "Transabdominal ultrasonographic findings in children with sickle cell anemia in Sokoto, North-estern Nigeria". *Nigerian Journal of Basic and Clinical Sciences* 9.1 (2012): 14-17.
3. Rusheke AH. "Abdominal Ultrasonographic Abnormalities in Patients with Sickle Cell Anaemia at Muhinbili National Hospital". M. Med (Radiology) Dissertation, Muhimbili University of Health and Allied Sciences (2010).
4. Al-Salem AH., *et al*. "Sonographic assessment of spleen size in Saudi patients with sickle cell disease". *Annals of Saudi Medicine* 18.3 (1998): 217-220.
5. Gaels J Lonergan., *et al*. "Sickle cell anemia". *Radiographics* 21.4 (2001): 971-974.
6. JE Roshkow., *et al*. "Acute splenic sequestration crisis in two adults with sickle cell disease: US, CT, and MR imaging findings". *Radiology* 177.3 (1990): 723-725.

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