Journal of Pharmaceutical Research International

33(47B): 17-21, 2021; Article no.JPRI.76044
ISSN: 2456-9119
(Past name: British Journal of Pharmaceutical Research, Past ISSN: 2231-2919, NLM ID: 101631759)

A Case of Budd-Chiari Syndrome – As an Unusual Early Manifestation of Systemic Lupus Erythematosis with Antiphospholipid Syndrome and Deep Vein Thrombosis

Gitanjali Dasari[1*], S. Selvamani[1], E. Dhivya[1] and V. R. Mohan Rao[1]

[1]Department of General Medicine, Chettinad Hospital and Research Institute, Kelambakkam, India.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/JPRI/2021/v33i47B33090
Editor(s):
(1) Dr. P. Veera Muthumari, V.V. Vanniaperumal College for Women, India.
Reviewers:
(1) Vaibhav Anjankar, Jawaharlal Nehru Medical College, DMIMS, India.
(2) Francesca D'Auria, AOU Maggiore della Carità of Novara, Italy.
Complete Peer review History: https://www.sdiarticle4.com/review-history/76044

Case Study

Received 11 August 2021
Accepted 23 October 2021
Published 30 October 2021

ABSTRACT

Budd-Chiari syndrome occurs due to obstruction of hepatic venous out-flow which in turn produces intense congestion of the liver. Systemic Lupus Erythematosus (SLE), which is an auto-immune disorder has protean manifestations such as easy fatiguability, arthralgia, photosensitivity, malar rash, fever, alopecia but our case had an unique association of ascites and hepatosplenomegaly due to Budd Chiari syndrome stemming from the root cause of Antiphospholipid antibody syndrome (APLA) is very rare. SLE accounts for 40% of the cases of APLA. SLE presenting with Budd-Chiari syndrome as an early manifestation is unusual and is rarely reported in the literature. Here we report a rare case of a young female who presented with abdominal distension, abdominal pain and fever diagnosed to have Budd-Chiari syndrome as a presenting feature of SLE and on further evaluation she was also found to have antiphospholipid syndrome secondary to SLE.

Keywords: Antiphospholipid syndrome; SLE; Budd-Chiari syndrome.

**Corresponding author: E-mail: drdg0709@gmail.com;*

1. INTRODUCTION

Autoimmune disorders like Systemic Lupus Erythematosus (SLE) usually presents with fatiguability, malar rash, fever, arthralgia, photosensitivity, alopecia but rarely presents with Budd-Chiari syndrome stemming from the root cause of Antiphospholipid antibody syndrome (APLA). Budd-Chiari syndrome is caused by obstruction of hepatic venous outflow which produces intense congestion of the liver [1]. In Budd-Chiari syndrome the smaller hepatic veins are often spared while involving all the three major hepatic veins. Antiphospholipid syndrome is characterised by the production of autoantibodies directed against phospholipids and phospholipid binding plasma proteins associated with multiple thromboembolic events. The common manifestations are recurrent foetal loss followed by portal vein thrombosis, mesenteric thrombosis, iliofemoral thrombosis and lastly Budd-Chiari syndrome [2]. SLE accounts for 40% of the cases of APLA. SLE presenting with Budd-Chiari syndrome as an early manifestation is unusual and rarely reported in the literature. Here, we report a young female who presented with abdominal distension, abdominal pain and fever diagnosed to have Budd-Chiari syndrome. On further evaluation, she was found to have antiphospholipid syndrome secondary to SLE.

2. CASE REPORT

A 21 year old married female with no known co-morbidities, presented with abdominal distension which was progressive over the period of 2 weeks which was associated with dull aching poorly localized abdominal pain. There was initially no history of swelling of legs, joint pain, photosensitivity, oral ulcers, jaundice, hematemesis, melena or altered sensorium. H/o cough with expectoration since 15 days which was mucoid in consistency. H/o vomiting since 15 days, 2-3 episodes/day, containing food particles, non-bilious, non-blood stained, or non-projectile. H/o recurrent headache since 2 weeks, on and off, bilateral and associated with nausea. She noticed right lower limb pain and swelling which was associated with low grade fever for past 2 days. H/o amenorrhea present for 3 months. Patient had H/o urinary tract infection 15 days back and was treated with antibiotics; details not available. H/o seizures 2 episodes at the age of 16 years and was not evaluated. H/o jaundice at the age 6 years and was treated and no further episodes of jaundice. H/o blood transfusions 2 times in the past 5 years; not fully evaluated. H/o weight gain (10 kgs in a month). H/o decreased appetite and disturbed sleep. On examination, Patient was conscious, coherent and oriented. She had pallor and unilateral tender pitting pedal edema of the right lower limb. Her heart rate was 123/min, BP was 100/80mmHg, respiratory rate was 21/min, SPO2 was 99% in R.A and JVP was not elevated. On systemic examination abdomen was distended, flanks were full, umbilicus was shifted downwards. She had tender hepatomegaly with a liver span of 19 cm and shifting dullness was present. Other systemic examination was normal.

Laboratory findings were as enclosed in Table-1. Peripheral smear shows microcytic hypochromic, anisopoikilocytosis, elliptocytes, platelets around 1 lakh/cu.mm. C-reactive protein (mg/dl) – 78.45(reactive), direct coomb's test – positive. QBC for MP, smear for MP/MF, dengue serology and urine pregnancy test were negative. Ascitic fluid analysis revealed high SAAG high protein ascites with normal cytology and no growth of organisms. Thyroid function test were normal.COVID-19 RTPCR, blood cultures and sputum for AFB were negative. USG abdomen showed hepatomegaly with liver size of 19cm with early chronic liver disease, moderate ascites, splenomegaly with normal status of the kidneys. CECT abdomen showed thrombosis of the right hepatic vein (Fig-1), features suggestive of Budd-chiari syndrome and a small splenic infarct. HRCT chest showed no significant abnormality detected. ECG shows sinus tachycardia. Lower limb venous Doppler study was done which revealed right lower limb deep vein thrombosis involving ilio-femoral veins. Thromboelastography showed a hyper-coagulable state. Platelet function assay showed prolongation of collagen/epinephrine and collagen/ADP test results ruling out aspirin induced platelet dysfunction and reinforcing the presence of anaemia and thrombocytopenia. ANA was positive and shows 3+ homogenous pattern. IgM and IgG antibodies to beta -2 glycoprotein complex, lupus anti-coagulant and anti-cardiolipin were positive. Ds-DNA (ELISA)(IU/ml) - 809 .C3 level(mg/dl) - 77.3 .c4 level(mg/dl) - 7.3. She was started on therapeutic anticoagulation and venoplasty of the right hepatic vein was done after the resolution of thrombocytopenia. On 1 month of follow up, she had improved clinically and there were no further thrombotic episodes. She is currently on anticoagulation, diuretics for ascites, hydroxychroloquine and prophylactic measures

for chronic liver disease. She has been vaccinated against hepatitis-B, streptococcus pneumonia and haemophilus influenza. She is on constant follow-up for the same. Patient is planned for repeat antiphospholipid antibody panel in near future.

Table 1. Investigations done on day -1 and day-3 of admission

Test	ON day 1	ON day 3
HB	7.8	8.1
PLATELET COUNT	67,000	32,000
ESR(mm)	83	
PT(seconds)	Control - 11.6, Test - 19.10	Control – 11.7-16.1,Test – 15.2
INR	1.66	1.29
AST(U/L)	59	52
ALT(U/L)	25	27
URINE ALBUMIN	TRACE	3+
URINE RBC'S		8-10/HPF
APTT(seconds)	Control – 26.4, Test – 50.30	Control – 27.8 – 40.4, Test - 61

Table 2. Investigations done on day – 1,2 and 3 of admission

TEST	Day 1	Day 3	Day 5
IRON (mcg%)		28	
TIBC (mcg%)		285	
VITAMIN-B12 (pg/ml)		710	
FOLIC ACID(ng/ml)		4.3	
FIBRINOGEN (mg/dl)	510		
24HRS URINE VOLUME			1040
24HRS URINE PROTEIN			1600

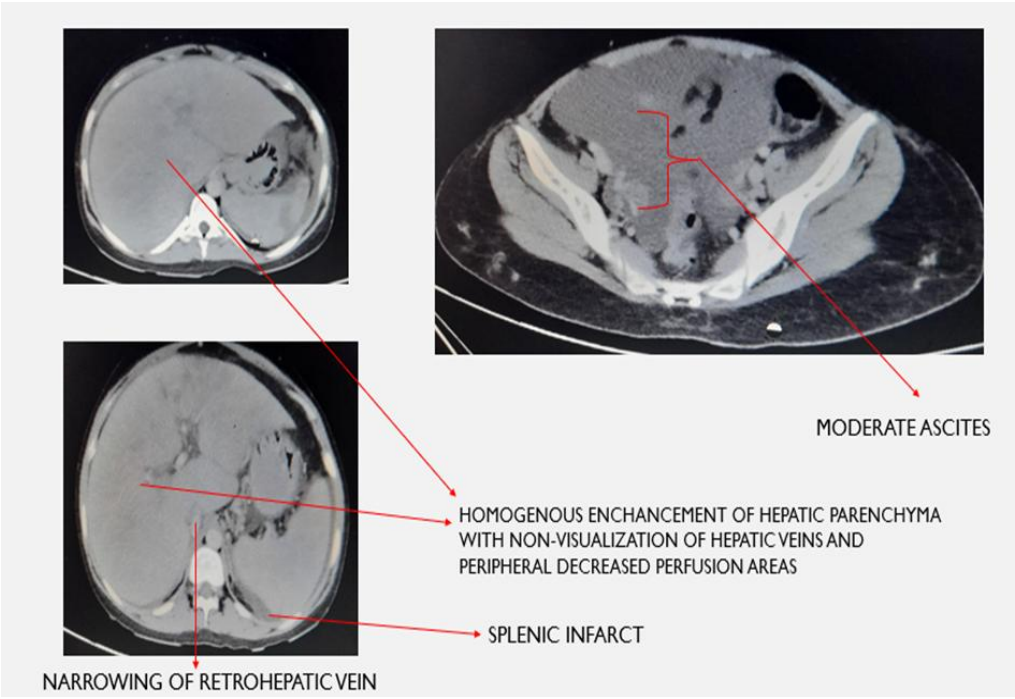

Fig. 1. CECT abdomen showing thrombosis of the right hepatic vein

3. DISCUSSION

Budd-Chiari syndrome occurs due to the obstruction of hepatic venous out-flow which in-turn produces intense congestion of the liver. Smaller hepatic veins are often spared in budd-chiari syndrome. Polycythemia rubra vera, aspergillosis , filariasis, amoebic liver abscess, schistosomiasis, membranous obstruction of IVC, adrenal adenoma, Hepatocellular carcinoma, renal cell carcinoma, antiphospholipid syndrome, leiomyosarcoma of IVC, pregnancy, HRT, oral contraceptives are some of the causes of budd-chiari syndrome.

Clinically patients may have abdominal pain, abdominal distension, weakness, anorexia, jaundice, abdominal venous distension, massive ascites, hepatomegaly, splenomegaly and oedema of thighs, legs and feet. Antiphospholipid syndrome is an autoimmune disorder characterised by the formation of auto-antibodies against the phospholipids leading to the occurrence of multiple thrombotic events [3].

Another rheumatic, autoimmune and multi-sytemic disorder is systemic lupus erythematosus characterised by the presence of auto-antibodies. The association of SLE with Antiphospholipid Syndrome (APS) is about 36% in reported cases [4].

The association of antiphospholipid antibody syndrome (APLA) and SLE with Budd-Chiari syndrome is very rare and only few cases have been reported in literature. Most of the cases reported till now had clinical manifestations of SLE before they were diagnosed with Budd-Chiari. Our patient presented with Budd-chiari which on further work up was found to have SLE Espinosa G et al.,[5] Jayabal et al., and Ilkgül O et al., also provided the data with the similar findings regarding the association of Budd- chiari syndrome as an early manifestation of SLE.. Patient with secondary antiphospholipid syndrome should be kept on long term antiplatelet or anticoagulants therapy to maintain the target INR between 3 to 4 [6]. Side to side porto-caval shunting can be done if there is inadequate response to medical therapy and in case of shunt failure [7],[8], Ortho topic liver transplantation can be considered. This is because of the thrombosis of the portal vein, splenic vein, and superior mesenteric vein which results in unshuntable portal hypertension.

4. CONCLUSION

Any patient presenting with features of Budd-chiari syndrome, should always be screened for Antiphospholipid syndrome as well as for SLE even in the absence of the other clinical features prior to anticoagulation therapy and steroids especially in young females[9]. Early screening and diagnosis will prevent the future complications like repeated miscarriages and other multi- systemic complications.

CONSENT AND ETHICAL APPROVAL

As per international standard or university standard guideline Patient's consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Pandiaraja J, Sathyaseelan A. Budd-Chiari Syndrome as an Initial Manifestation of Systemic Lupus Erythematosus. J Clin Diagn Res. 2016;10(4):OD01-OD2.
2. Dizkucukkaya R, Demir K, Yenerel MN, Nalcaci M, Kaymakoglu S, Inanc M, Coexistence of homozygous factor V Leiden mutation and antiphospholipid antibodies in two patients presented with Budd-Chiari syndrome *Haematologia (Budap)* 2002;32(1):73-77.
3. Kuhn A, Bonsmann G, Anders H-J, Herzer P, Tenbrock K, Schneider M, The Diagnosis and Treatment of Systemic Lupus Erythematosus *Dtsch Ärztebl Int* 2015;112(25):423-32.
4. A case of antiphospholipid antibody syndrome with Budd-Chiari and colonic ulcers complicated with gastrointestinal hemorrhage.Ilkgül O, Içöz G, Dayangaç M, Tokat Y, Ozütemiz O Turk J Gastroenterol. 2004;15(2):115-6.
5. Espinosa G, Font J, García-Pagan JC, Tàssies D, Reverter JC, Gaig C, et al. Budd-Chiari syndrome secondary to antiphospholipid syndrome: clinical and immunologic characteristics of 43 patients. Medicine (Baltimore) 2001;80(6):345–54
6. Duarte C, Couto M, Inês L, Malcata A. [Budd-Chiari syndrome and severe

thrombocytopenia in a patient with Systemic Lupus Erythematosus and Secondary Antiphospholipid Syndrome] Acta Reumatol Port. 2007;32(2):175–80.

7. Pavri TM, Herbst A, Reddy R, Forde KA. Budd-Chiari syndrome: a single-center experience. World J Gastroenterol: WJG. 2014;20(43):16236–44.
8. Rizzati R, Sala S, Tilli M, Marri I, Galeotti R. Budd-Chiari syndrome associated with systemic lupus erithematosus: diagnostic imaging and percutaneous treatment. Radiol Med (Torino) 2003;105(5-6):519–23.
9. Joffe GM, Aisenbrey GA, Argubright KF. Budd-Chiari syndrome, systemic lupus erythematosus, and secondary antiphospholipid antibody syndrome in pregnancy. Obstet Gynecol. 2005;106(5 Pt 2):1191–94.

Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle4.com/review-history/76044