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Case Report

Systemic Lupus Erythematosus Complicated by Portal Vein Thrombosis: A Case Report

*Abdal SJ[1], Shahin MA[2], Islam MA[3]

Abstract:

A 24-year-old female got admitted in a remote hospital with one and half month previous history of gradually increasing swelling of the abdomen followed by swelling of the both legs and face without any discernable etiology. Later she is diagnosed as a case systemic lupus erythematosus (SLE) based on clinical features and serology. She had neither any history of having pro-thrombotic risk factor nor any history of deep vein thrombosis. Her Anti-β2 Glycoprotein 1 (Ab IgM & IgG) was marginally positive. Her ultra-sonogram of abdomen showed moderate ascites, gross thrombosis & stenosis in portal system (main trunk of portal vein, both proximal branches of portal vein, splenic vein at pancreatic area, superior mesenteric vein *at confluence) that were 100% blocked that also supported by CT scan of abdomen and Doppler ultra-sonogram. Her Doppler ultra-sonogram of lower limbs were normal. She was treated initially with albumin infusion, enoxaparin, warfarin, aspirin, hydroxychloroquine and medium dose prednisolone.*

Keywords: *portal hypertension, systemic lupus erythematosus, anticardiolipin antibody*

INTRODUCTION

Thrombotic events are well-known complications of systemic lupus erythematosus (SLE) and have been largely related to the presence of the lupus anticoagulant.[1,2] Portal vein thrombosis (PVC) results from a combination of local and systemic prothrombotic factors. In acute portal vein thrombosis, patients may be asymptomatic or present with life-threatening intestinal ischemia and infarction. In the chronic stage, patients generally present with complications related to portal hypertension, such as variceal bleeding and hypersplenism.[3] However, no cause is identified in more than 25%of patients.[4] The case report will highlight the abdominal swelling as a remarkable symptom in portal vein thrombosis. The main objective to report the case is unusual thrombotic feature of SLE.

1. *Dr. Syed Jamil Abdal, Assistant Professor, Department of Rheumatology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka. E-mail: sjabdal@gmail.com
2. Prof. Md. Abu Shahin, Professor, Department of Rheumatology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka.
3. Dr. Md. Ariful Islam, Associate Professor, Department of Rheumatology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka.

*For correspondence

CASE REPORT

A 24-year-old female, homemaker from southwestern district of Bangladesh got admitted in a remote hospital with one and half month previous history of gradually increasing swelling of the abdomen followed by swelling of the both legs and face without any history of jaundice or melena, breathlessness, fever. She was treated then conservatively with diuretics without any diagnosis or any resolution of her ascites. Six months later, again, she had started to have abdominal and pedal swelling with a gradually increasing puffy face; then she had got herself admitted in the same hospital for her diagnosis. This time extensive investigations were carried out and found raised ESR (45 mm in 1st hour), homogenous pattern of ANA positivity (28.5 u/mL) (reference value: >10 U/ml) with raised Anti-ds DNA Anti-ds DNA – 18.7 (reference value: <20 IU/ml), low C3 (43) (reference value: 80-170 mg/dL) and C4 (19) (reference value: 20-50 mg/dL). Her Anti-β2 Glycoprotein 1 (Ab IgM & IgG) were marginally positive but Lupus anticoagulant and Anti-Cardiolipin (Ab IgM & IgG) were negative. The hepatitis viral markers were negative. Her serum albumin (2, 2.4 and 1.47 g/dL in three occasions reference value is 3.5–5.0 g/dL), D-dimer (2.62 mcg/mL reference value is <0.5), INR (1.0), aPTT (30 seconds reference value is 30-40 seconds), UTP (0.14 g/day reference value is <0.150 gm/d) were carried out. The ascitic fluid study showed protein (3.0 g/dL reference value is 2.5 grams/dL) and ADA (5.18 IU/L reference value is 36 to 40 IU/L). The upper GI endoscopy revealed congestive gastropathy with duodenal ulcer, her x-ray chest PA view was normal.

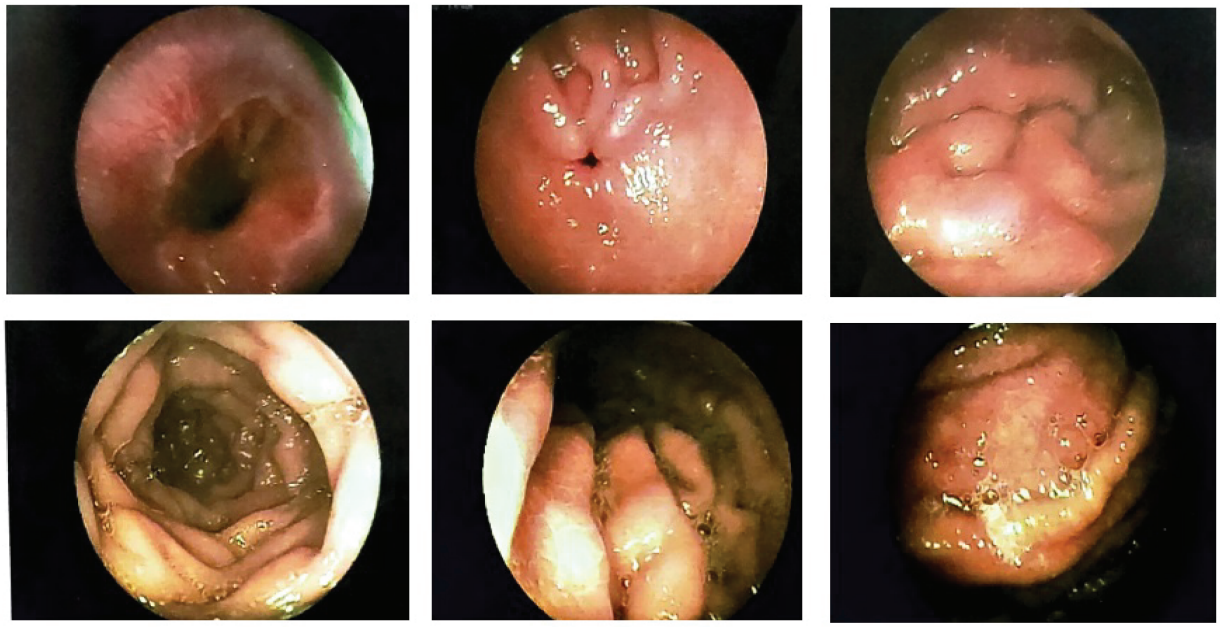

Figure 1 Upper GI endoscopy

At that time, ultra-sonogram of abdomen showed moderate ascites, gross thrombosis & stenosis in portal system (main trunk of portal vein, both proximal branches of portal vein, splenic vein at pancreatic area, superior mesenteric vein at confluence) which were 100% blocked. The upper GI endoscopy (Figure 1)

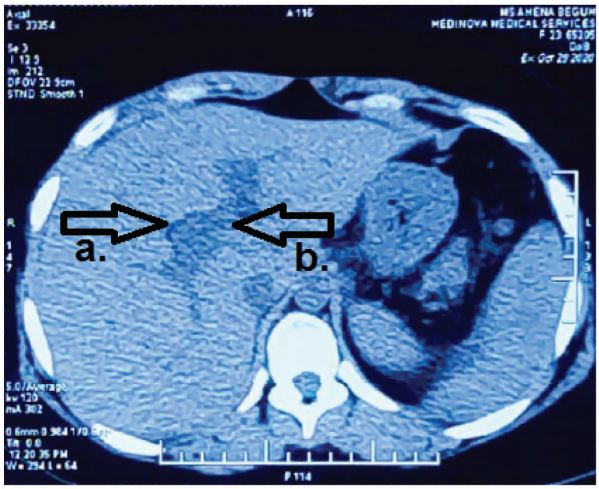

Figure 2 CT Abdomen

Upper GI endoscopy revealed congestive gastropathy as a normal esophagus without any varix where stomach revealed features of congestive gastropathy seen at the body and fundus and duodenum revealed multiple small superficial ulcer seen at the bulb, post-bulbar area and second part revealed portal hypertensive gastropathy. The CT scan of abdomen (**Figure 2**)

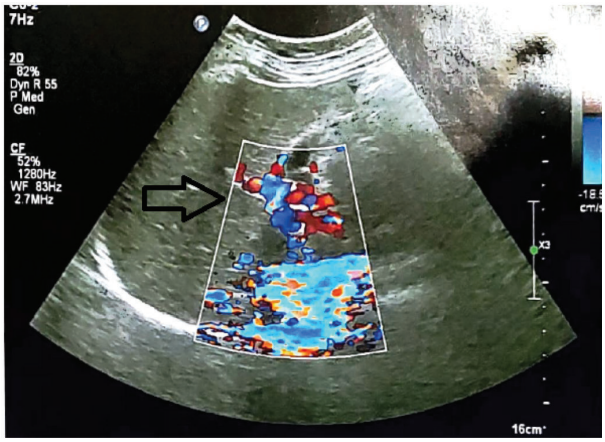

Figure 3 Doppler Ultrasound of portal and hepatic vein

Unenhanced axial CT scan of the abdomen showing an increase in the caliber of the portal vein (>13 mm) (a) that contains hyperdense material of portal vein thrombosis (b) revealed portal vein thrombosis with an enlarged portal vein. The Doppler ultra-sonogram of portal and hepatic vein revealed portal venous thrombosis with no flow with a peri-portal collateral circulation. (**Figure 3**)

The color Doppler ultrasound in a case of portal vein thrombosis, showing serpiginous vessels (arrow) in the periportal region.

Her Doppler ultra-sonogram of lower limbs were normal. She got symptomatic treatment without any improvement. Following primary management, she was referred to

tertiary hospital with new complaints of constant epigastric pain of recent onset with variable intensity that aggravates with food intake, associated with nausea and occasional vomiting. This time she also gave history of alopecia and photosensitivity. She was also suffering from constipation. She had received albumin infusion twice during her course of illness. She had neither any history of having pro-thrombotic risk factor nor any history of deep vein thrombosis. Her clinical examinations revealed she is overweight (BMI 27 kg/m2) with a puffy face. She was anemic, non-icteric, having bilateral pitting edema. Her abdominal examination revealed epigastric and bilateral hypochondriac tenderness without any organomegaly. Moderate ascites was present evidenced by shifting dullness. The bowel sound was audible. The examinations of all other systems including fundoscopy were unremarkable. During her hospital stay, she was initially treated with enoxaparin as 1 mg per kg/day, warfarin 5 mg/day, aspirin 75 mg/day, hydroxychloroquine 300 mg/day and medium dose prednisolone 0.5 mg/kg/day. After two weeks of the treatment her the patient symptomatically improved much. After readjustment of warfarin according to INR, the patient was discharged with a proper follow up schedule along with patient education about SLE and warfarin.

DISCUSSION

The PVT refers to the development of thrombosis within the extra-hepatic portal venous system draining into the liver.[5] It has been classified into 4 anatomic groups[6]:

(1) Thrombosis confined to the portal vein beyond the confluence of the splenic and superior mesenteric vein (SMV);

(2) Extension of thrombus into the SMV but with patent mesenteric vessels;

(3) Diffuse thrombosis of splanchnic venous system but with large collaterals;

(4) Extensive splanchnic venous thrombosis but with only fine collaterals.

The main mechanisms of PVT are due to disturbance of any element of the Virchow triad results in sluggish portal blood flow that occurs commonly in liver cirrhosis, hepatobiliary malignancies, gastric carcinoma, or extrinsic compression by lymph node or tumor.[7]

There are many medical conditions including Factor V Leiden deficiency, G20210A prothrombin gene mutation and surgical conditions can lead to thrombosis of the portal vein.[8, 9]

Table 1. Risk factors for portal vein thrombosis[10]

1. Acquired thrombophilia
   a. Antiphospholipid syndrome
   b. Paroxysmal nocturnal hemoglobinuria
2. Inherited thrombophilia
   a. Antithrombin deficiency
   b. Factor V Leiden
   c. Prothrombin gene G20210A mutation
   d. Protein C and S deficiency
3. Local factors
   a. Abdominal trauma
   b. Abdominal malignancy (eg, pancreatic cancer, hepatocellular carcinoma)
   c. Abdominal surgery (eg, splenectomy, liver transplantation)
   d. Endoscopic sclerotherapy
   e. Intra-abdominal inflammatory process (eg, cholecystitis, diverticulitis, pancreatitis)
   f. Transjugular intrahepatic portosystemic shunt
4. Systemic disorders
   a. Behçet syndrome
   b. Cirrhosis*
   c. Collagen vascular disease (eg, systemic lupus erythematosis)
   d. Inflammatory bowel disease
   e. Myeloproliferative syndrome (eg, polycythemia vera)

f. Pregnancy or exogenous hormone use

* Patients with decompensated cirrhosis are at higher risk for developing portal vein thrombosis than patients with compensated cirrhosis.

Our patient fulfilled the first classification. No definite time-frame found in the literatures to differentiate acute from chronic PVT, but studies of the chronic PVT considered in patients who developed symptoms <60 days prior to hospital assessment.[11]

Both acute and chronic PVT may be clinically asymptomatic and diagnosed incidentally during a radiologic examination for other reasons. The acute PVT patients may present with acute abdominal pain associated with or without fever. Our patient presented with epigastric pain that was not clinically specific for the portal

vein thrombosis. Sometimes, superior mesenteric vein thrombotic patients may have colicky abdominal pain and diarrhea. On the other hand, chronic PVT patients may present with symptoms related to complications of chronic PVT e.g. portal hypertension or portal cholangiopathy. The physical examination in most patients with acute PVT reveal no abnormalities though some patients with acute PVT may have an ileus and abdominal distension without other signs of intestinal obstruction.[12] Our patient presented with abdominal distention due to ascites. On the contrary, the patients with chronic PVT, even if asymptomatic, frequently have esophageal or gastric varices, and mostly of those present with gastrointestinal bleeding.[13-16]

Acute portal vein thrombosis (PVT) is diagnosed with abdominal imaging that demonstrates portal venous occlusion without the radiographic findings of chronic PVT. Radiographic findings in chronic PVT include demonstration of cavernous transformation of the portal vein and filling defects within the portal vein. A contrast-enhanced abdominal computed tomography (CT) scan can confirm the diagnosis, evaluate for predisposing conditions, assess the extent of the thrombosis, delineate the anatomical details, and detect evidence of intestinal infarction as well. Less suspicious patient can do a Doppler ultrasound. If the ultrasound is suggestive of acute PVT, an abdominal CT scan will be the next option for confirmatory diagnosis. Abdominal magnetic resonance imaging (MRI) is an alternative in patients who neither can undergo CT scan nor accept radiation exposure. Ultrasound with Doppler is an alternative; if CT and MRI are contraindicated or not available. Finally, a diagnosis of PVT is possible with portal venography; or by invasive procedure like superior mesenteric angiography that is generally not required.[17]

The primary management of acute portal vein thrombosis (PVT) is anticoagulation and when found; treatment of predisposing conditions. Anticoagulation is required to prevent extension of the clot and prevention of development of portal hypertension. The presence of predisposing conditions and the patient's comorbidities dictate the management of chronic portal vein thrombosis (PVT). Anticoagulant therapy should be initiated promptly in all patients, unless contraindicated because spontaneous restoration of patency is rare and recanalization of the portal vein prevents complication like portal hypertension that dictates the prognosis of the patient. Early anticoagulation results in better outcome e.g. chances of recanalization is 60% if therapy begins within the first week after symptom onset where only 20% if started within the first month. Anticoagulation needs to be continued indefinitely in patients with an underlying thrombophilic disorder. Therefore, our patient needs to be on life-long anticoagulation. Surgical thrombectomy is not recommended.[18] Predictors of survival have not been properly studied, the main determining factor appears to be advanced age and mesenteric venous thrombosis.[19]

For reduction of PVT-associated morbidity and mortality there are two broad objectives to be achieved, firstly, to reverse or prevent the advancement of thrombosis within the portal venous system; and secondly, to treat the complications of established PVT, most specifically gastrointestinal varices or biliary complications. (G. J. M. Webster et al.)

CONCLUSION

The PVT is a rare disease. A high index of suspicion needs to reach a clinical diagnosis of PVT considering the multiple risk factors, including inherited and acquired thrombophilic predispositions. Early diagnosis will certainly prevent both morbidity and mortality.

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I acknowledge the contribution of our former Nepalese resident Dr. Bidhan Neupane, now practices as rheumatologist in Nepal after passing MD course in rheumatology.

CONFLICT OF INTEREST STATEMENT

The authors have no conflict of interest.

REFERENCES

1. Boey ML, Colaco CB, Gharavi AE, Elkon KB, Loizou S, Hugues GRV: Thrombosis in systemic lupus erythematosus: striking association with the presence of lupus anticoagulant. Br Med J 1983; 287: 1021-1023.
2. Pauzner R. Rosner E, Many A: Circulating anticoagulant in systemic lupus erythematosus: clinical manifestations. Acta Haematol (Basel) 1986; 76: 90-94.
3. Parikh S, Shah R, Kapoor P: Portal Vein Thrombosis. The American Journal of Medicine, Vol 123, No 2, February 2010.

4. Valla D, Casadevall N, Huisse MG et al., "Etiology of portal vein thrombosis in adults. A prospective evaluation of primary myeloproliferative disorders," Gastroenterology, vol. 94, no. 4, pp. 1063–1069, 1988.
5. Webster GJM, Burroughs AK, Riordan SM, "Portal vein thrombosis—new insights into aetiology and management," Alimentary Pharmacology and Therapeutics, vol. 21, no.1, pp. 1–9, 2005.
6. N. V. Jamieson, "Changing perspectives in portal vein thrombosis and liver transplantation," Transplantation, vol. 69, no. 9, pp. 1772–1774, 2000.
7. DeLeve LD, Valla DC, Garcia-Tsao G (2009) Vascular disorders of the liver. Hepatology 49: 1729–1764.
8. Sobhonslidsuk A, and Reddy KR, M.D. Portal Vein Thrombosis: A Concise Review, Center For Liver Diseases, University of Miami School of Medicine, Miami, Florida AJG Vol. 97, No. 3, 2002
9. Case Reports in Vascular Medicine Volume 2015, Article ID 823063, 5 pages http://dx.doi.org/10.1155/2015/823063
10. UpToDate Version 28.0
11. Malkowski P, Pawlak J, Michalowicz B, et al. Thrombolytic treatment of portal thrombosis. Hepatogastroenterology 2003; 50: 2098–100.)
12. Primignani M. Portal vein thrombosis, revisited. Dig Liver Dis 2010; 42:163.
13. Sogaard KK, Astrup LB, Vilstrup H, Gronbaek H. Portal vein thrombosis; risk factors, clinical presentation and treatment. BMC Gastroenterol 2007; 7:34.
14. Amitrano L, Guardascione MA, Brancaccio V, et al. Risk factors and clinical presentation of portal vein thrombosis in patients with liver cirrhosis. J Hepatol 2004; 40:736.
15. Webb LJ, Sherlock S. The aetiology, presentation and natural history of extra-hepatic portal venous obstruction. Q J Med 1979; 48:627.
16. Kameda H, Yamazaki K, Imai F, et al. Obliterative portal venopathy: A comparative study of 184 cases of extrahepatic portal obstruction and 469 cases of idiopathic portal hypertension. J Gastroenterol Hepatol 1986; 1:139.
17. Subramanyam BR, Balthazar EJ, Lefleur RS, et al. Portal venous thrombosis: correlative analysis of sonography, CT and angiography. Am J Gastroenterol 1984; 79:773.
18. Llopa E, Seijo S. Gastroenterol Hepatol. 2016; 39 (6):403-410.
19. Chawla YK, Bodh V. Portal vein thrombosis. J Clin Exp Hepatol.2015;5:22-40.