International Journal of Research in Medical Sciences
Polavarapu N et al. Int J Res Med Sci. 2019 Sep;7(9):3550-3552
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20193653

Case Report

Recurrent abdominal and lower limb venous thrombosis due to inherent protein-C and protein-S deficiency

Naren Polavarapu, Senthil Narayanasamy*, Naveen Arunachalam Subramanian

Department of General Medicine, Sri Ramachandra Institute of Higher Education and Research, Porur, Chennai, Tamil Nadu, India

Received: 28 July 2019
Accepted: 05 August 2019

***Correspondence:**
Dr. Senthil Narayanasamy,
E-mail: narenpvr12@gmail.com

ABSTRACT

Protein-C and protein-S deficiency is associated with a hypercoagulable state which usually presents with recurrent venous thrombosis as a common complication. But extensive thrombosis involving all the major abdominal and lower limb veins is quite rare. Here, we report a case of a 27-year-old woman presented with engorged veins all over abdomen and chest since 20 days. Clinical examination revealed signs suggestive of portal venous hypertension with moderate splenomegaly. Protein-C and protein-S levels were found to be low. Portal venous doppler ultrasound and CT-venogram revealed chronic thrombosis of portal vein, inferior vena cava, bilateral iliac and femoral veins with extensive collaterals formation and partial thrombosis in collaterals as well. The patient was successfully managed with anti-coagulating agent (acenocoumarol) and has been maintained on regular follow-up to avoid reoccurrence of thrombosis.

Keywords: Protein-C, protein-S, Venous thrombosis, Portal vein thrombosis

INTRODUCTION

Hypercoagulable state is a condition in which patient is prone to thrombus formation due to inherent or acquired abnormalities in coagulation and anti-coagulation pathway. Protein-C, protein-S and antithrombin-III are natural anti-coagulation proteins and deficiency of these proteins result into recurrent venous thrombosis development.[1-3] Thrombosis can occur in any vein of the body such as cortical veins, portal vein, deep veins of lower limb, pulmonary veins, cerebral veins, etc. and patients may present with wide array of clinical symptoms based on the site of involvement. Here, we are reporting a case of protein-C and protein-S deficient patient with chronic thrombosis of all major abdominal and lower limb veins including portal vein, inferior vena cava, and bilateral iliac and femoral veins.

CASE REPORT

A 27-year-old women was presented with complains of engorged veins over whole abdomen and chest wall since 20 days. She had a history of deep vein thrombosis which was treated by anti-coagulants but was stopped by her own without physician's assistance. She was not a vegetarian by diet and her siblings had no similar complaints. She had no known co-morbidities or had no history of hematemesis or melena. On examination she was afebrile, with 90/50 mmHg blood pressure at the time of presentation and her other vital were normal. Left lower limb pitting pedal edema was observed and thus

bilateral lower limb compression stockings were used by the patient. Detailed abdominal examination revealed moderate splenomegaly and engorged dilated and tortuous veins all over abdomen, chest and back (Figure 1).She had no evidence of systemic involvement.

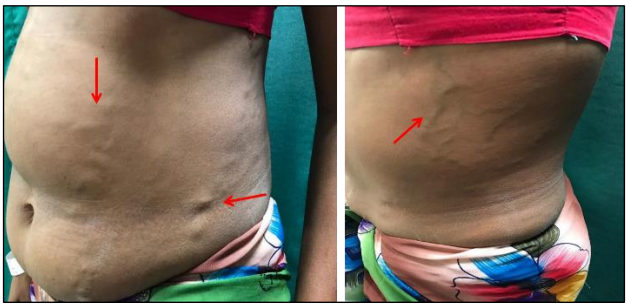

Figure 1: Engorged prominent veins all over abdomen, chest and back.

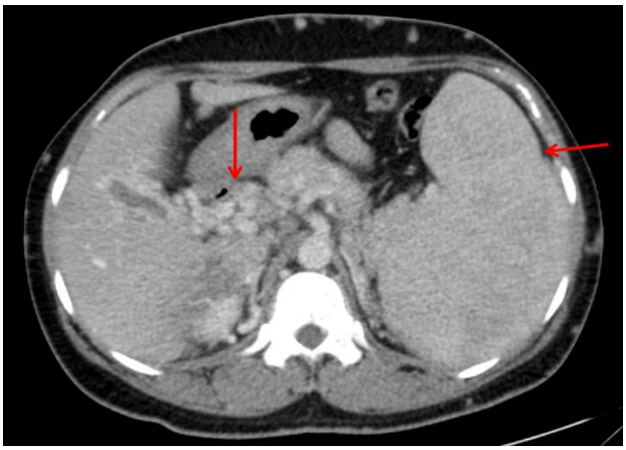

Figure 2: Computed tomography-venogram showing portal cavernoma at porta hepatis and splenomegaly.

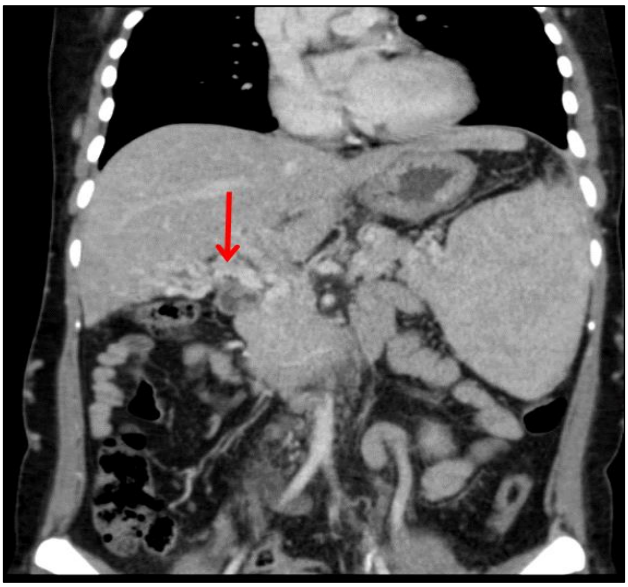

Figure 3: Thinned-out main portal vein with thrombus within.

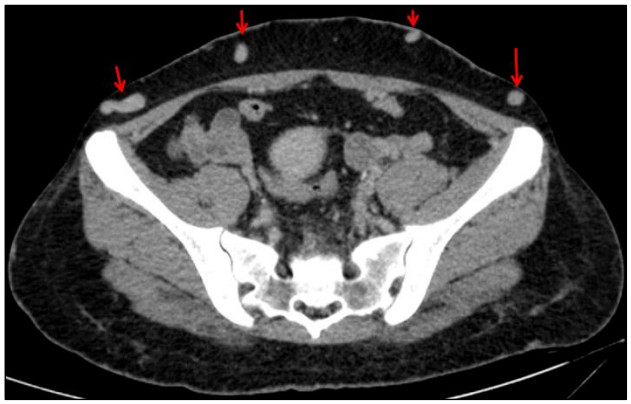

Figure 4: Multiple dilated veins (collaterals) in anterior abdominal wall.

Blood investigations revealed decreased haemoglobin level (7.5 gm/dL; N: 12.1-15.1 g/dL) and platelet counts (71 × 109 cells/L; N: 150-400 x 109/L). Her renal and electrolyte examination showed blood urea nitrogen 15 mg/dL (N: 7-20 mg/dL), serum creatinine 0.8 mg/dL (N: 0.5 to 1.1 mg/dL), sodium 134 mmol/L (N: 135-145 mmol/L), potassium 4.2 mmol/L (N: 3.5-5.0 mmol/L), chloride 102 mmol/L (98-106 mmol/L) and bicarbonate 24 mmol/L (23-30 mmol/L). Her prothrombin time (PT) (17.3 seconds; N: 11-13.5 seconds) and international normalized ratio (INR) (1.51; N: 0.8-1.1) both were higher. Activated partial thromboplastin time (aPTT) was 24.6 seconds (N: 30-40 seconds). Coagulating protein-C activity was 61.9% (N: 70- 140%) and protein-S activity was 44.7% (N: 65-140%). Portal venous doppler ultrasound and computed tomography-venogram revealed chronic thrombosis of portal vein, inferior venacava, bilateral iliac and femoral veins with extensive collaterals formation and partial thrombosis in collaterals as well.

The patient was started on tablet acenocoumarol-3 mg at bedtime to control the condition. Serial PT/INR monitoring was done and the dose of acenocoumarol was gradually increased to 4 mg and then to 5 mg at bedtime. Patient was advised to continue the use of bilateral lower limb compression stockings. She was discharged with tablet acenocoumarol-5 mg once daily at bedtime. The prominent veins over the abdomen and chest have decreased and she seems to be doing fairly well with her daily activities without worsening of symptoms. Patient has been maintained on regular follow-up.

DISCUSSION

Protein-C and protein-S both are vitamin-K dependent glycoproteins that co-ordinately work as natural anti-coagulants which degrade coagulating factor Va and VIIIa to prevent thrombin formation. The deficiency of protein-C and/or protein-S, either congenital or acquired, results into episodes of venous thrombosis that generally begin in the late teens and twenties. In patients with venous thromboembolism, the prevalence of deficiency of protein-C and protein-S was 2-5%.[4-7] Here, we report a

case of a women who is in her twenties, already had a history of deep vein thrombosis, and yet again diagnosed with chronic abdominal venous thrombosis including portal vein, bilateral iliofemoral vein and inferior venacava.

Various causes and predisposing factors of venous thrombosis have been reported in literature.[3,8] The diagnosis of inherent protein-C and protein-S deficiency is very crucial and most of the patients present with other predisposing factors too. In the present case, the clinical, radiological, and laboratory evaluations and all the unusual sites of thrombosis guided the physician towards the diagnosis of protein-C and protein-S deficiency. Although spontaneous resolution of most vein thrombosis have been reported, specific conservative management is mandatory to prevent recurrent thrombosis, to prevent extension of thrombosis and to re-establish the patency of all the affected veins.

Currently, anti-coagulant therapy has been used to recanalize thrombotic veins and have displayed good outcomes with least complications. However, the patients with chronic venous thrombosis are often at higher risk of recurrent thrombosis. Thus, the decision to start and to continue anti-coagulating agents must be made on a case-by-case basis. In the current case, acenocoumarol was used as an anti-coagulating agent. In patient with protein-C and protein-S deficiency, it is better to use acenocoumarol rather than warfarin because of its shorter duration of action, less variability in anti-coagulation and require less monitoring. In addition, it is important to prevent over correction of the condition as it can cause bleeding manifestations such as most dangerous intracranial bleeding (usually when INR goes above 4). The patient with such inherent deficiency should be advised to take anti-coagulating agents indefinitely and sharp monitoring of PT/INR is mandatory to avoid any future bleeding complications.

CONCLUSION

The reported case is very rare with recurrent abdominal and extensive lower limb venous thrombosis in a young woman with protein-C and protein-S deficiency. Whenever any patient presents with recurrent thrombosis especially in younger individuals, it is important to rule out other conditions that can cause hypercoagulable state such as liver diseases, malignancy or disseminated intravascular coagulation before attributing it to protein-C/protein-S deficiency. Regular follow-up with PT/INR monitoring (once in a month) is recommended.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not Required

REFERENCES

1. Nachman RL, Silverstein R. Hypercoagulable states. Annals of Internal Medicine. 1993;119:819-27.
2. Qi X, De Stefano V, Wang J, Bai M, Yang Z, Han G, et al. Prevalence of inherited antithrombin, protein C, and protein S deficiencies in portal vein system thrombosis and B udd-C hiari syndrome: A systematic review and meta-analysis of observational studies. Journal of gastroenterology and hepatology 2013;28(3):432-42.
3. Hotoleanu C. Genetic risk factors in venous thromboembolism. Thrombosis and Embolism: from Research to Clinical Practice: Springer; 2017:906;253-72.
4. Cho Y, Kwon T, Ahn J, Kang G, Han M, Kim Y, et al. Protein C and/or S deficiency presenting as peripheral arterial insufficiency. The British J radiol. 2005;78:601-5.
5. Doganay S, Sonmezgoz F, Sen Y, Kocakoc E. Deep vein thrombosis associated with protein C and protein S deficiency: An unusual cause of acute abdomen. J Clinical Ultrasound. 2011;39(5):287-9.
6. Mondal R, Nandi M, Dhibar T. Protein C and protein s deficiency presenting as deep venous thrombosis. Indian pediatr. 2010;47(2):188-9.
7. Deshpande S, Patil S, Pendse M, Kashyap P, Dave D. Portal vein thrombosis with protein CS deficiency in a non-cirrhotic patient. International Journal of Research in Medical Sciences 2016;4(11):5061.
8. Rodríguez-Leal GA, Morán S, Corona-Cedillo R, Brom-Valladares R. Portal vein thrombosis with protein CS deficiency in a non-cirrhotic patient. World J hepatol. 2014;6(7):532.

Cite this article as: Polavarapu N, Narayanasamy S, Naveen AS. Recurrent abdominal and lower limb venous thrombosis due to inherent protein-C and protein-S deficiency. Int J Res Med Sci 2019;7:3550-2.