Case Report

A Rare Case of Splenic Vein Thrombosis

Anitta Varghese[1]*., Ancy George[2]., Juvita Joy[3]., Abila T Babu[4]., Lakshmi R[5], Arthanareeswaran S[6]

[1-5]Department of Pharmacy Practice, Swamy Vivekanandha College of Pharmacy, Elayampalayam, Tiruchengode, Tamil Nadu, India.

[6]Department of General Medicine, Vivekanandha Medical Care Hospital, Elayampalayam, Tiruchengode, Tamil Nadu, India.

***Corresponding author's E-mail:** anittav5@gmail.com

Received: 12-10-2019; **Revised:** 25-11-2019; **Accepted:** 05-12-2019.

ABSTRACT

Splenic vein thrombosis (SVT) is a blood clot that is obstructing the splenic vein, which is located on the surface of the spleen. SVT in non-cirrhotic and non-malignant individuals can occur in the setting of coagulation disorders, abdominal inflammatory processes such as pancreatitis, infections, and inflammatory bowel diseases, and after abdominal surgeries. Individuals with protein S deficiency are at risk for developing blood clots, specifically blood clots that begin in veins (venous thromboembolisms). Duplex, Doppler ultrasound, and abdominal CT scan can be used to make an accurate diagnosis of SVT in the early stage, and abdominal MRI can be another diagnostic tool. There is no therapy particular to the patients with protein S deficiency. The use of anticoagulant therapy however is highly effective in the treatment and prevention of blood clots in patients with the common type of protein S deficiency.

Keywords: Splenic vein, Thrombosis, Protein S, Anticoagulant.

INTRODUCTION

Splenic vein thrombosis (SVT) is a blood clot that is obstructing the splenic vein, which is located on the surface of the spleen. There are chances of internal bleeding as a result of the raised pressure in the splenic vein which in turn leads to the enlargement of spleen and other veins to dilate and twist in the esophagus and stomach.[1] It is relatively rare and has been associated with the presence of an underlying liver disease or prothrombotic disorders.[2]

The meta-analysis on thrombophilia in portal venous system thrombosis showed the prevalence of protein S deficiency in 2.6% with Odds ratio of 8.00.[3] Indian data on hypercoagulable states in splenic vein thrombosis reveal that the inherited thrombophilia is infrequent in the Indian population and protein S deficiency was seen in 3–4%.[4]

CASE REPORT

A 46 year old female was admitted to the casualty department with complaints of pain in left hypochondrium for past 3 days. She had a significant past medical history of type 2 diabetes mellitus and hypertension. She underwent hysterectomy 7 years back. She was slightly overweight with a BMI of 27.4 kg/m^2 and had no history of similar illness in the past and in her family.

Vitals revealed a BP of 160/100 mm Hg, Pulse rate: 82 beats/ min, SpO_2 : 95%. Her random blood glucose was 179 mg/dl, blood urea was 15.10 mg/dl, Serum amylase 133 U/L and lipase: 89 U/L. Her Hb level was 11.3 g/dl, PCV 35.10%, prothrombin time 18.6 seconds. CT abdomen-contrast revealed focal partial thrombosis of splenic vein. Her procoagulant state was investigated and it was found that she had declined protein S activity of 42%.

Patient was treated with low molecular weight heparin followed by oral anticoagulants. Her conditions improved on the following days.

DISCUSSION

SVT in non-cirrhotic and non-malignant individuals can occur in the setting of coagulation disorders, abdominal inflammatory processes such as pancreatitis, infections, and inflammatory bowel diseases, and after abdominal surgeries.[5] It seems that a combination of general and local factors is needed to enable the development of SVT, thus establishing the importance of a thorough investigation of those factors when facing a diagnosis of SVT.[6] These factors includes either inherited or acquired prothrombotic disorders, other thrombophilic factors, and local factors.[7]

This patient was diagnosed with PVT due to protein S deficiency. Thrombus formation is governed by the principles of the Virchow's triad namely, hemostasis, endothelial injury and hypercoagulability.[8]Extrinsic obstructions have been ruled out in our patient and inherited hypercoagulable states due to protein S deficiency is identified as the cause of SVT.

Individuals with protein S deficiency are at risk for developing blood clots, specifically blood clots that begin in veins (venous thromboembolisms). The exact risk of a blood clot, the age of onset of the disease, the severity of the disease, and the number and frequency and location of blood clots will vary greatly among affected individuals.

Some indications of a possible protein S deficiency include blood clots that develop before the age of 50 without an obvious cause, recurrent blood clots, blood clots in a person in a family with history of blood clot formation, and

blood clots that occur in sites that are not normally affected by blood clots including veins in the small bowel (mesenteric veins), veins of the liver (portal veins), and veins in the brain (cerebral veins).[9]

SVT may be symptomatic or asymptomatic and the GI bleeding at varying severity is the most common manifestation of this syndrome.[10] Duplex, Doppler ultrasound, and abdominal CT scan can be used to make an accurate diagnosis of SVT in the early stage, and abdominal MRI can be another diagnostic tool. [11]

Current guidelines recommend anticoagulation treatment for all patients with SVT to be administered for atleast 3 months and to be extended beyond this period in patients with permanent prothrombotic conditions and an acceptably low risk of bleeding. [12,13]In this case of acute SVT, however, early anticoagulant therapy did prevent the spread of thrombi and pro-moted blood reperfusion.

There is no therapy particular to the patients with protein S deficiency. The use of anticoagulant therapy however is highly effective in the treatment and prevention of blood clots in patients with the common type of protein S deficiency. When anticoagulation is pursued, warfarin is frequently the anticoagulant of choice, although LMWH can be safely considered as an alternative. The choice of drug, specific dosage, and duration of anticoagulant therapy will vary among affected individuals.[9]

Table 1: Suggested anticoagulant therapy for the acute-phase treatment

Symptom	Treatment
Noncirrhotic, symptomatic SVT patients with no signs of active bleeding	Consider full therapeutic dose LMWH (eg, 1 mg/kg twice daily). Start VKA after 48-72 h if no bleeding occurs.
Cancer-associated SVT	Full therapeutic dose LMWH (eg, 1 mg/kg twice daily) for 1 mo and then reduce to 75% of the initial dose for at least 3–6 mo.
Estimated glomerular filtration rate <30 mL/min	Consider UFH or reduce the dose of LMWH by 50% and consider anti-Factor Xa level monitoring. If creatinine clearance is lower than 15 mL/min, abstain from using LMWH. VKA should be started as soon as possible to reduce the duration of heparin treatment (consider VKA also for cancer patients with chronic severe renal insufficiency).
Platelet count >30 000 and <50 000 per mm^3	Reduce the dose of LMWH by at least 50%, (more if additional risk factors for bleeding are present) and delay VKA initiation (if VKA is indicated) until the cause of thrombocytopenia is diagnosed and managed. Consider abstaining from anticoagulant therapy if a single vessel clot with limited extension and reassess when platelet count improves.
Platelet count <30 000 per mm^3	Abstain from using anticoagulant drugs or consider prophylactic dose LMWH if acute thrombosis in multiple vessels and platelet count more than 20 000 per mm^3.
Cirrhotic, symptomatic PVT patient	Consider full therapeutic dose LMWH (eg, 1 mg/kg twice daily) after careful assessment (and treatment, if necessary) of esophageal varices. Empiric dose reductions (50% of therapeutic dose or more, based on individual risk assessment) if additional risk factors for bleeding are identified. Delay starting of LMWH if major risk factors for bleeding coexist, until successfully managed. Delay VKA initiation and start only when the patient is stable and no additional major bleeding risk factors are identified, also according to patient preference.
Concomitant renal failure	Same as for the noncirrhotic patient, but consider abstaining from any anticoagulant treatment if poor short-term prognosis or if concomitant major risk factors for bleeding are identified. VKA is to be considered if no severe concomitant coagulopathy exists.
Platelet count <50 000 per mm^3	Abstain from anticoagulant treatment or reduce the dose of LMWH by 50% (or more, based on individual risk assessment) if extended and occlusive thrombosis.
Incidentally detected SVT	Apply the same treatment regimens proposed for the symptomatic patient unless 1 or more of the following conditions are identified: thrombosis is nonocclusive, likely not recent, and limited to a single vein segment; no permanent risk factors or no recent (<1 mo) removable risk factors for thrombosis are identified; bleeding risk is moderate to high; and prognosis of underlying disease is poor.

Table 2: Suggested strategies for the long-term treatment

Patient	Strategy
All patients with SVT	At least 3 mo of anticoagulant treatment
Patients with SVT secondary to surgery	Treatment discontinuation after 3-6 mo
Patients with PVT, MVT secondary to nonsurgical transient risk factors (eg, hormonal therapy, abdominal infections)	Treatment discontinuation after 6-12 mo
Patients with PVT, MVT secondary to permanent risk factors (eg, cirrhosis, chronic inflammatory disorders, cancer, MPNs, autoimmune disorders, major thrombophilia*)	Indefinite anticoagulant treatment with periodic reassessment of bleeding profile
Patients with unprovoked SVT	Indefinite anticoagulant treatment with periodic reassessment of bleeding profile
Patients with BCS	Indefinite anticoagulant treatment with periodic reassessment of bleeding profile

*Antiphospholipid antibodies, double heterozygosity for Factor V Leiden mutation and G20210A mutation in prothrombin gene or single homozygosity.

CONCLUSION

Splenic vein thrombosis is a rare disease, but our understanding of this disorder has improved during the last few years. It continues to be a complicated and difficult condition with significant morbidity and mortality. In patients with SVT, local factors such as liver cirrhosis and pancreatitis should be suspected. It is necessary to investigate genetic or acquired thrombophilic factors if no local factor can be identified. This case appears to be a SVT provoked by protein S deficiency. Though the patient had multiple predisposing factors as past history of hysterectomy, fibroid uterus, fatty liver disease and pancreatitis, these were least likely to have caused it. The presence of SVT should be regarded as an indicator for prothrombotic disorders, liver disease, and other local and general factors that must be carefully investigated.

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Source of Support: Nil, **Conflict of Interest:** None.