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CT-151 A Rare Bleeding Diathesis With Subacute Splenic Hematoma: PAI-1 Deficiency

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INTRODUCTION

Plasminogen activator inhibitor 1 (PAI-1) deficiency is a rare hyperfibrinolytic bleeding disorder characterized by mild to moderate bleeding symptoms. It can present similarly to other bleeding disorders such as platelet dysfunction, coagulation factor deficiencies, or von Willebrand factor deficiency. Early diagnosis of PAI-1 deficiency is crucial, as it can be easily treated with antifibrinolytic agents, improving patient care and the psychological well-being of both the physician and the patient. .

AIM

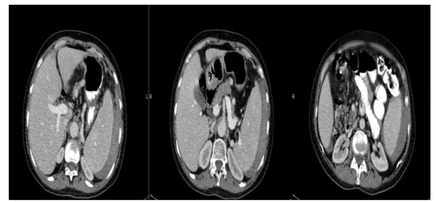
In our case report, we present a case of PAI-1 deficiency who developed spontaneous visceral organ hematoma as a major bleeding attack.

CASE REPORT

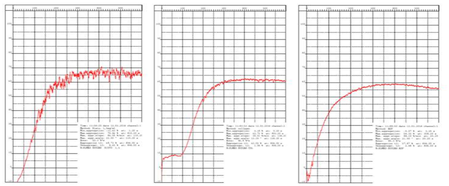
A 55-year-old female patient was admitted to the hospital in March 2018 with abdominal pain radiating to the shoulder. She had a total abdominal hysterectomy and bilateral salpingoophorectomy operation due to menometrorrhagia in her history. There were no additional features in her family history. Abdominal ultrasonography revealed hepatosplenomegaly (spleen and liver, both 160 mm) and a hypodense area compatible with a subacute hematoma of approximately 130 x 200 mm in the subcapsular localization of the spleen and fluid in the douglas and perihepatic region (Picture 1.).

RESULTS

The patient received 2 units of erythrocyte suspensions and a unit of fresh frozen plasma (FFP). She was referred and consulted to our hematology department. Peripheral smear showed no morphological abnormalities and confirmed a normal count regarding platelets. A coagulation panel was studied in terms of bleeding diathesis due to the history of menometrorrhagia in her medical history. Prothrombin time (PT), activated partial thromboplastin time (PTT), vWF antigen, ristocetin cofactor activity, factor VIII level, factor XIII level, platelet function tests, platelet aggregometry tests and thromboelastography (Picture 2.) were performed Since Glanzman Thrombasthenia and Bernard Soulier Syndrome were included in the differential diagnosis, CD41, CD61 and CD42 levels were measured by flow cytometry. Routine coagulation studies were normal including the PT and PTT. There was no evidence of ongoing disseminated intravascular coagulation with a normal fibrinogen level (386 mg/dl, normal 200-400). The bleeding time and vWF antigen, ristocetin cofactor, and factor VIII coagulant activity were normal, suggesting that a diagnosis of von Willebrand's disease was unlikely. As a lack of a potential explanation for her ongoing spontaneous subcapsular hepatic and splenic bleeding, the PAI-1 Activity of the patient was ordered and was found to be <0.05, and she was diagnosed with PAI-1 deficiency. The patient was started on tranexamic acid (TXA) 500 mg tablet 4x1 per day. Subcapsular hematoma was followed up with daily ultrasonography. 20 days later, her abdominal findings had completely disappeared in imaging.
One year later, she had a major implant surgery under the treatment of TXA without complications and without a need for transfusion.



Picture 1. Subcapsular splenic hematoma, computed tomography (CT), transverse section



Picture 2.
Thromboelastography curves: Non-pathological thromboelastogram result of patient

CONCLUSIONS

In conclusion, PAI-1 deficiency is a rare bleeding disorder and may present with mild to severe bleeding clinic. In our opinion, a hyperfibrinolytic bleeding disorder should be suspected in patients with BUC with recurrent, abnormal posttraumatic, postsurgical, and/or mucocutaneous bleeding; a positive family history; reproductive failure; and/or bleeding in unusual sites. Treatment with TXA is safe and sufficient and avoids unnecessary transfusion and surgical procedures. PAI-1 deficiency should be kept in mind in bleeding disorders of unknown origin. Our patient, who presented with spontaneous major organ bleeding because of PAI-1 deficiency, constitutes an important contribution to the literature..

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