Factor VII Deficiency: A Paradoxical Bleeding Disorder with Prothrombotic State

Factor VII deficiency is a rare bleeding disorder which can vary in both its severity and signs and symptoms on presentation. Common presentations include excessive bruising, hemarthrosis, or hematuria. Spontaneous thrombosis occurs in 3-4% of the factor VII deficiency patients. This case is an example of a 47 year old male who presented with coronary artery thrombosis and progressive ischemic cardiomyopathy and was later found to have factor VII deficiency.

A 47 year old male with a past medical history of coronary artery disease status post a coronary artery stent placement after a myocardial infarction two years prior, hypertension, non-insulin dependent diabetes, and transient ischemic attack presented to the hospital with progressively worsening shortness of breath. Echocardiogram showed an ejection fraction of 25% which was significantly reduced for a prior echo obtained one year prior raising concern for obstructive coronary artery disease. On admission, it was noted that he was found to have an isolated elevated PT and INR of 1.7 without the use of anticoagulation at home. The patient's INR increased to as high as 2.1. With an elevated INR, cardiac catheterization could not immediately be performed. Despite multiple trials of Vitamin K, his INR did not change. Further testing with coagulation mixing studies and factor activity testing revealed a factor VII activity level of 13% compatible with factor VII deficiency. In addition, no factor VII inhibitor was detected. The patient subsequently underwent cardiac catheterization after receiving recombinant factor VII and was found to have multi vessel disease as well as stent restenosis. No post procedural hemorrhagic events were noted.

Factor VII is a vitamin K dependent factor produced by the liver. When it binds to tissue factor, its activation begins the intrinsic coagulation cascade leading to thrombin production. Deficiency may result in excessive bleeding since thrombin production is impaired. Thrombosis associated with Factor VII is uncommon and the mechanism is not well understood. This case is a rare presentation of a young male who presented with recurrent thrombosis despite appearing hypocoaguable by in vitro coagulability testing. The presence of a bleeding disorder does not necessarily rule out prothrombotic states and must still be considered. The management of such patients who are prothrombic yet appear to be hypocoaguable is complicated as a balance needs to be maintained between risk of bleeding versus thrombus formation.