TM, arterial thrombosis was observed in 2 (3.3%) patients, venous thrombosis in 3 (5.0%) patients, and signs of chronic venous insufficiency in 4 (6.7%) patients. Out of 70 non-splenectomized patients with TM, venous thrombosis was observed in 1 (1.4%) patient, and signs of chronic venous insufficiency in 3 (4.3%) patients. Of the 40 splenectomized TI patients, arterial thrombosis was observed in 2 (5.0%), venous thrombosis in 3 (7.5%), and signs of chronic venous insufficiency in 4 (10%). Of 55 non-splenectomized TI patients, venous thrombosis was observed in 2 (3.6%), and signs of chronic venous insufficiency in 3 (5.4%). Assessment of thrombotic tendency was conducted among non-splenectomized patients. HS (the total score for the PTT questionnaire >30) was detected in 36.0 ± 6.8% of TM patients and 40.0 ± 7.7% of TI patients. In patients with TI and in BD, increased thrombotic tendency was not detected (the sum of the scores for the PTT questionnaire is <30).

Conclusion: TCs detected in patients with homozygous β-thalassemia was more common in patients with TI compared with patients with TM ($p \geq 0.05$). In patients, cases of venous thrombosis were detected 2 times more often than arterial thrombosis ($p \geq 0.05$). Chronic venous insufficiency was detected identically in patients with TM and TI. TCs was observed more often in splenectomized patients with TM and TI compared with non-splenectomized patients ($p \geq 0.05$). It was established that some patients with β-thalassemia who did not have clinical thrombotic complications had prethrombotic state. A study of clinical and anamnestic risk factors revealed a tendency to HS in 1/3 of patients with β-thalassemia. Based on the results of the survey, the risk factors (predictors) of HS were determined. The tendency to form blood clots in patients with anemia was associated with two groups of clinical and anamnestic symptoms: “comorbidity” and “chronic stress conditions”.

https://doi.org/10.1016/j.htct.2020.09.080

PP 19

Factor XIII deficiency case with posttravmatic subcutaneous bleeding

M. Reis Aras[1,*], M. Tıglıoğou[1], B. Sağlam[1], P. Akyol[1], Ü. Malkan[1], A. Alan[2], M. Albayrak[1]

[1] *University of Health Sciences, Ankara Dışkapı Yıldırım Beyazıt Training and Research Hospital, Hematology Department, Ankara, Turkey*
[2] *University of Health Sciences, Ankara Dışkapı Yıldırım Beyazıt Training and Research Hospital, Internal Medicine Department, Ankara, Turkey*

Objective: Factor XIII deficiency; is a rare hereditary bleeding disorder caused by heterogeneous mutations that can lead to life-threatening bleeding. Hereditary factor XIII deficiency's inheritance is autosomal recessive and its incidence is about 1–3/1,000,000. The form of bleeding can be seen in a wide spectrum, from life-threatening bleeding (such as intracranial bleeding) to skin bleeding. Umbilical cord hemorrhage and soft tissue hematoma is the most common and often first symptom of factor XIII deficiency (1). Lifelong bleeding diathesis can be seen in hereditary FXIII deficiency. Especially subcutaneous bleeding (57%), delayed umbilical cord bleeding (56%), muscle hematoma (49%), postoperative bleeding (40%), intracerebral bleeding (34%) and recurrent abortion can be seen. Bleeding after trauma or surgery (12–36 h) is pathognomonic in factor XIII deficiency. (2) Diagnosis of factor XIII deficiency is difficult due to its rarity. Because standard clotting screening tests including prothrombin time (PT), activated partial thromboplastin time (aPTT), thrombin time (TT), platelet count or bleeding time are normal; therefore, specific factor XIII assays are required. For all these reasons, factor XIII deficiency remains one of the least diagnosed rare bleeding disorders (1).

Case report: 34-year-old male patient applied to the emergency department due to the swelling that developed after hitting his right arm on the door. He stated that he had a history of factor 13 deficiency. Fracture or fissure line was not observed in the patient's physical examination and direct radiography. Bleeding observed in skin and subcutaneous region. In the anamnesis, the patient stated that he had a history of skin-subcutaneous bleeding and hematoma after trauma. In hospital records, it was observed that he had posttraumatic intramuscular hematoma two times in the last 5 years (the largest is 75 mm × 25 mm × 40 mm). In these hematomas treatment; there was no need for factor XIII concentrate, it was regressed with fresh frosen plasma replacement. In the laboratory tests performed in emergency department; leukocyte value 12,370/μL, neutrophil 6720/μL, hemoglobin 16.7 g/dL, platelet 315,000, PT: 9.12 s, aPTT 23.2 s, INR 1.02 was detected. Fresh frozen plasma was replaced at a dose of 15 mL/kg. The patient, who did not have any additional systemic problem, was discharged by recommending polyclinic control.

Conclusion: Hereditary factor XIII deficiency is an autosomal recessive bleeding disorder with a serious course (4). Unlike other hereditary hemostatic protein deficiencies, clotting tests and platelet function tests are normal in factor XIII deficiency. For this reason, specific factor XIII assays should be performed and the factor XIII level should be checked. The basis of treatment is replacement of the missing factor with plasma, cryoprecipitate and FXIII concentrates (2). However, in cases where there is a serious decrease in factor XIII levels, prophylaxis strategies with factor XIII concentrate can be applied to minimize bleeding events (5). In cases with recurrent delayed bleeding after trauma, factor XIII deficiency should be considered if the clotting profile is normal (2).

https://doi.org/10.1016/j.htct.2020.09.081