Estados de Hipercoagulabilidad y Eventos Isquémicos en Hemoglobinopatías

Eliezer A. Rachmilewitz

CONFERENCIA

the Edith Wolfson Medical Centrer, Holon, Israel

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ABSTRACT

Thalassaemia and sickle cell disease (SCD) represent the most common forms of hereditary haemolytic anaemia and result from a partial or complete lack of synthesis of one of the major α or β-globin chains of haemoglobin (Hb) A or from a single amino acid mutation (β6Glu →Val) of the β-globin chain, respectively. Although they have different pathophysiologies, patients with these conditions manifest both biochemical and clinical evidence of hypercoagulability. While the frequency of various thrombotic complications may vary in β-thalassaemia and homozygous sickle cell disease (sickle cell anaemia [SCA]), patients with both diseases manifest decreased levels of anticoagulant proteins, such as proteins S and C as well as increased markers of thrombin generation and platelet activation such as thrombexane A2, thrombin, antithrombin (TAT), prothrombin fragment 1.2 and D-dimer. The abnormal phospholipid membrane assymetry present in the red blood cells of β-thalassaemia and SCA patients are induced by oxidative stress documented by increased reactive oxygen species and decreased gluthatione peroxidase. Phosphatidylserine exposure on the external lipid bilayer, appears to play a significant role in the etiology of the observed hypercoagulable state by triggering formation of prothrombinase complex and increased thrombin generation which promotes fibrin clot formation and platelet activation. The available data on the aetiology and clinical manifestations of the coagulation and platelet activation that exist in both β-thalassaemia and SCA will be presented, as well as the potential therapeutic implications resulting from this hypercoagulability.