ABSTRACT

Systemic lupus erythematosis (SLE) is a multisystem disorder. Although Anti Nuclear Antibody (ANA) positivity is described as an essential criterion of diagnosis in 2019 EULAR guideline, ANA negative lupus is recognized upto 2% of all SLE patients who otherwise fulfill clinical and immunological criteria.

We describe a 20 year old female diagnosed with ANA negative SLE who presented with nephritic/nephrotic SLE relapse and adult onset seizures with high blood pressures; MRI was more in favor of lupus cerebritis than posterior reversible encephalopathy syndrome, facilities to detect CSF anti ribosomal P antibodies were not available. During hospital stay she developed severe bleeding manifestations with normal prothrombin time (PT) and activated partial thromboplastin time(APTT). However, the thrombin time(TT) was prolonged with normal fibrinogen levels and low fibrinogen clauss which was partially corrected with cryoprecipitate. Clot solubility was not performed. A diagnosis of acquired hypodysfibrinogenemia with possible factor X111 deficiency was made.

ANA-negative SLE is a clinical rarity. Differentiation between cerebral lupus and pure hypertensive encephalopathy may be challenging. Acquired coagulation disorders, which may exhibit diverse clinical and biochemical presentations, are well recognized in SLE.

Key words - hypodysfibrogenemia, systemic lupus erythematosis, ANA negative lupus, cerebral lupus