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CHARACTERIZATION AND STUDY OF CONGENITAL DEFICIENCY OF PROTEINS C AND S ASSOCIATED WITH SYSTEMIC LUPUS ERYTHEMATOSUS, SECONDARY ANTIPHOSPHOLIPID ANTIBODY SYNDROME AND MONOCLONAL GAMMOPATHY OF UNDETERMINED SIGNIFICANCE

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INTRODUCTION: The incidence of monoclonal gammopathy of undetermined significance (MGUS) was 3% in patients with systemic lupus erythematosus (SLE).

DESIGN: A 40-year-old-woman with a history of bilateral lung thromboembolism (1995) and repeated episodes of oral thrush.

METHOD: Clinical characteristics and laboratory data included: antiphospholipid antibodies (enzyme immunoassay), MTHFR gen (PCR and hybridization with specific probes for 677 position), prothrombin gen (20210 position), factor V Leiden gen, autoantibodies , immunoglobulins, capillary electrophoresis and special coagulation were obtained.

RESULTS: - Clinical characteristics : SLE, secondary antiphospholipid antibody syndrome.
- Laboratory data: leukopenia (3100), lymphopenia (1200), total protein: high (9), immunoglobulin G 3’08 g/l (similar results in 11 years). Capillary electrophoresis: monoclonal gammopathy. Antiphospholipid antibodies: Positive (ACA Ig G 63’8). MTHFR gen, prothrombin gen and Factor V Leiden gen: no mutation. Special coagulation: deficiency of proteins C and S. Antinuclear antibodies: positive (1/5120) speckled pattern. Anti-DNA autoantibody: positive (92). Anti-Sm autoantibody: positive (155). Anti-SSA: positive (121). Anti-SSB: positive (136)

CONCLUSION: 1- We describe for the first time the association between SLE, secondary antiphospholipid antibody syndrome, congenital deficiency of proteins C and S and MGUS type Ig G. 2- It is necessary to follow the evolution of MGUS every 6 months. 3- We achieved prophylactic treatment with acenocumarol (1995). 4- Antiphospholipid antibody syndrome and congenital deficiency in patients taking coagulation inhibitors is associated with an increased risk of vascular disease and is a cause of hereditary thrombotic disease. 5- Monoclonal human immunoglobulins may contain multiple autoantibody specificities including anti-DNA and anti-Sm. Such relationships could contribute to idiotypic immune regulation and control. 6- Previous reports of autoantibody properties characteristic of immunoglobulins revealed a high incidence of anti-SSA or anti-SSB activity in the sera of patients with monoclonal gammopathies; in our study the activity of both are increased.

Keyword: monoclonal gammopathy, congenital deficiency in coagulation inhibitors, systemic lupus erythematosus