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THE RELEVANCE OF "NON-CRITERIA" CLINICAL MANIFESTATIONS OF ANTIPHOSPHOLIPID SYNDROME: A CASE REPORT.

Walquíria Santos Davi (Universidade do Estado de Mato Grosso - UNEMAT, Cáceres, MT, Brasil), Bruna Pereira Pereira Santos (Universidade do Estado de Mato Grosso - UNEMAT, Cáceres, MT, Brasil), Caio Carvalho Castilho Avellar (Universidade do Estado de Mato Grosso - UNEMAT, Cáceres, MT, Brasil), Matheus Rodrigues Souza (Universidade do Estado de Mato Grosso - UNEMAT, Cáceres, MT, Brasil), Verônica Palmiro Silva Lima (Universidade do Estado de Mato Grosso - UNEMAT, Cáceres, MT, Brasil)

BACKGROUND

Antiphospholipid syndrome (APS) classification process is characterized by the presence of antiphospholipid antibodies (aPL) associated with thrombosis of arteries, veins or the microcirculation and/or well defined obstetrical manifestations. To date, there are no accepted criteria for diagnosis of this condition, although classification criteria tend to be used for definition and for diagnostic purposes. Current classification criteria for definite APS were established in a workshop, preceding the 11th International Congress on aPL in Sydney. These criteria derived from the Sapporo preliminary classification criteria for APS. However, several clinical manifestations associated with aPL are not included, and the revised classification criteria may result in negligence in the diagnosis of some cases.

CASE REPORT

Female, 51 years old, a previous history of aortic stenosis, surgery of valve replacement by metal prosthesis two years ago, already using anticoagulant therapy, she was referred to the rheumatology clinic due to polyarthralgia, chronic renal failure and positive antinuclear antibody (ANA). She denied previous history of thrombosis and abortion. At physical examination: blood pressure: 170/100 mmHg; cardiac auscultation with metallic click; absence of synovitis and presence of reticular livedo. Basic biochemistry tests: Hemogram with normocytic and normochromic anemia; Leukometry and unchanged lymphocyte counts; thrombocytopenia 79 000; increased reticulocytes; positive direct COOMBS; Urea 35; Creatinine 1.7; Proteinuria of 24 hours: 1222mg. Serological tests collected after suspension of anticoagulants: ANA 1/160 dense fine speckled nuclear pattern; complements no change; Anti-SSA negative; Anti-SSB negative; Anti-Sm negative; Anti-double helix DNA negative; Anti-RNP negative; lupus anticoagulant with strong presence; anticardiolipin IgG 130 / IgM 15; Anti-beta 2 glycoprotein 1 IgG> 100 / IgM negative; Long activated partial thromboplastin time (aPTT); Veneral Disease Research Laboratory false positive. Image exams: Doppler ultrasound of the renal arteries with bilateral parenchymal nephropathy and absence of thromboses. Renal biopsy unavailable at our service. After the manifestations: livedo reticular + valvulopathy + autoimmune hemolytic anemia + thrombocytopenia + chronic renal failure due to probable thrombotic microangiopathy? + Anti-phospholipid antibodies triple positives in high titles, we conclude that it is a primary antiphospholipid syndrome of poor prognosis.

CONCLUSION

Several times in the medical literature, it was discussed the relevance of clinical manifestations not included in the classification criteria of APS. The case analyzed, shows that none of the clinical manifestations presented by the patient are included in the revised Sapporo criterion and reveals, the need to re-discuss the inclusion of these manifestations in the criteria for classification of APS.