SBR 2019
36º CONGRESSO BRASILEIRO DE REUMATOLOGIA
CENTRO DE EVENTOS DO CEARÁ
04 A 07 DE SETEMBRO
FORTALEZA 2019
PROMOÇÃO
70 anos Sociedade Brasileira de Reumatologia
REALIZAÇÃO
SCR SOCIEDADE CEARENSE DE REUMATOLOGIA

HYPOCOMPLEMENTEMIC URTICARIAL VASCULITIS WITH RARE EXTRACUTANEOUS MANIFESTATIONS: A CASE REPORT.

Walquiria Santos Davi (Universidade do Estado de Mato Grosso - UNEMAT, Cáceres, MT, Brasil), Bruna 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), Rebeca Antunes Oliveira (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

Hypocomplementemic urticarial vasculitis (HUV) is an uncommon vasculitis of unknown etiology, rarely described in the literature. The 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitis defined the HUV like an immune complex small vessels vasculitis, because patients with HUV typically presented with low C1q complement level and normal C1 inhibitor level, in association with anti-C1q antibodies in 55% of patients. Urticarial lesions were typically erythematous papules more pruritic than painful, associated with angioedema in 51%, purpura in 35% and livedo reticularis in 14%. Extracutaneous manifestations included constitutional symptoms (56%), musculoskeletal (82%), ocular (56%), pulmonary (19%), gastrointestinal (18%) and kidney involvement (14%). Pulmonary problems of chronic obstructive airways disease and asthma occurred in 20% of patients. Rarely does pulmonary effusion or severe emphysema occur. In this case, we presents a patient with HUV associated to rarely extracutaneous manifestations.

CASE REPORT

Female, 39 years, unemployed, presenting 3 months ago recurrent urticarial lesions pruritic in the whole body, which lasted more than 24 hours in the skin and often resolves with faint residual hyperpigmentation, associated with 3 episodes of lips angioedema, polyarthralgia with arthritis affecting the hands, elbows and knees, comproved by magnetic resonance imaging. It evolved during this period with progressive dyspnea and evidence in computed tomography the lymph node enlargement in mediastinal, hilar and axillary regions and bilateral and massive pleural effusion, characterized after thoracocentesis as exudate (excluded causes as congestive heart failure with echocardiography). Infectious causes were excluded by the ADA and PCR tests performed in the pleural fluid. Laboratory data revealed decreased C1Q complement levels = 38,7 mg/ml (normal 83 - 125); another complements no changes; normal complete blood count; ANA test was positive at a titer of 1:320 and dense fine speckled nuclear pattern; Anti - SSA/SSB/RNP/DNAds/Sm non-reactive (NR); ANTI - SCL– 70 NR; Anticardiolipin antibodies and lupus anticoagulant were negative.; ANCA – P NR; ANCA – C NR. Preserved kidney function without proteinuria. Skin biopsy with evidence of leukocytoclastic vasculitis. Started treatment with corticosteroids, antihistamines and methotrexat presenting parcial response requiring association with Azathioprine immunosuppressive doses, with good follow up for 1 year.

CONCLUSION

HUV represents an uncommon systemic and relapsing vasculitis with various manifestations. This report presents a confirmated case of HUV according to Schwartz diagnoses criteria, with rare extracutaneous manifestations like pulmonary effusion and lymphadenomegaly, serving to consolidate references for the specialists as well as to extend possibilities of diagnostic criteria.