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

A RARE ASSOCIATION BETWEEN MYELODYSPLASTIC SYNDROME AND IGA VASCULITIS

THAIS CHAVES BELISARIO (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITÓRIA, ES, Brasil), KETTY LYSIE LIBARDI LIRA MACHADO (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITÓRIA, ES, Brasil), LUIZA CORRÊA RODRIGUES (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITÓRIA, ES, Brasil), ISABELLA RABELO FARIA (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITÓRIA, ES, Brasil), MARIA CARMEN LOPES FERREIRA SILVA SANTOS (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITÓRIA, ES, Brasil), LETÍCIA FONSECA FAVARATO (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITÓRIA, ES, Brasil), WEIDER ANDRADE TOMÉ (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITÓRIA, ES, Brasil), LUIZA VALLORY ALOCHIO (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, vitória, ES, Brasil), ERICA VIEIRA SERRANO (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITORIA, ES, Brasil), RUBEN HORST DUQUE (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITÓRIA, ES, Brasil), LÍDIA BALARINI SILVA (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITÓRIA, ES, Brasil), ANA PAULA ESPÍNDULA GIANORDOLI (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITÓRIA, ES, Brasil), MARIA BERNADETE RENOLDI DE OLIVEIRA GAVI (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITÓRIA, ES, Brasil), VALQUIRIA GARCIA DINIS (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITÓRIA, ES, Brasil), MIRIAM KUSTER HUBER (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITÓRIA, ES, Brasil), VALÉRIA VALIM CRISTO (UNIVERSIDADE FEDERAL DO ESPÍRITO SANTO, VITÓRIA, ES, Brasil)

BACKGROUND

Myelodysplastic syndromes (MDS) are a heterogeneous group of clonal diseases of hematopoietic stem cell characterized by functional abnormalities of hematopoietic progenitors, morphologic dysplasia, peripheral cytopenias and risk of progression to acute myeloid leukemia. This disease, however, has an exceptionally association with vasculitis,like leukocytoclastic vasculitis, polyarteritis nodosa and IgA Vasculitis (previously Henoch-Schönlein Purpura). In this following case, we present a MDS combined with IgA Vasculitis in the elderly.

CASE REPORT

A 60-years-old man presenting history of polyarthritis since 2012. In 2016 he presented fatigue, dizziness, palpitation, dyspnea, intermittent fever, dry cough and polyarthralgia. A bone marrow biopsy showed a diagnostic of MDS, being needed blood transfusions and the use of erythropoietin. In the same year, among blood transfusions, had emerged violet lesions with characteristics suggestive of vasculitis, in arms, legs and trunk. In 2017 he was referred to rheumatology of the university hospital for presenting adynamia, inflammatory polyarthralgia of small and large joints, purpura like lesions in arms and legs, occasional low fever and elevated acute-phase reactants. New analyze of bone marrow was made on this occasion, whose result confirmed MDS. Subsequently, the patient did laboratory tests, which showed thrombocytopenia, anemia, elevation of inflammatory proofs, ANA cytoplasmic dot pattern 1:320, and serum protein electrophoresis with polyclonal hypergammaglobulinemia.Thus were made the diagnostic of MDS with autoimmune polyarthritisand was initiated corticotherapy and hydroxychloroquine, in addiction two sessions of pulsetherapy. Patient had maintained follow-up, when in march of 2019, despite clinical improvement, he showed intense abdominal pain and distension, liquid-pasty diarrhea, hematochezia and purpura lesions, leading to his hospital admission. During hospitalization, evolved with acute renal failure. Skin biopsy showed a leukocytoclastic vasculitis with mesangial IgA deposits. Renal biopsy showed segmental proliferative glomerulonephritis with mesangial IgA, C3, kappa and lambda deposits. Urinanalysis were performed and presented both proteinuria and haematura. Those results confirmed the diagnosis of IgA Vasculitis.

CONCLUSION

MDS are often associated with autoimmune disease and autoimmune phenomena. Thus an investigation for such comorbidities is essential. For patients with purpura and with indefinite diagnosis, skin biopsy with immunofluorescence is recommended.