SBR 2019
36º CONGRESSO BRASILEIRO DE REUMATOLOGIA
CENTRO DE EVENTOS DO CEARÁ
04 A 07 DE SETEMBRO
PROMOÇÃO
REALIZAÇÃO

PANCREATITIS AND MACROPHAGE ACTIVATION SYNDROME AS INITIAL MANIFESTATIONS OF ADULT SYSTEMIC LUPUS.

EDUARDA ROVERE GRILL (Hospital São Lucas da PUC , PORTO ALEGRE, RS, Brasil), JOELSON FELIPE DA SILVA DE OLIVEIRA (Hospital São Lucas da PUC , PORTO ALEGRE, RS, Brasil), VANESSA DE QUADROS MARTINS (Hospital São Lucas da PUC , PORTO ALEGRE, RS, Brasil), BIANCA LOPES NOGUEIRA (Hospital São Lucas da PUC , PORTO ALEGRE, RS, Brasil), DANIELE CRISTOVAO ESCOUTO (Hospital São Lucas da PUC , PORTO ALEGRE, RS, Brasil), HENRIQUE LUIZ STAUB (Hospital São Lucas da PUC , PORTO ALEGRE, RS, Brasil)

BACKGROUND

Clinical presentation of systemic lupus erythematosus (SLE) is knowingly polymorphic. The interface of SLE with macrophage activation syndrome (MAS) is nebulous and tends to occur in juvenile SLE. We here highlight a case of pancreatitis, MAS and rapid evolution for severe SLE in an adult.

CASE REPORT

The patient, a 50 year-old African-Brazilian female, had presented a sudden abdominal pain two months before admission. Laboratory and radiological findings were compatible with pancreatitis, with no apparent cause and with spontaneous resolution. Thirty days later, she was readmitted with intense asthenia. Hyperchromic lesions were noted on extensor surfaces of hands and in buttocks. The hemogram revealed pancytopenia (hemoglobin: 7.7g/dL, leukocytes: 2130/mm³, lymphocytes: 731/mm³ and platelets: 114.000/mm³). Hyperferritinemia (2780 ng/mL) was also demonstrated. Testing for antinuclear antibodies (ANA) was initially negative. The complement profile was normal. The creatinine was 1.6 mg/dL, and non-nephrotic proteinuria was present (1 g daily). The bone marrow biopsy showed hemophagocytosis and global hypocellularity. The renal picture was responsive to corticotherapy, differently from the hematological manifestations. Four weeks after initial testing, ANA in high titers was detected (1/5120 speckled and cytoplasmic pattern). Circulating anti-Sm and anti-Ro/SSA antibodies were present, and C3 and C4 levels were now very low. Even though on corticotherapy, the patient developed features of myelopathy (paraparesis of lower limbs, urinary incontinence and protein-cytological dissociation in cerebrospinal fluid). Magnetic resonance of neuroaxis is normal for the time being. Skin biopsy was consistent with subacute lupus. Pancytopenia and myelopathy were both responsive to pulsetherapy of methylprednisolone and intravenous immunoglobulin.

CONCLUSION

The intersection of pancreatitis, MAS and SLE is intriguing. In this case, pancreatitis can be interpreted either as a manifestation of MAS or initial lupus. Of interest, the manifestations of MAS clearly preceded the laboratory diagnosis of SLE. Nevertheless, by the time pancytopenia was disclosed, skin lesions were already present, and a prozone phenomenon might explain a initial negative result for ANA. The diagnosis of SLE was thereafter based in skin, renal and neurological features, plus autoantibodies. In summary, we emphasize the presence of pancreatitis and MAS working as an atypical presentation of adult SLE.