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DIAGNOSIS OF EVANS SYNDROME PRECEDING THE DIAGNOSIS OF JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS

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BACKGROUND

Systemic lupus erythematosus (SLE) is a chronic disease that results in an inflammatory process of the blood vessels and connective tissue, associated with immunological abnormalities with alteration of the complement system and formation of immunocomplexes. The clinical presentation is varied and can occur insidiously and oligosymptomatic to an acute, potentially fatal, acute form. Evans syndrome (SE) is a rare disorder characterized by the occurrence of two or more immune hematological cytopenias, commonly autoimmune hemolytic anemia (AIHA) and autoimmune thrombocytopenia (ICP). The development of hematological changes may occur simultaneously or sequentially, usually associated with a direct positive Coombs test.

MATERIALS AND METHODS

A retrospective study was carried out, in which the medical records of 48 patients were reviewed at the Pediatric Rheumatology Service of the Children`s Hospital Darcy Vargas (HIDV) with diagnosis of JSLE established between 2007 and 2015. The study was approved by the ethics of HIDV. The diagnosis of JSLE was established according to the 1997 ACR criteria. Patients who met four or more criteria for SLE according to ACR and who had previously been diagnosed with ES were included in the study group.

RESULTS

Of the 48 patients diagnosed with JSLE between 2007 and 2015 accompanied in HIDV, five (10.4%) had previous diagnosis of ES. In all cases, the diagnosis of ES was established by the concomitant presence of AHAI and TPI. The mean platelet count at the SE diagnosis was 33,400 and the mean hemoglobin concentration was 7.38 g / dl. Only one patient had plateletometry above 50,000. The Coombs test was positive in four of the five patients (80%). Four patients (80%) had immunological exams collected at the diagnosis of ES that showed ANA positivity in all cases; in three patients the pattern described was fine-dyed and only patient 4 presented a homogeneous pattern of ANA.

CONCLUSION

We conclude that the presence of ES can precede the diagnosis of JSLE in months or years, and that in the presence of the syndrome the collection of immunological tests aiming at the early diagnosis of JSLE is extremely important, especially in girls in the pubertal phase. Although the presence of positive ANA is non-specific, even in low titers, it should be considered as an important factor in the diagnosis of SLE and indicates periodic and frequent medical follow-up in the search for other criteria for early diagnosis and treatment of JSLE.