GENETICS AND GENODERMATOSES

HLA ALLELES IN BRAZILIAN PATIENTS WITH MELKERSSON-ROSENTHAL SYNDROME

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Introduction: Melkersson-Rosenthal syndrome (MRS) is a rare neuromucocutaenous disease characterized by the triad of recurrent orofacial edema, relapsing facial paralysis and plicated tongue. Incomplete cases are common (cheilitis granulomatosa). The cause of MRS is unknown, but relation with inflammatory bowel disease is suspected.
Objective: The aim of our study was to describe the HLA alleles of patients with MRS using modern methodology (PCR-SSO).

Materials and Methods: A case-control study was performed to compare the frequency of class I and II HLA alleles of Brazilian patients with MRS with those of a healthy control group. Full local Ethical Committee approval was obtained. The chi-square test was used to compare alleles frequencies.

Results: The MRS and control group comprised 36 and 297 patients, respectively. There was an increase in the expression of HLA A*02 (p = 0.0269; OR: 1,79 [1,045-2,973]), HLA DRB1*11 (p<0,0001; OR: 4,009 [2,214-7,277]) and HLA DQB1*03 (p=0,0177; OR: 1,829 [1,122-2,978]) and an decrease in the expression of HLA A*01 (p = 0.0046, OR: 10.26 [1,859-105]), HLA DRB1*04 (p = 0.0274, OR: 4,375 [1,184-18,56]), HLA DRB1*07 (p=0,0091, OR: 5,447 [1,491-23,04]) and HLA DQB1*02 (p = 0.0051, OR: 3,204 [1,386-6,978]) in the syndrome group when compared to the control group.

Conclusions: Our study demonstrates that HLA A *02, HLA DRB1 * 11 and HLA DQB1 * 03 are associated with MRS and that HLA HLA A*01, HLA DRB1*04, HLA DRB1*07 and HLA DQB1*02 might act as protective genes. The significance of these findings remains under discussion. The main theories are that the HLA polymorphisms may present altered or microbial peptides at the site of disease, may influence on the T-cell repertoire, resulting in potential autoreactivity or may be in linkage disequilibrium with other genes, which merely acts as a marker. This is the first genetic study on MRS performed in Latin-America.