The relation between HLA-DRB1 alleles and the outcome of therapy in children with idiopathic thrombocytopenic purpura

Wafaa Ahmed El Neanaey¹, Shahira Salah Barakat, Mohamed Abdel Rahman Ahmed, Wafaa Mohamed Hasab El Nabie, Mohamed Ebrahim Sayed Ahmed

Department of Clinical Pathology, Faculty of Medicine, University of Alexandria, Alexandria, Egypt.

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Idiopathic thrombocytopenic purpura (ITP) is a common hematologic disease. The pathogenesis involves formation of autoantibodies against platelet glycoproteins. The mechanism of autoimmunity might involve binding of antigenic peptides to HLA antigens. In this study, we tried to find out if a specific HLA allele might be associated with the occurrence of ITP, and whether or not this specific allele, if present, is related to the response to treatment. We investigated the frequency of HLA-DRB1 alleles in 30 Egyptian children with documented diagnosis of ITP. All patients were followed up for at least 6 months. Ten healthy children of matched age and sex served as a control group. The alleles were identified using polymerase chain reaction (PCR) sequence specific primers. The median age of the study patients with good response was 3.94 +/- 2.31 years (range 2-10 years, female to male ratio was 2.6:1 and platelet count at presentation was 17.91 +/- 9.1 x 10(9)/L (range 10-36 x10(9)/L). For patients with poor response, female to male ratio was 3.8:1 the median age and platelet count at presentation were 4.85 +/- 2.57 years (range 2-10 years) and 29.36 +/- 24.02 x 109/L (range 10-81 x 109/L) respectively. The median duration of disease for clinically responding patients was 10.29 +/- 2.75 months (range: 6-15 months) and for non responding patients was 29.84 +/- 16.30 months (range: 6-60 months). It was found that HLA-DRB1 *14 was significantly increased in ITP patients with good response (P<0.001) while HLA-DRB1 *13 was significantly decreased in patients with good response (P=0.002, OR=0.07, CI=0.01-0.69). In conclusion, HLA-DRB1 *07 allele seems to be protective marker against ITP, HLA-DRB1 *14 allele can be used as a predictive marker for therapy in ITP patients with good response and for favourable outcome after splenectomy. Moreover, HLA-DRB1 *13 allele has an important role in resistance to therapy. Our findings indicate that genetic factors might influence the clinical course of ITP.