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Response

Immune thrombocytopenic purpura: terminology and definitions

We thank Professor Marmont for his interest in our work and for the opportunity to further discuss some controversial aspects on terminology and definitions in immune thrombocytopenic purpura (ITP).[1]

Primary ITP is a diagnosis of exclusion, characterized by a great heterogeneity in the pathogenesis and clinical outcomes.[2] The International Working Group (IWG) is aware that certain cases of primary ITP may be accompanied by coexisting antibodies such as antiphospholipid or antinuclear antibodies (ANA). However, the IWG classifies as secondary ITP only those cases in which the underlying disorder modifies the natural course or influences the treatment approach. A significant proportion of patients diagnosed with ITP has been found to have ANA. For example, in a prospective study in 186 adult patients,[3] weak positivity (titer from 1:40 to 1:80) or definite positivity (titer higher than 1:80) were found in 18 (10%) and 7 (4%) of cases, respectively. However, the impact of ANA as an adjunctive prognostic marker in isolated thrombocytopenia, otherwise meeting our criteria for primary ITP, is not defined. Although the development of other autoimmune disorders, including systemic lupus erythematosus, has been reported in a minority of cases during prolonged follow-up (around 5%),[4] in a more recent retrospective analysis of 108 adult ITP patients the presence of ANA (titer higher than of 1:80) was found in 36 (33%), but no case of systemic lupus erythematosus was recorded after a mean follow-up of 3.6 years (range, 2.1-7 years).[5] This finding was also confirmed in a prospective evaluation in patients with high ANA titer (1:160 or higher) after a similar follow-up period.[6] Regarding the less favorable response to steroid therapy, the study cited by Marmont[7] refers to a small cohort of patients (41 cases, 10 with ANA). In a larger study,[8] 39 patients with a positive test for ANA showed a response to steroids similar to that of 506 negative cases. Thus, IWG maintains that isolated ANA positivity at diagnosis should not shift toward a secondary form of ITP, unless large-scale prospective studies will provide evidence of a significant clinical impact of this finding.

We hope that Professor Marmont's comments will raise interest in such studies.

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