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Evans syndrome secondary to chronic lymphocytic leukaemia: presentation, treatment, and outcome

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Abstract

Evans syndrome (ES) is defined by the combination (either simultaneous or sequential) of immune thrombocytopenia (ITP) and autoimmune haemolytic anaemia (AIHA). When related to secondary conditions, ES may arise in patients with chronic lymphocytic leukaemia (CLL), which is frequently associated to autoimmune cytopenias (AIC). We analysed 25 patients with ES secondary to CLL, which were identified from a large series of consecutive patients with CLL, diagnosed and followed up in two institutions. They represented 2.9 % of the whole series. Thirteen patients presented with concurrent ITP and AIHA (simultaneous ES), while others developed the two AIC sequentially. Occurrence of ES was associated with unfavourable biological prognostic factors like ZAP-70 expression, unmutated immunoglobulin heavy chain variable region gene status, 17-p13 deletion and TP53 gene mutations. Of note, the majority of patients with ES (66 %) had stereotyped B cell receptor configuration. Most patients had short-lasting remissions and required second-line treatments to control the autoimmune manifestations of ES. Patients with ES were associated with inferior survival compared to patients not developing AIC, especially when ES developed early in the course of CLL, although the reduced survival was not confirmed by multivariate analysis. In conclusion, ES secondary to CLL is a difficult-to-treat complication, characterised by adverse biological features and clinical outcome.