GENETICS AND GENODERMATOSES

MACROPHAGE ACTIVATION SYNDROME IN DERMATOLOGY

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Introduction:Macrophage activation syndrome (MAS) is a severe, life-threating hematologic condition and is classified in the group of secondary hemophagocytic lymphohistiocytosis. It is observed mainly in internal medicine and reanimation. The dermatologist may be confronted with this clinical entity.

Objective: to draw the dermatologist's attention to the possible occurrence of the MAS during some dermatosis in hospitals, hence the need to know his criteria in order to establish an early diagnosis and avoid unnecessary further explorations.

Materials and methods:We collected six cases of MAS met at the dermatology department of the university hospital of Rabat, over a period of 3 years from January 2015 to June 2017.

Results: The average age of patients was 56 years. Hypertension was noted in 2 patients and diabetes in 3 patients. The MAS occurred in 3 cases of pemphigus vulgaris, 2 cases of toxidermia (Lyell and Dress syndrome) and 1case of dermatomyositis. Paraclinical investigations revealed a biological inflammatory syndrome in all cases, associated to pancytopenia in one case,bicytopenia in five cases, hypertriglyceridemia in all cases, an increase in LDH in four cases, hyperferritinemia and disruption of liver function in five cases, and blood-crust balance in three cases. The medullogram was performed in 3 patients showing signs of haemophagocytosis in a single patient. The etiological investigation revealed a tuberculosis infection in 2patients. Treatment with corticosteroids was initiated in 2 patients with immunoglobulin in a single patient and etiological treatment with anti bacillary agents in 2 patients. The evolution was favorable in 33% of the cases and the death occurred in 67% of the cases.

Conclusions:Autoimmune bullous dermatosis were ranked first in our series, followed by toxidermias. Through this study we can conclude that the MAS may be considered as a complication of the autoimmune condition, however it could be triggered by the immunosuppressive therapy as well.