CHANGES IN LYMPHOCYTE SUBPOPULATIONS AND CD3* HLA-DR+ IN CHILDREN WITH GAUCHER DISEASE

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Abstract:

Background: Gaucher Disease (GD) is the most prevalent lysosomal storage disease. GD is associated with remarkable alterations in the immune system, and GD patients are more susceptible to infections and are at a higher risk of developing autoimmune disorders and malignancies. Aim: to determine the changes in lymphocyte subpopulations and activated T lymphocytes (CD3+ HLA-DR+) in children with GD under enzyme replacement therapy (ERT) managed in Assiut Children university hospitals. Methods: This prospective case-control study was conducted among 18 children aged from 2-14 years (10 males and 8 females) with GD type 1 under enzyme replacement therapy (ERT) admitted to Assiut children university hospitals. Three-color flow cytometric immunophenotyping was used for determining the frequency of lymphocyte subpopulations and activated T lymphocytes in these patients. Results: A significant increases was found in the frequencies of total lymphocytes, CD19+, CD3+, CD4+ and CD8+ in children with GD1 when compared to healthy control. The frequencies of activated T-Lymphocytes (CD3+ HLA-DR+), activated CD4 (CD4+ HLA-DR+) and activated CD8 (CD8+ HLA-DR+) were significantly higher in GD1 as compared to healthy children. Conclusion: The increased proportion of activated T-lymphocytes in children with GD1 raises the issue of their involvement in the pathogenesis of the immune dysfunction seen in these patients. Activated T-lymphocytes could play a role in the clinical course of GD1.

Keywords:

Activated T Lymphocytes; Children; Gaucher Disease

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