Autoimmunity in Patients Hyper IgM Syndrome

Samaneh delavari[a]

a. Research Center for Immunodeficiencies, Pediatrics Center of Excellence, Children's Medical Center, Tehran University of Medical Sciences, Tehran, Iran

Abstract

Introduction: hyper-IgM (HIGM) syndrome is characterized by normal to increased serum IgM and very low or undetectable IgG, IgA, and IgE. HIGM (also known as class-switch recombination (CSR) defects) patients manifest different clinical manifestations such as autoimmune disorders. Aim of present study was to evaluate demographic data, clinical manifestation and immunological findings in HIGM patients.

Materials and Methods: Clinical features and immunological data were collected from the 79 Iranian HIGM patients' medical records diagnosed in Children's Medical Center in Iran. To compare clinical records and laboratory data, all HIGM patients were classified into two different groups patients with autoimmune disease and patients without autoimmune diseases.

Results: A total of 79 patients (60 male and 19 female) with median (IQR) age at the time of the study of 12 (6-22.45**).** Autoimmunity diseases were seen in 19 patients (23.75% , 3 females and 16 males). Among the noninfectious manifestations, the hepatomegaly and spelenomegaly were significantly higher in patients with autoimmunity (*p= 0.006)* than patients without autoimmunity (p=0.006). The most common autoimmune presentations among HIGM patients were ITP (32%), juvenile rheumatoid arthritis (16%), autoimmune hemolytic anemia (11%) and Sclerosing cholangitis (11%), Gullain-Barré syndrome, Evans syndrome, diabetes mellitus and chrohn's disease. **Conclusions:** The relation between HIGM syndrome and autoimmunity disorders could lead to sever clinical complications. So, we suggested that immunologists to be aware of this complications.

Keywords: Hyper IgM Syndrome, Autoimmunity, Immune trobocytopenia purpura.