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Case Report

Autosomal Recessive Chronic Granulomatous Disease, IgA Deficiency and Refractory Autoimmune Thrombocytopenia Responding to Anti-CD20 Monoclonal Antibody

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Abstract:

Immunodeficiency and autoimmune disease may occur concomitantly in the same individual. Some of the immunodeficiency syndromes, especially humoral defects are associated with autoimmune disorders. Hematological manifestations such as thrombocytopenia and hemolytic anemia are the most common presentations. Persistent antigen stimulation due to an inherent defect in the ability of the immune system to eradicate pathogens is the primary cause leading to autoimmunity in patients with primary immunodeficiency states.

We describe a 10 year old Iranian girl with chronic granulomatous disease -the autosomal recessive type with mutation of NCF1 gene P47- associated with selective IgA deficiency, refractory immune thrombocytopenia that showed an excellent response to Rituximab (Anti-CD20 monoclonal antibody).

Patients with primary immunodeficiencies may have variable autoimmune manifestations. So for early detection and appropriate treatment, autoimmune diseases should always be suspected in such patients.

Keywords:

Anti-CD20 ، Autoimmune thrombocytopenia ، Chronic granulomatous disease ، IgA deficiency ، Primary immunodeficiency diseases

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