The clinical significance of immunoglobulin A deficiency

ABSTRACT

IgA deficiency is the most common primary immunoglobulin deficiency. The prevalence in Caucasians is around one in 500, whereas in some Asian populations it is very uncommon. Most individuals with IgA deficiency are clinically asymptomatic. Those with symptoms of immunodeficiency have predominantly sinopulmonary or gastrointestinal infections, which are more severe when associated with IgG2, IgG4 or specific antibody deficiency. IgA deficiency is believed to be one end of a spectrum of immunodeficiency with common variable immunodeficiency at the most severe end. Although primary IgA deficiency is the most commonly encountered form, secondary deficiencies due to drugs or viral infections are recognized. IgA deficiencies can be partial or transient.

Primary IgA deficiency is caused by a defect of terminal lymphocyte differentiation, which leads to underproduction of serum and mucosal IgA; affected individuals have normal IgA genes. A number of non-immunoglobulin genes have been implicated in IgA deficiency. There have been many diseases reported in association with IgA deficiency, particularly autoimmune diseases. The most common association is with coeliac disease (CD), which has special significance since CD is usually diagnosed by detection of specific IgA antibodies that are obviously lacking in IgA deficiency. There is no specific treatment for patients with symptomatic IgA deficiency. Antibiotics are prescribed in those with acute infections. A significant proportion of IgA-deficient individuals are reported to have anti-IgA antibodies in their serum. Although blood or blood products given to IgA-deficient individuals can lead to severe, even fatal, transfusion reactions, such reactions are rare.

Keyword: IgA deficiency; Immunoglobulin A deficiency; Autoimmune disease