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Selective immunoglobulin A deficiency associated with autoimmune diseases

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Introduction: Selective immunoglobulin A deficiency (SIgAD) is the most common primary immunodeficiency. Although most patients remain asymptomatic, SIgAD is frequently associated with autoimmune and allergic diseases, while infections occur considerably less often than in common variable immunodeficiency (CVID).

The aim of the study was to examine the prevalence of SIgAD among patients with inflammatory and autoimmune diseases, to characterise their infectious risk and immunological abnormalities, and to compare their clinical and laboratory parameters with those observed in CVID. The SIgAD was diagnosed when the serum sensitive IgA level was 0.7 g/l.

Material and methods: Among patients treated for autoimmune/inflammatory conditions at the Rheumatology and Immunology Clinic of the University of Debrecen, SIgAD was confirmed in 33 patients (27 women, 6 men; average age 54.15 ±13.4 years, and average sensitive IgA 0.31 g/l). The CVID (defined as IgG 6 g/l with low IgA, IgM levels) was diagnosed in 10 patients (8 women, 2 men; average age 60.6 ±15.6 years).

Results: In the SIgAD group, recurrent non-severe infections (mainly upper respiratory and urinary tract infections) occurred in 11 patients (33.3%). Allergic diseases (asthma, pollen allergy, atopic dermatitis) were present in 19 cases (57.6%), and concomitant malignancy in 4 patients (12.1%). The underlying autoimmune diseases included rheumatoid arthritis (n = 12), systemic lupus erythematosus or rheumatoid arthritis–systemic lupus erythematosus overlap syndrome (n = 7), undifferentiated connective tissue disease (n = 8), spondyloarthropathy (n = 2), myositis (n = 1), polymyalgia rheumatica (n = 2), and scleromyxedema (n = 1). Allergic and celiac diseases occurred significantly more frequently in SIgAD than in CVID, whereas malignant diseases were significantly less common. Immunophenotypic analysis revealed that the proportion of CD8+ naïve T cells (40.44%) was significantly higher in SIgAD compared to CVID (34.4%, p = 0.05). The proportion of CD19 naïve B cells was significantly lower and showed a strong positive correlation with decreased IgA levels (R = 0.52, p = 0.003). In contrast, CD19+ IgM memory B cells were significantly increased and were associated with the more frequent occurrence of allergic and neoplastic diseases.

Discussion: Patients with SIgAD exhibit significant abnormalities in both humoral and cellular immune responses.

Conclusions: Determining the subtypes of T- and B-cells may contribute to a better understanding of disease pathogenesis and may help identify patients at increased risk for additional comorbidities.