Autoimmune Hemolytic Anemia in Primary Sjögren’s Syndrome

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Sjögren’s syndrome (SS) is an autoimmune disease characterized by the inflammation of salivary and lacrimal glands, which presents as dry eyes and dry mouth (1, 2). Such inflammation was thought to be caused by B lymphocyte hyperreactivity (1, 2). However, extra-glandular involvement such as pulmonary, renal, neurologological, and muscle involvements are also found in primary SS. Occasionally in primary SS, lymphoma, autoimmune liver disease, and autoimmune thyroid disease are noted, but autoimmune hemolytic anemia (AIHA) is rarely reported in the literature (1-3).

SS is commonly associated with leukopenia but rarely associated with other hematological abnormalities such as AIHA and idiopathic thrombocytopenic purpura (ITP). The latter complications are common findings in systemic lupus erythematosus (SLE). Some clinical and laboratory findings are found in both SLE and SS, such as arthralgia, lymphadenopathy, leucopenia, rheumatoid factor, hypergammaglobulinemia, anti-SS-A antibody. If a patient with primary SS has AIHA, it is critical to differentiate that the cytopenic complications are not due to SLE.

There are several reports of primary Sjögren’s syndrome associated with AIHA (3-8). In some cases, sicca symptom is not predominant, and the symptoms of AIHA develops earlier than the diagnosis of SS (8). In the report of Kikawada et al primary SS with AIHA which developed in an 81 year old is characteristic (9).

AIHA is a group of disorders characterized by an abnormality of the immune system that produces autoantibodies, which attack red blood cells. In AIHA, the destruction of red blood cells by autoantibodies may occur suddenly, or it may develop gradually. Some patients with autoimmune hemolytic anemia may have no symptoms, especially when the destruction of red blood cells is mild and develops gradually. When patients are elderly, they may have mild anemia. It is important to pay attention to the existence of AIHA.

AIHA, which is reported in the literature, can occur at any age from children to elderly patients (10). This autoimmune disease may associate with different kinds of autoimmune diseases such as collagen vascular diseases and organ-specific autoimmune diseases. The frequency of primary SS appears to increase with age. It is not clear whether there is any difference of clinical symptoms of SS between elderly onset and young onset SS (11). Haga and Jonsson (12) reported that young patients tend to have more frequency of autoantibodies in primary SS. Tishler et al (13) indicated that elderly onset SS has somewhat milder clinical symptoms with fewer immunological manifestations than young age onset SS.

AIHA may develop earlier than the onset of SS. The clinical presentation of SS is often quite non-specific and diverse (1, 2), suggesting that even in aged AIHA patients we should pay attention to the possible association with other autoimmune disease like primary SS.

References

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