KU Antibodies

Clinical Significance of Antibodies to the Ku Antigen

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Ku antibodies are seen in mixed connective tissue disorders, Graves' disease, and they are thought to contribute to multiple autoimmune disorders and overlap syndromes.

An Antinuclear Antibody

The anti-Ku antibody is a rare type of antinuclear antibody with either a speckled or nucleolar ANA pattern that is seen in several different autoimmune diseases. In Japan, Ku antibodies are highly associated with systemic autoimmunity, whereas in the United States Ku antibodies are most often seen in autoimmune overlap syndromes particularly scleroderma/polymyositis or dermatomyositis, mixed connective tissue diseases, and multiple autoimmune syndrome.

Diagnostic Significance

Ku antibodies are not diagnostic for any one disease. Besides occurring in multiple autoimmune and overlap syndromes, Ku antibodies are seen in scleroderma, poliomyelitis, Graves’ disease and primary pulmonary hypertension. Ku antibodies are thought to contribute to the development of multiple autoimmune conditions.

Ku antibodies have also been reported in overlap syndromes of systemic lupus erythematosus (SLE), scleroderma, and rheumatoid arthritis in patients that did not form a distinctive clinical pattern who had only mild symptoms. Anti-Ku antibody is also reported to be found in 50 percent of patients with polymyositis–scleroderma overlap syndromes and in a high number of scleroderma patients with primary pulmonary hypertension.

Symptoms

Symptoms commonly seen in patients with anti-Ku antibodies include Raynaud’s phenomenon (79 percent of patients), myositis (50 percent of patients), pulmonary fibrosis (40 percent of patients), arthralgia (86 percent of patients), esophageal reflex or esophageal dysmotility (in 35 percent of patients, and skin thickening.

Ku antibodies have been reported in a patient with Graves’ disease and autoimmune thrombocytopenic purpura who later developed a scleroderma-dermatomyositis overlap syndrome. Besides having Ku antibodies, the patient had relatively low natural killer T-cell counts. Both of these immunological features are thought to contribute to the development of multiple autoimmune diseases.
The Ku Antigen

The Ku antigen (p70/p80 antigen) is a nucleolar protein that occurs as a component of a DNA-dependent protein kinase enzyme complex involved in double-stranded DNA repair and V(D)J recombination in protein linkages. Ku antigen is induced in responses to the immune system cytokines IL-13 and IL-4 within the region of the 15-lipoxygenase-1 promoter. Dysregulated function of Ku antigen is seen in various autoimmune diseases, including systemic lupus erythematosus and thyroiditis, and it’s associated with the anti-inflammatory role of the 15-lipoxygenase-1.

Who is Affected?

Anti-Ku antibodies are primarily reported in adults between the ages of 19-80 years with a peak incidence at age 60 who have mixed connective tissue diseases, multiple autoimmune diseases, Graves’ disease, or conditions of inflammatory myositis. Females are affected more often than males. However anti-Ku antibodies may also occur in children and have been reported in a 16-year-old patient with polymyositis who developed SLE at age 7 and scleroderma at age 10. Ku antibodies and other antibodies associated with SLE have also been reported in patients with African sleeping sickness (*Trypanosoma brucei gambiense* infection).

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