

6th Annual Surgeons & Orthopedics Conference & Expo

September 12-14, 2016 Atlanta, USA



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Sjogren or sicca syndrome and IgG4 positive multiorgan lymphoproliferative syndrome

Sjogren syndrome [SS] is an inflammatory autoimmune disease affecting primarily the exocrine glands. Lymphocytic infiltrates replace functional epithelium leading to decreased exocrine secretions (exocrinopathy). Characteristic autoantibodies, anti-Ro (SS-A) and anti-La (SS-B) are produced. Mucosal dryness manifests as xerophthalmia (keratoconjunctivitis sicca), xerostomia, xerotrachea and vaginal dryness. The periepithelial extraglandular manifestations are the results of lymphocytic invasion in epithelial tissues of the lungs, kidneys and the liver. The extraepithelial manifestations, such as skin vasculitis, peripheral neuropathy and glomerulonephritis with low C4 levels, are associated with increased morbidity and high risk for lymphoma. Clinical manifestations include glandular involvement such as ocular involvement and oropharyngeal involvement; and extraglandular manifestations such as arthritis, skin involvement (purpura, annular erythema and Raynaud's phenomenon); pulmonary involvement (bronchial abnormalities and parenchymal changes), gastrointestinal and hepatobiliary features; neuromuscular involvement (mononeuritis multiplex, polyneuropathy); and renal involvement (renal tubular acidosis, glomerulonephritis). Minor salivary gland biopsy from the inferior lip is a cornerstone for the diagnosis of SS. In microscopic examination, the focal score in an area of 4 mm² of focal aggregates of at least 50 lymphocytes is sufficient for diagnosis. Another possibility to recognize SS is with an ocular staining score of 3 or greater. IgG4 positive multiorgan lymphoproliferative syndrome like Mikulicz disease (MD) is characterized by high serum levels of IgG4 and tissue biopsies showing an infiltration of IgG4+ plasma cells coupled with fibrosis or sclerosis. Compared with SS, MD does not show the same female predominance and is associated with a lower frequency of dry eyes and dry mouth, arthralgia, and serum antinuclear antibody (ANA) test positivity.

Biography

Margaret Wisłowska is the Head of the Department of Internal Medicine and Rheumatology at Szpital Kliniczny MSW, Poland. She is a Specialist in Internal Medicine, Rheumatology, Rehabilitation Medicine and Hypertension. She is the author of over 200 scientific papers and books. She has participated in numerous scientific meetings and is a Promoter of 12 PhD thesis. She took trainings at Guy and St. Thoma's Hospitals in London, Charity Hospital in Berlin and Rheumatology Institutes in Prague and Moscow. In 2003, she started the Department of Internal Medicine and Rheumatology; and in 2010 the Clinic of Internal Medicine and Rheumatology Szpital Kliniczny MSW, Poland. She is a Professor at the Warsaw Medical University, Poland.

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