中文題目：病例報告－修格蘭氏症候群表現為淚腺炎、唾液腺炎及乳腺炎，仿似IgG4相關性疾病

英文題目：Sjogren's syndrome with dacryoadenitis, sialadenitis, and mastitis mimicking IgG4-related disease: A case report

作　　者：劉家豪[1]，石硯如[2]，張詩欣[2]，吳柏樟[2]

服務單位：[1]中國醫藥大學附設醫院內科部，[2]中國醫藥大學附設醫院風濕免疫科

Abstract:

A 55-year-old woman visited our clinic with a complaint of painless lumps in bilateral upper eyelids and submandibular areas for the past 10 years. She also mentioned that she had dry eye for two years and dry mouth for two months. Magnetic resonance imaging (MRI) revealed enlargement of bilateral lacrimal, parotid, and submandibular glands; moreover, multiple enlarged lymph nodes were noted on both sides of the neck. Soft tissue sonography showed multiple hypoechoic areas with hypervascularization in the four main salivary glands and lacrimal glands, indicating sialadenitis and dacryoadenitis. The serum anti-SSA and anti-SSB antibodies were negative, while elevated serum IgG4 (IgG4: 500mg/dL, IgG: 1550mg/dL) was noted. Whole body PET-CT demonstrated intense FDG uptake in bilateral lacrimal glands, four main salivary glands, and multiple cervical lymph nodes; in addition, there was moderate FDG uptake in bilateral breasts and axillary lymph nodes. A submandibular gland biopsy and core needle biopsy of breast nodule was performed. Histopathological examination of the submandibular gland showed dense infiltration of lymphocytes, with reactive lymphoid follicles formation, which was compatible with Sjogren's syndrome. The diagnosis of IgG4-related disease was less likely since there was insufficient plasma cells infiltrations (30/HPF) in the submandibular gland. Histopathological examination of the breast nodule showed moderate lymphocytic infiltrations. Based on above findings, Sjogren's syndrome with dacryoadenitis, sialadenitis, and mastitis were considered.

Immunosuppressive therapy with pulse methylprednisolone and cyclophosphamide was given. Her enlarged lacrimal glands, salivary glands, and lymph nodes resolved thereafter.

Discussion:

IgG4-related disease is a chronic fibro-inflammatory disorder which is characterized by elevated levels of serum IgG4 and infiltration of IgG4-bearing plasma cells in the involved organs. This disease has now been described in virtually every organ system, including the salivary glands, lacrimal glands, lymph nodes, kidneys, lungs, and retroperitoneum. IgG4-related disease shares similar clinical

manifestations with primary Sjogren's syndrome, both conditions may have sicca symptoms, lacrimal and salivary glands enlargement, and multiple lymphadenopathy. In this case, image studies of the glandular lesions with markedly elevated serum IgG4 levels initially suggest the diagnosis of IgG4-related disease. However, after performing histopathological and immunohistochemistry analyses, the final diagnosis was changed to Sjogren's syndrome. These results give us information on diagnostic procedures and stress the importance of obtaining biopsies.