中文題目：第四型免疫球蛋白 G 相關疾病併淋巴腺腫大：個案報告
英文題目：IgG4 related disease with lymphadenopathy mimics with malignant - A Case Report
作　　者：蔡旻軒[1]，莊政皓[1.2]，洪仁宇[1.2],
服務單位：高雄醫學大學附設醫院[1]內科部，[2]胸腔內科

Introduction:

IgG4-related disease is a newly recognized fibro-infilammatory condition and may involve virtually every organ system. It was first time recognized as a systemic condition until 2003, when extrapancreatic manifestations were identified in patients with autoimmune pancreatitis. The diagnosis of IgG4-related disease is based on the combination of characteristic histopathologic, clinical, serologic, and radiologic findings. We reported a case of a 59-year-old male with IgG4-related disease with initial presentation as multiple lymphadenopathy.

Case report:

A 59 year-old male was a current smoker with 1 pack daily for 30 years. He had underlying disease of type 2 diabetes mellitus under medical control. Cough with sputum was complained for 1 month, accompanied with erythematous round plaques over forehead, lateral face, upper back and left lower leg. The physical examination showed lymphadenopathy over neck and right axillary, which was moveable and without tenderness. Laboratory test showed elevated eosinophil count (983/uL). Chest computed tomography(CT) revealed lymphadenopathy in the right axillary region, bilatereal upper mediastinum, precarinal space, subcarinal space and bilateral pulmonary hilae.
Therefore, the erythematous skin lesion biopsy and axillary lymph node biopsy were performed. The skin biopsied specimen revealed psoriasis vulgaris and the lymph nodes of right axilla biopsy specimen revealed IgG4-related lymphadenitis. Microscopic examination revealed reactive follicular hyperplasia with intact sinuses. Immunostaining shows numerous IgG4+ cells in the interfollicular zone with IgG4/IgG ratio more than 40%. The CD3, CD20 and Bcl-2 immunostains reveal no evidence of malignant lymphoma. We checked the IgG profile, which showed elevated IgG (3410mg/dL) and elevated IgG4 (>1300mg/dL). The diagnosis of IgG4-related disease was confirmed. Then the patient started treatment with prednisolone monotherapy (30 mg daily), and followed-up at our outpatient department regularly.

Discussion:

IgG4-related disease is characterised by tumefactive and fibro-inflammatory involvement of multiple organs or tissues with lymphoplasmacytic cells. The IgG4-related disease has been described in several organs and tissues: pancreas, biliary tract, salivary glands, periorbital tissue, kidneys, lungs, lymph nodes, meninges, aorta, breast, prostate, thyroid, pericardium, and skin.The exact prevalence of IgG4-related disease (IgG4-RD) and the involvement organ is unknown. This should be related to underestimate the true prevalence, especially because the study from which this estimate derived was done early in the development of knowledge about IgG4-related disease. The lymphadenopathy is frequently observed in patients with immunoglobulin G4-related disease (IgG4-RD) and sometimes appears as the first manifestation of the disease. In some patients with IgG4-related disease, erythematous papules, typically involving head and neck, but have also been described on the trunk and limbs. In the other hand, the clinical presentations of IgG4-related disease are heterogeneous, non-specific and subacute. The diagnosis of

IgG4-related disease under such condition can be challenging and need differentiating IgG4-related disease from malignancies and other autoimmune diseases.

Most clinical manifestations of IgG4-related disease respond to glucocorticoids, and glucocorticoids is usually the standard-of-care as the initial therapy for IgG4-related disease. Sometimes additio+nal immunosuppressive drug, such as biologic agent, particularly rituximab, is required to achieve disease remission.