中文題目：以多發性圓狀毛玻璃樣斑塊表現的 IgG4 相關性肺病

英文題目：IgG4-related lung disease presenting as multiple round shaped ground glass opacity

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Introduction

IgG4-related disease (IgG4-RD) is a rare recognized immune-mediated fibroinflammatory disease that is capable of involving various organs. Autoimmune pancreatitis, sclerosing cholangitis, chronic sclerosing sialadenitis and dacryoadenitis, orbital disease, retroperitoneal fibrosis are common and typical presentation forms. The pulmonary manifestations of IgG4 disease are rare and often nonspecific. The diagnosis depends both on pathologic and serologic reports. The IgG4-rich plasma cell lymphoplasmacytic infiltration of involved site with elevated serum IgG4 concentrations are essential for definite diagnosis. There are 4 different patterns of computed tomography (CT) image findings among patients with pulmonary IgG4 disease, including solid nodular, bronchovascular, alveolar interstitial, and round-shaped ground glass opacity

We herein present the clinical course of an IgG4 related lung disease mimics as lung cancer from an asymptomatic patient.

Case Report

A 52-year-old Asian, heavy-smoker male, with history of hypertension and gastric ulcer. He underwent a health examination and chest plain film revealed several irregular nodular opacity in the left lower lung (LLL) field. He didn't have any symptoms before examination. Chest computed tomography (CT) revealed multiple round-shaped ground-glass opacities at bilateral lung fields especially LLL. The laboratory data at baseline were within normal range. No endobronchial lesion was identified from bronchoscopic examination, and negative result of Acid-fast stain, TB-PCR (寫全名), Aspergillus Galactomannan Antigen test and cytology result showed negative for malignant cells. He then received VATS biopsy (寫全名) for further evaluation. The pathologic study revealed plasma cell infiltration of resected tissue and IgG4-positive plasma cells were found under immunohistochemical stain. Moreover, the serum IgG4 level was >1300mg/dL(寫 normal range) and all other autoimmune disease titer were within normal range. IgG4 -related pulmonary disease was finally diagnosed. After a treatment with prednisolone 30mg/day for 3 months, the follow-up serum IgG4 level decrease to ?? and Chest CT gets improving.

Discussion

IgG4-related lung disease has few subjective symptoms, and is usually detected during workup of patients with extra-thoracic lesions of IgG4 related disease.

The prevalence of IgG4 related lung disease is unclear, previous study indicate that an

assessment of 125 patients with IgG4-related disease found lung involvement in 22 (17.6%). The pathogenesis of IgG4 related lung disease remained unclear. The diagnosis is based upon histopathological and immunohistochemical assessment. The treatment of IgG4-related lung disease complies with Japanese consensus guidelines for the treatment of autoimmune pancreatitis. Initial treatment with 0.6 mg/kg/day prednisolone is recommended, followed by a gradual tapering to 5 mg/day as a maintenance dosage, depending on clinical improvements, biochemical markers include serum IgG4 and sIL2-R concentrations, and imaging.

This case reminds us that the differential diagnosis of bilateral lung multiple round-shaped-ground-glass opacity CT examination should not neglect the possibility of IgG4 related disease, a treatable disease mimicking malignancy.