■ Sun-471 ■

A case of mixed connective tissue disease with mesangial nephritis at onset

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Mixed connective tissue disease is characterized by overlapping clinical features of systemic lupus erythematosus, systemic sclerosis, polymyositis, and very high titers of anti-U1 RNP antibodies. Among MCTD, lupus nephritis is very rare, according to a population-based epidemiology data from Norway estimated the point prevalence of adult-onset MCTD to be 3.8 per 100.000 and the mean annual incidence to be 2.1 per million per year. Renal involvement is rare at the time of initial diagnosis, and pathologically, it is the first time for the domestic announcement of mesangial nephritis. We report a case of early MCTD in a patient with asymptomatic proteinuria associated with Raynaud's phenomenon, and experienced mesangial nephritis on renal biopsy. A 52-year-old man was admitted with a 3 month history of weakness and Raynaud's phenomenon. The patient did not have alcohol or tobacco, and there was no specific medical history. Both shoulder and elbow strength G3, both hip G2 muscle strength decreased, bilateral peripheral edema and cyanosis on both toes. The patient was anti-RNP antibody-positive (P: >600) with a speckled antinuclear antibody of 1:1,280. Laboratory test results included the following: LDH, 4434U/L; CK, 8990U/L; myoglobin, 2135ng/ml; Anti SS-A(Ro) Ab, positive(> 240); Anti ds-DNA IgM, P:143.6; Direct Coombs' test, positive; blood urea nitrogen, 11.6 mg/dL; creatinine 0.39 mg/dL; eGFR 137.99 mL/min/1.73m^2; proteinuria 1+; and red blood cell, 0-1/HPF in urinalysis. The amount or 24-hour proteinuria was 933mg. Thigh muscle biopsy shows dense inflammatory infiltrate and destroyed myofibril. Kidney biopsy was done, and result is Mesangial widening by increased mesangial matrix and mesangial cell proliferation (lupus nephritis class II). Consequently, the patient was diagnosed with MCTD with mesangial nephritis and we start to treat 30mg of prednisolone and 50mg of azathioprine.

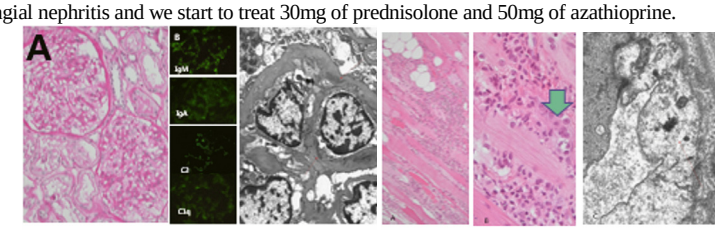

Fig.1. Kidney Biopsy shows as follows
A. Mesangial widening by increased mesangial matrix (blue arrows) with mild mesangial cell proliferation.
B. Diffuse mesangial pattern of IgM, IgA,C3 & C1q with intensity 1+ to 2+.
C. Scanty mesangial electron-dense deposit (red arrows).

Fig.2 Muscle biopsy
A. Dense inflammatory infiltrate and degenerated myofibrils.
B. Destroyed myofibril (between arrows).
C. Tubuloreticular inclusion bodies (red arrow).

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Follicular bronchiolitis mimicking lung cancer in a patient with primary Sjogren's syndrome

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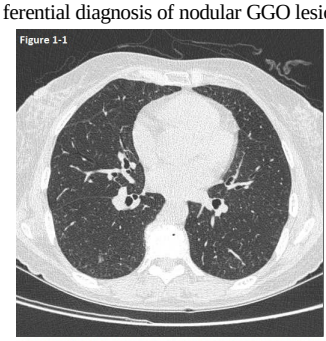
Follicular bronchiolitis (FB) is an uncommon pulmonary lymphoproliferative disorder that characterized by the presence of hyperplastic lymphoid follicles with reactive germinal centers along bronchiolar walls. FB could be associated with systemic illnesses including immunodeficiency, infection, and autoimmune diseases. But, there has been no reported case of FB associated with primary Sjogren's syndrome (SS) in Korea. We describe a case of FB presenting nodular ground-glass opacities (GGO), which mimicked lung cancer, in patients with primary SS. A 67-year-old female patient had been diagnosed with SS with interstitial lung disease 7 years ago, based on dry mouth, recurrent parotid gland swelling, hyposalivation, positive anti-Ro/SSA, and focal sialadenitis. She had an uneventful course with oral mizoribine and artificial saliva. Serial pulmonary function tests had remained stable and a nodular GGO lesion in the upper lobe of the left lung was stable over the recent 2 years. Seven months prior to admission, chest high resolution computed tomography (HRCT) showed a newly developed GGO lesion in the lower lobe of the right lung and its size increased to 11- mm over 6 months. Whole-body positron emission tomography/CT demonstrated a mild hypermetabolic focus corresponding to the GGO in the right lower lobe (RLL). She underwent superior segmentectomy of the RLL with selective mediastinal lymph node dissection under the suspicion of lung cancer. However, pathological examination revealed peribronchioloar lymphoid tissue hyperplasia that is consistent with FB. This is the first case of FB associated with primary SS patients in Korea. The differential diagnosis of nodular GGO lesion in SS patients should include FB although FB is a rare pulmonary manifestation of SS.



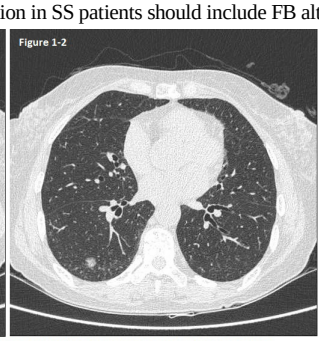


Figure 1. Serial chest CT images revealed a progressively increasing GGO nodule in the superior sement of RLL.

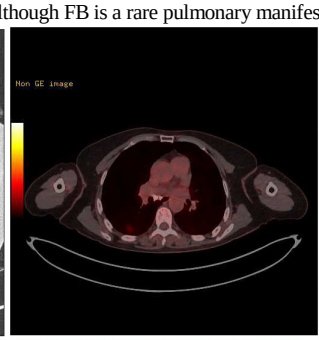


Figure 2. The PET-CT showed a GGO nodule in RLL with mild hypermetabolism

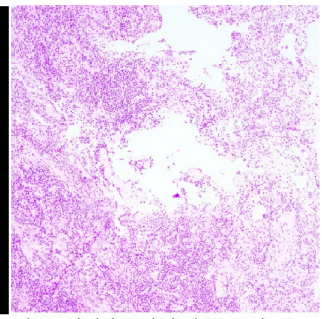


Figure 3. Histologic examination demonstrated peribronchiolar lymphoid tissue hyperplasia with polymorphous lymphoid populations (H&E, x40)