中文題目：原發性 修格連氏症候群 合併次發性器質化肺炎之罕見病例分享

英文題目：Primary Sjögren's syndrome with secondary organizing pneumonia: a case report

作 者：李文濱[1]，蔡英明[2]，張景棠[3]，曾家駿[4]，楊志仁[2]

服務單位： 高雄醫學大學附設中和醫院 內科部[1]

高雄市立大同醫院 胸腔內科[2] 心臟內科[3] 風濕免疫科[4]

BACKGROUND

Sjögren's syndrome (SS) is a complex autoimmune disease, presenting with glandular and extra-glandular manifestations. In pulmonary involvement, clinical presentations vary from dry cough to respiratory failure. In recent review of 146 histological cases, primary SS with pulmonary involvement includes non-specific interstitial pneumonitis (45%), usual interstitial pneumonitis (16%), lymphocytic interstitial pneumonitis (15%), and organizing pneumonia (OP, 11%).

CASE PRESENTATION

A 52-year-old woman presented with dry eyes, dry mouth, fever, and dyspnea. Initial chest radiography showed bilateral pulmonary consolidation, while the chest high resolution computed tomography indicated pulmonary consolidation, coral reef sign along the visceral pleura, and ground-glass pattern. Reviewing back her profiles, the laboratory tests revealed positive anti-RO antibody and negative procalcitonin. Schrimer's test confirmed the dry eye syndrome (4mm after 5 minutes). The pathologic report showed uniform temporal appearance, polypoid plugs of loose organizing connective tissue protrude into the distal airspaces, establishing the diagnosis of cryptogenic organizing pneumonia. Primary Sjögren's syndrome with secondary organizing pneumonia was impressed. The clinical condition and chest radiography got improvement after pulse therapy with methyl-prednisolone (1000mg/day) for 3 days, followed by low dose methyl-prednisolone (80mg/day) .

DISCCUSION

Current standard treatment strategy of organizing pneumonia is long-termed corticosteroids (prednisolone, 0.75 - 1.5 mg/kg/day) for 6 to 12 months. Certain evidences suggest azathioprine, cyclophosphamide, and rituximab (Anti-CD20) as the second line treatment. Pulse steroid therapy plays a role in salvage of severe or rapidly deteriorated cases.