A polyangiitis overlap syndrome: third cranial nerve palsy and dyspnea as only two manifestations

계명대학교 의과대학 내과학교실

이승준, 박재석, 김현정, 박순효, 권용식

Antineutrophil cytoplasmic autoantibody (ANCA) associated vasculitides (AAV) is a subset of small vessel vasculitis and AAV includes granulomatosis with polyangiitis (GPA), eosinophilic granulomatosis with polyangiitis (EGPA), and microscopic polyangiitis. Although previous studies have suggested diagnostic criteria, there are no definitive diagnostic criteria, and the sensitivity and specificity of these criteria are not satisfactory. Because of the difficulty in establishing appropriate diagnostic criteria, Chapel Hill Consensus Conference (CHCC) presented only the nomenclature according to disease definition. The majority of AAV was diagnosed with diagnostic criteria or categorized with nomenclature system, a polyangiitis overlap syndrome was rarely reported. We report a patient diagnosed with GPA and EGPA whose manifestations were only third cranial nerve palsy and dyspnea. The patient was a 30-year-old man with a history of asthma. He visited the emergency room due to diplopia and dyspnea. Right third cranial nerve palsy was found without ischemic brain lesion and with normal cerebrospinal fluid study. Eosinophil count was 4873/μL, Multiple consolidation and ground grass opacity were found on chest computerized tomography scan. The patient fulfilled the American College of Rheumatology diagnostic criteria for EGPA and showed positive proteinase antigen (10.9 U/mL). Diplopia and radiologic findings were not improved despite treatment of glucocorticoid (1mg/kg). We suspected the overlap of GPA and EGPA with positive proteinase antigen, surrogate maker for GPA (bloody nasal discharge), and poor response to glucocorticoid. He has received the 5 cycles or cyclophosphamide (15mg/kg) and oral glucocorticoid (1mg/kg and tapered to 20 mg daily for 5 months). The improvement of radiologic findings(Fig. 1) and diplopia(Fig.2) was found. When AAV was suspected, diagnostic criteria and nomenclature remain confusing and imperfect. Cranial nerve involvements have been reported in AAV, however, it is rare reported in overlap syndrome of GPA and EGPA. Overlap syndrome of GPA and EGPA is a rare disease that requires appropriate diagnosis and treatment.

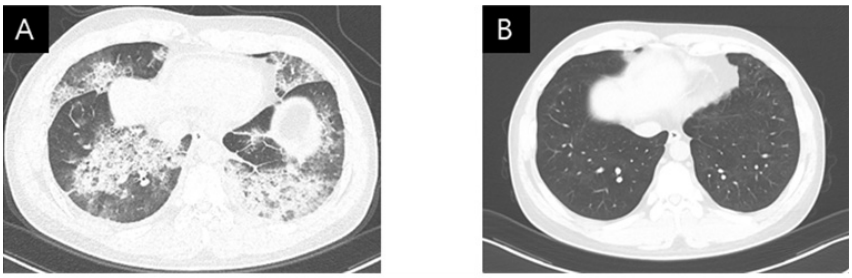

Figure 1. chest CT before(A) and after(B) treatment

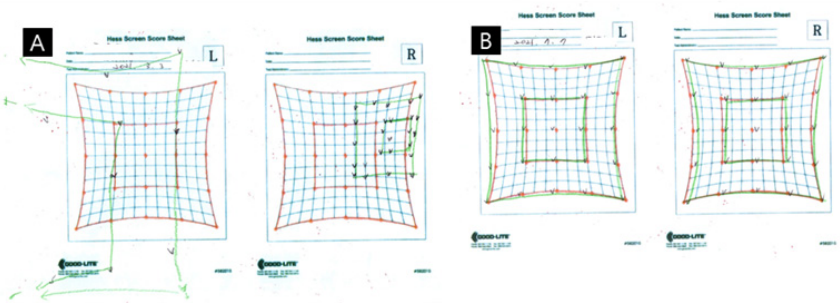

Figure 2. HESS screenig test before(A) and after(B) treatment