DOI: 10.5152/TurkThoracJ.2019.362

[Abstract:0743] PP-263 [Accepted:Poster Presentation] [Clinical Problems - Pulmonary Vascular Diseases Occupational Lung Diseases]

A Rituximab Response Resistant Granulomatois Patient with Polyanjitis

Reyhan Yıldız, Meltem Ağca, Büşra Bağlar, Deniz Uysal, Tülin Sevim

Clinic of Pulmonology, Süreyyapaşa Chest Diseases and Thoracic Surgery Training and Research Hospital, İstanbul, Turkey

Polianjiitis-associated granulomatosis (Wegener granulomatosis) is an unknown, multisystemic and necrotizing granulomatous vasculitis. It often affects the upper and lower airways, lungs and kidneys. Chest X-ray and CT scan may show granulomatous disease nodules or diffuse interstitial infiltrates that reflects alveoli and haemorrhage. Nodules are located on both sides, and can be often cavitated. Combined therapy with cyclophosphamide and methylprednisolone is recommended. Anti-TNF therapy (infliximab) or Rituximab (anti-CD20 antibody) in combination with cyclophosphamide are alternative treatments for active and resistant cases against treatment. We wanted to share the rituximab response in our case resistant to steroid and cyclophosphamide combined treatment.

60-year-old woman presented with cough, dyspnea, and chest pain. She had hypertension and diabetes and was being followed up because of diagnosis of asthma. In her physical examination, she had bilateral expirium rhonchus. In chest X-ray, intestinal bilateral infiltration was found in the lower right zone. In lung tomography, as the biggest one is on right lower lobe, a large number of diffuse parenchymal nodules and consolidation areas partially become cavitary, were observed. Bronchoscopy; In the left bronchial system, the upper lobe mucosa had oedema, mucosa at the entrance of lingula had irregular appearance, the lingula entrance was severely narrowed, right upper lobe mucosa was seen in edematous and irregular appearance (indirect tm findings), middle lobe entrance was completely closed with submucosal tm, right lower lobe segments were observed as open. Pathology reported as inflammation accompanied by eosinophils. C-ANCA (++) was diagnosed with granulomatous polyanjitis and 1 mg/kg methylprednisolone was started and clinical and radiological improvement was observed. Cyclophosphamide and pulse steroid treatment was initiated after the 6th month of treatment because of observing radiological progression. The patient was re-evaluated by rheumatology because of the continuation of radiological progression and rituximab was started. A significant regression was observed in bilateral cavitary - noncavitary lesions of the patient. The patient is presently continuing treatment with rituximab and low dose prednisolone Wegener progression is a bad vasculitis and alternative therapies should be considered in resistant cases.

Keywords: Polianjiitis-associated granulomatosis, response resistant, rituximab