A case of myelodysplastic syndrome associated with polymyalgia rheumatica and aortitis

Departments of Internal Medicine, Laboratory Medicine, University of Ulsan College of Medicine, Ulsan University Hospital, Ulsan, Korea

Joong Gi Bae, Ji Seon Oh, Hawk Kim, Seung Won Choi, Sung-Ryul Kim

Myelodysplastic syndromes (MDS) are a group of clonal hematopoietic stem cell disorders leading to peripheral cytopenias. A variety of systemic autoimmune disorders have been reported in patients with MDS. Polymyalgia rheumatica (PMR) is an inflammatory rheumatic condition characterized clinically by aching and morning stiffness in the shoulders, hip girdle, and neck. It is almost exclusively a disease of adults over the age of 50 and occurs in about 50 percent of patients with giant cell arteritis (GCA). We report a case of a 67-year-old female who presented with general weakness, weight loss, polymyalgia including the shoulders and hips, back pain with stiffness, and headache for 1 year. Laboratory investigation revealed WBC 5,840/ul, hemoglobin 8.8 g/dl, platelet 384,000/uL, ferritin 2034 ng/mL, transferin saturation 11.2%, CRP 18 mg/dL, ESR 60 mm/hr, antinuclear antibody 1:80. There was no evidence of dysplastic changes and blast in peripheral blood smear. There was no evidence of gastrointestinal bleeding in endoscopy. Chest CT revealed diffuse wall enhancement of aorta and large branching vessels, Brain MRI with angiogram showed multifocal stenosis in left superficial temporal artery. Temporal artery biopsy failed to demonstrate GCA. The patient was diagnosed with PMR and aortitis (GCA based on the American College of Rheumatology classification criteria for GCA), and the cause of anemia was considered to be related to systemic inflammatory condition. With systemic corticosteroids (methylprednisolone 1 mg/kg), her symptoms and inflammatory makers (ESR, CRP) improved rapidly, but persistent anemia was noted. Bone marrow biopsy revealed marked dysplastic changes in erythroid (40%) and megakaryocyte (16%) lineage and myeloblast (6.5%). Blasts (6%) were present in the peripheral blood. The diagnosis was made as myelodysplastic syndrome (refractory anemia with excess blasts-2). Although she was treated with 7 cycles of azacitidine, her disease evolved into acute myeloid leukemia (57% of blasts in the peripheral blood and 49% of myeloblasts in the bone marrow aspirate). The patient was treated with induction and consolidation chemotherapies with cytarabine and daunorubicin.

Etanercept treatment in a patient with ankylosing spondylitis on peritoneal dialysis

Department of Internal Medicine, Pusan National University School of Medicine, Busan, Korea

Department of Internal Medicine, Kosin University College of Medicine, Busan, Korea

Sang A Choi, Seung Geun Lee, JiMin Kim, GeunTae Kim

Treatments for patients with ankylosing spondylitis (AS) include non-steroidal anti-inflammatory drugs (NSAIDs), disease-modifying antirheumatic drugs (DMARDs) and anti-tumor necrosis factor-alpha (TNF-α) agents. However, owing to well-known nephrotoxicity of NSAIDs and some DMARDs, the use of these drugs is limited in AS patients with renal insufficiency. Meanwhile, as pharmacokinetics and metabolism of anti-TNF-α agents in patients of end stage renal disease (ESRD), especially those receiving peritoneal dialysis (PD), has not been investigated well, little is known about treating them with anti-TNF-α agents. Herein, we described the safety and efficacy of etanercept, a soluble fusion protein comprising the TNF receptor 2 in linkage with the Fc portion of immunoglobulin G, in a 40-year old male AS patient on PD. The patient was referred to our clinic due to severe inflammatory back pain, peripheral arthritis and morning stiffness despite the use of NSAIDs, sulfasalazine and glucocorticoids. At presentation, the Bath Ankylosing Spondylitis Disease Activity Index (BASDAI) and Ankylosing Spondylitis Disease Activity Score C-reactive protein (ASDAS-CRP) of the patient were 7.7 and 4.55 respectively and he was considered to have active AS. Etanercept 25mg was initiated subcutaneously twice a week. After 9 months of etanercept treatment, his symptoms improved without adverse effects on renal function and both BASDAI and ASDAS-CRP decreased to 1.2 and 1.8 respectively. Further, he achieved “major improvement” considering that difference of ASDAS-CRP between baseline and 9 month was greater than 2.0. In conclusion, our case suggests etanercept can be an effective and safe approach to treat AS patients undergoing PD.