Microscopic polyangiitis (MPA) is classified as ANCA associated vasculitis and affects many organs and/or system [1]. The clinical findings of MPA has a broad spectrum and revised 5 factor score (FFS), Birmingham vasculitis activity score (BVAS) and vasculitis damage index (VDI) are useful in follow up, monitoring disease activity and predicting prognosis. Herein, we present a case of MPA with cutaneous, articular, renal and gastrointestinal involvement (GIS), who was followed up in guidance of disease activity scores.

**CASE REPORT**

A 44-year-old male patient admitted to emergency department due to hemoptysis and palpable purpura in both legs for the last 2 days. He also had pain, cough and fever for 3 months. He had a history of otitis media operation 1 year ago and right facial paralysis was developed after that. His body temperature was 38.2 °C. Physical examination revealed right peripheral facial paralysis, hypertrophy and nodularity in both tonsils, rales in both hemithorax, arthritis in both wrists and palpable purpura in both lower extremities. In the laboratory tests; hemoglobin was 7.3 gr/dl (13.5-16.9), leucocytes 13.1 x10^3 / µL (3.91-10.9), eosinophils 0.2 x10^3 / µL (0-0.4), platelets 210 x10^3 / µL (166-308), blood urine nitrogen 115 mg/dl (0-20), creatinin 6.1 mg/dl (0.4-1.2), erythrocyte sedimentation rate (ESR) 98 mm/hour (0-20), C-reactive protein (C-RP) level was 15 mg/dl (0-5), respectively. Urine analysis revealed 4.7 gr/day proteinuria and hematuria (34 RBC/HPF), 80% of the erythrocytes were dysmorphic in microscopic evaluation. Bilateral reticulity was increased in chest X-ray (Figure 1). There was nodules, infiltration and cavity lesions in thorax computed tomography. The ANCA test was evaluated with indirect immunofluorescent assay and p-ANCA was positive. Neutrophilic small vessel vasculitis was detected in skin biopsy. Ear-nose-throat department consulted and the patient was having conductive hearing loss. Renal biopsy was performed and 3-4 of the 15 glomerulus had fibrocellular crescent and the remaining glomerules had cellular crescents. While the results were pending, he had hematemesis and melena. Multiple ulcers in different sizes were established in endoscopic evaluation (Figure 2). CMV colitis was ruled out and GIS bleeding was considered as a manifestation of MPA. So, the patient was diagnosed as MPA with cutaneous, renal, articular and GIS involvement. The patients revised FFS was 2 and BVAS score was 33.

**Figure 1**: Chest X-ray of the patient

Methylprednisolone 1 gr intravenously (3 days) followed by 1 mg/kg maintenance was initiated. Because of the impaired renal function, dose of the cyclophosphamide was calculated according to glomerular filtration rate and 500 mg/month for induction therapy was started. Therapeutic plasmapheresis was applied for 9 days. Due to decreased urine output and renal functions, hemodialysis was applied. Even the patient received cyclophosphamide, creatine levels were still increasing and he developed GIS bleeding. Considering the high FFS and BVAS scores, rituximab was initiated.

GIS, cutaneous and articular manifestations were recovered. After 2 months, creatine levels were regressed to 2.6 mg/dl and he did not need hemodialysis anymore. On the 29th month of follow-up, creatine levels were increased, metabolic asidosis despite treatment developed and he underwent hemodialysis 3 days a week. His VDI score was 2.

**DISCUSSION**

MPA generally presents with renal and pulmonary involvement. Cutaneous, nervous system and GIS involvement is relatively rare [1]. GIS involvement results with increased morbidity and mortality. The most common GIS manifestation is abdominal pain (30-58%) and GIS bleeding is reported in 21-29% of the patients [2]. As the GIS bleeding has a poor prognosis it requires early and aggressive treatment. The presenting patient's revised FFS and BVAS scores were high, and there was no improvement in clinical signs so rituximab treatment was initiated. In the published studies, switching to targeted therapy is recommended, especially the response to induction therapy is not satisfactory and the patients BVAS score is high, [3]. In patients with a revised FFS 2 and higher, 5 year mortality is defined as 45.9% [4]. Aggressive immunosuppression in such patients may be life-saving.

**KEY MESSAGES**

- GIS involvement in MPA is a relatively rare condition, and has high mortality if it is not diagnosed and treated rapidly.

- Response to standard therapy regimens is usually not satisfactory. Because of the high morbidity and mortality rates, alternative treatment options must be kept in mind.
References