acta medica

Case 3: Microscopic Polyangiitis (MPA)

Assoc. Prof. N. Şule Yaşar Bilge, MD,
ORCID: 0000-0002-0783-1072
Erdal Bodakçı, MD,
ORCID: 0000-0002-0402-1525
Timuçin Kaşifoğlu, MD
ORCID: 0000-0003-2544-8648

Microscopic polyangiitis (MPA) is classified as ANCA associated vasculitis and effects many organ 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 guidence 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, hypertrophi 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 reticularity was increased in chest X-ray **(Figure 1)**. There was nodules, infiltration and cavitary lesions in thorax computarised tomography. The ANCA test was evaluted with indirect immunofluorescent assay and p-ANCA was positive. Neutrophilic small vessel vasculitis was detected in skin biopsy. Ear-nose-throat department consultated and the patient was having conductive hearing loss. Renal biopsy was performed and 3-4 of the 15 glomerulus had fibrocellular cresent and the remaining glomerules had cellular cresents. 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.

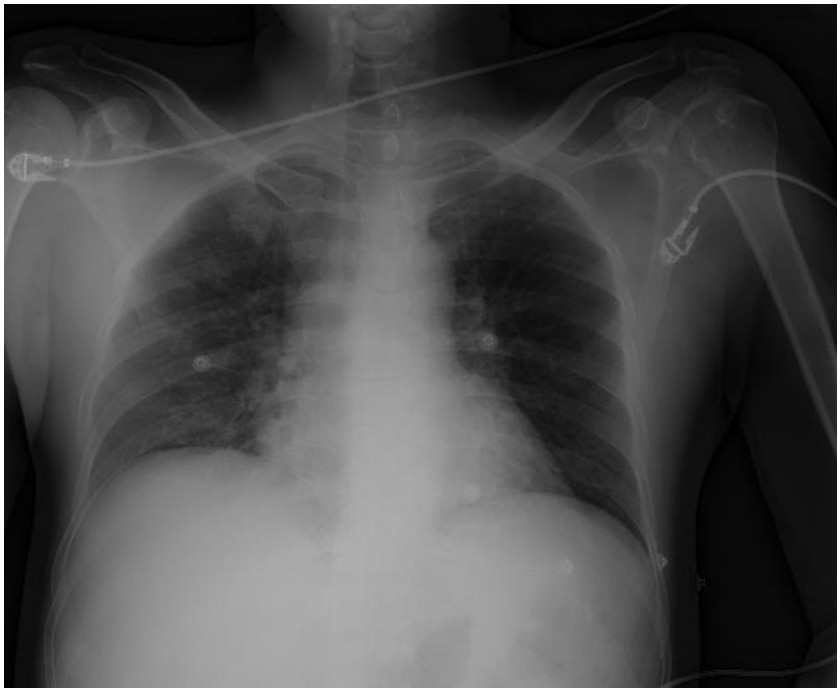

Figure 1: Chest X-ray of the patient

Eskisehir Osmangazi University School of Medicine, Department of Internal Medicine, Division of Rheumatology, Eskisehir

https://doi.org/10.32552/2021.ActaMedica.605

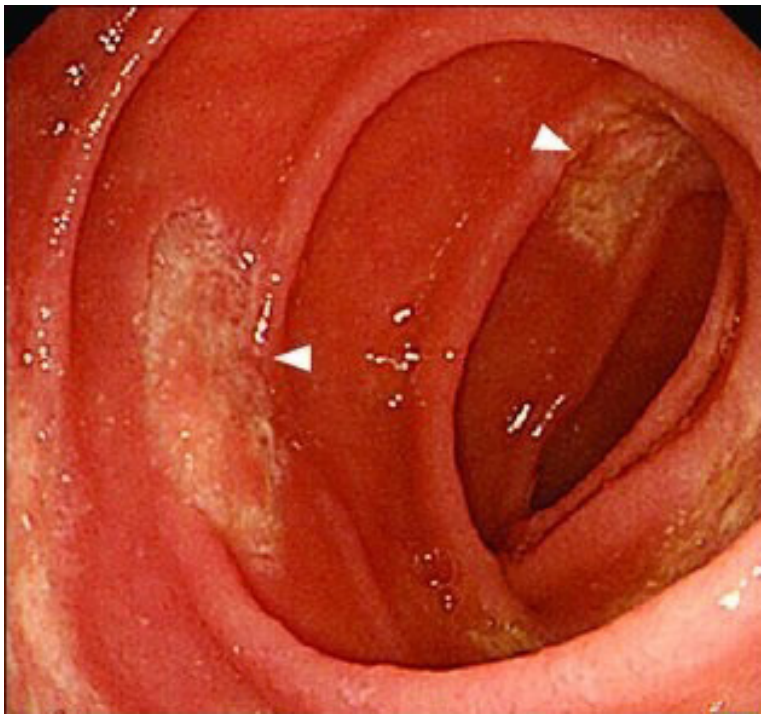

Figure 2: Endoscopic view of the jejunal ulcers

Methylprednisolone 1 gr intravenously (3 days) followed by 1 mg/kg maintance 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 recieved cyclophosphamide, creatine levels were still inreasing 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 agresive 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]. Agresive immunosupression 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 standart therapy regimens is usually not satisfactory. Because of the high morbidity and mortality rates, alternative treatment options must be kept in mind.*

References

1. Chung SA, Seo P. Microscopic polyangiitis. Rheum Dis Clin North Am. 2010 ;36(3):545-58.
2. Latus J, Koetter I, Fritz P, Kimmel M, Biegger D, Ott G, Stange EF, Amann K, Alscher DM, Braun N. Gastrointestinal involvement in granulomatosis with polyangiitis and microscopic polyangiitis: histological features and outcome. Int J Rheum Dis. 2014;17(4):412-9.
3. Eriksson P, Segelmark M, Hallböök O. Frequency, Diagnosis, Treatment, and Outcome of Gastrointestinal Disease in Granulomatosis with Polyangiitis and Microscopic Polyangiitis. J Rheumatol. 2018;45(4):529-537
4. Guillevin L, Pagnoux C, Seror R, MAhr A, Mouthon L, Le Toumelin P; French Vasculitis Study Group. The Five-Factor Score revisited: assesment of prognoses of systemic necrotising vasculitis based on the French Vasculitis Study Group cohort. Medicine (Baltimore).2011;90(1): 19-27