

## Case 2: Eosinophilic Granulomatosis with Polyangiitis (EGPA)

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**A** 74-year-old-male patient who had a history of chronic obstructive lung disease for 23 years and hearing loss in the left ear for 1 year, presented to our clinic in 2014 with fever, headache, myalgia, cough, non-productive sputum, purpuric rash, tingling and burning sensation and loss of strength in left leg for the last 20 days.

The remarkable findings upon physical examination were barrel chest deformity, prolongation expiration in breath sounds and left sided drop foot abnormality. Blood tests revealed elevated C-reactive protein (CRP, 78.4 mg/dl, N<5) and erythrocyte sedimentation rate 55 mm/h, leukocytosis (34,730/mm<sup>3</sup>), neutrophilia (6500/mm<sup>3</sup>) and eosinophilia (25,610/mm<sup>3</sup>), Proteinase 3 anti-neutrophil cytoplasmic antibody (PR3-ANCA) positivity. Electromyography (EMG) showed sensorimotor neuropathy (mononeuritis multiplex in the peroneal nerve) in the lower and upper limbs. Diffusion and cranial magnetic resonance imaging demonstrated acute thalamic infarction. Temporal artery biopsy was consistent with acute temporal arteritis and Mönckeberg medial calcific sclerosis. Given the clinical manifestations, laboratory findings, temporal artery biopsy and EMG results the diagnosis of Eosinophilic Granulomatous Polyangiitis (EGPA) and temporal arteritis was made.

The Birmingham Vasculitis Activity Score (BVAS) was calculated as 14 with clinical findings of myalgia (1), fever (2), purpura (2), headache, cerebrovascular accident and neuropathies (9). Five Factor Score (FFS) was calculated as 2 with age>65 years (1) and absence of ear, throat and nose involvement (1). The patient was treated with pulse methyl-prednisolone 1000mg daily for 3 consecutive days followed by 0.5 mg/kg/day methyl-prednisolone and intravenous cyclophosphamide 500 mg fortnightly. Six months later patient went into remission and azathioprine was used for two years as maintenance regimen. In 2017 patient presented with incident right wrist drop and recurrence of eosinophilia. Rituximab was given with fix-schedule protocol as 1000 mg fortnightly every 6 months for four cycles until his last regular follow up visit in 2018. Patient was lost to follow up until May 2020. In his last visit, patient was in remission but he was confined to wheel chair and unable to do activities of daily living due to his neuropathic sequela. Vasculitis Damage Index (VDI) was calculated as (3) with clinical findings significant muscle atrophy and weakness (1), peripheral neuropathy (1) and cerebrovascular disease (1).

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