:hronic
rt

Abstract:

Mrs.M, 50-year-old housewife and mother of 3 children was a previously known hypertensive complicated to stroke. She presented with loss of appetite and loss of weight for 2months, frothy urine with mild abdominal distension, periorbital oedema in mornings and B/L lower limb oedema for 1 month and intermittent low grade fever with constitutional symptoms for 2weeks. She had a past history of cutaneous vasculitis, maxillary sinusitis and bronchiectasis with negative TB screening. On examination she was cachectic, pale with B/L ankle oedema and mild abdominal distension.

Her investigations revealed pauci-immune crescentic glomerulonephritis, dual ANCA positivity para septal emphysema with multiple bullae and blebs involving both lung fields, which led the diagnosis of ANCA associated vasculitis possibly granulomatosis with polyangiitis (GPA) based on the American College of Rheumatology (ACR) / European League Against Rheumatism (EULAR) Provisional 2017 classification criteria for GPA.

She was started on IV Methyl prednisolone pulse followed by oral prednisolone and two weekly regime of low dose cyclophosphamide pulse with the monitoring of renal function. After 3months (following completion of 6cycles of cyclophosphamide pulse) the outcome was poor as her repeat renal biopsy showed progressive sclerotic changes and reducing renal function. Though she ended up in end stage kidney disease, she is asymptomatic clinically. At present she is on maintenance dose of oral prednisolone and close monitoring of renal function with the future plan of renal replacement therapy if necessary.