Abstract

Granulomatosis with Polyangiitis (GPA) is one of the Anti-Neutrophil Cytoplasmic Antibody (ANCA) associated vasculitis of small to medium sized vessels with characteristic upper and lower respiratory tract and renal disease with multi-system involvement.

Histologically GPA is characterised by the presence of pauci-immune necrotizing granulomatous inflammation of the blood vessels which produces symptoms related to respiratory tract and kidneys.

Here we describe a middle-aged Sri Lankan female with a history of inflammatory type polyarthritis who had vasculitic rash in extremities, constitutional symptoms, developing upper and lower respiratory tract symptoms suggestive of GPA with a solitary pulmonary cavity. There can be multiple differential diagnoses for saddle nose deformity and solitary cavitary lung lesion, including infectious, inflammatory and neoplastic disorders.

She had excellent clinical response to standard regime of corticosteroid therapy coupled with cyclophosphamide pulses leading to remission.