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An Elderly Women With Uncommon Presentation Of Granulomatosis With Polyangitis

Dr Sinchu Marium Philip[1]
Dr Antonious Maria Selvam[2]
Dr Madhan Subramanian[3]
Dr Arun Prasath[4]
Dr Jackin Moses[5]
Dr Sebin John Thampan[6]
Department Of Pulmonary Medicine, Pondicherry Institute Of Medical Sciences , Puducherry- 605014-India

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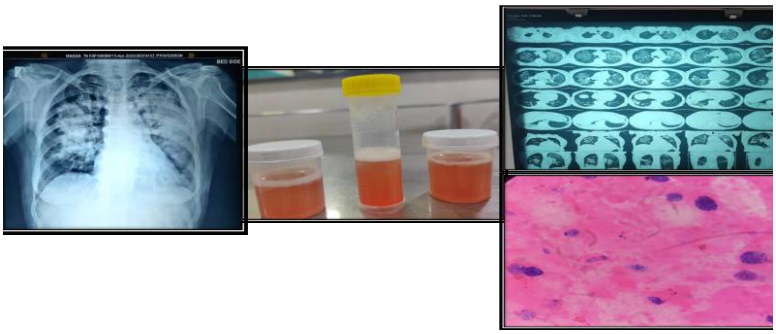
I. Background and Objectives

The annual incidence of Granulomatosis with polyangiitis (GPA) has been estimated as 4 to 12 cases per million. It is defined as a multi-organ system disease characterized by granulomatous inflammation, tissue necrosis and vasculitis with presenting age group between 40 to 50 and the sexes equally. We are hereby presenting a case scenario of an elderly female with atypical presentation of GPA.



II. Case Report:

A 79 years old female presented with complaints of dyspnoea, cough with blood stained sputum, fever and malaise for three days. On examination patient was tachypnoeic with room air saturation 85%.Routine investigations showed leukocytosis with neutrophilic predominance and Hb 5, Urine Routine showed increased RBCs. CXR showed diffuse fluffy opacities with peripheral sparing. CT scan showed pattern consistent with diffuse alveolar haemorrhage. Diagnostic Bronchoscopy was done and BAL showed haemorrhagic returns on serial aliquots. BAL cytology revealed hemosiderin laden macrophages. Further evaluation with ANA was negative and C-ANCA was found to be positive.With diagnosis of GPA patient was started on pulse steroid therapy followed by maintenance dose of cyclophosphamide. Patient improved clinically and on regular follow up CXR showed complete resolution with patient maintaining room air saturation 96%.

III. Discussion

Granulomatosis with polyangitis is defined as necrotizing granulomas in respiratory tract with focal necrotising vasculitis and glomerulonephritis. Diagnosis is based on a combination of clinical manifestation, positive C- ANCA and histological evidence of necrotising patterns. Owing to outstanding efficacy and less morbidity immunosuppresants is the treatment of choice along with steroids. With the current therapy there is a survival rate of 74-91% with mortality risk > 5 years.

IV. Conclusion

Even though usual age of presentation of DAH in GPA is around 40-50 years, we have to anticipate atypical presentation of the same in all age groups to start optimal treatment for better survival.

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