



A 26 Year Old Male with Fever, Productive Cough, Hemoptysis and Epistaxis

Authors

M. Umar. Majid M.D DNB FCCP¹, C.P. Rauf M.D²

¹Department of Pulmonology, SKIMS MC Hospital, Sringar Kashmir, India

²Chief Consultant Pulmonologist, Department of Pulmonology, Chest Hospital, Calicut Kerala India

Corresponding Author

Dr. M. Umar. Majid

Department of Pulmonology, SKIMS MC Hospital, Bemina Srinagar Kashmir India.

Email: drumarmajidrx@yahoo.com

Abstract

We are presenting a case of Granulomatosis with polyangitis (GPA) formerly known as Wegener's granulomatosis in a young non smoker male who presented with fever, productive cough, hemoptysis, headache, and epistaxis of two months duration. He was earlier treated as a case of pulmonary tuberculosis with no significant improvement. On evaluation he was found to have paranasal sinus and lung involvement and his laboratory findings showed C ANCA (PR3) =102 u/ml, tissue biopsy from a polypoidal nasal mass showed granuloma with vasculitis, based on these findings he was diagnosed as a case of GPA and treated with Cyclophosphamide and Steroids with good response.

Keywords: GPA= Granulomatosis with polyangitis, C ANCA= cytoplasmic antineutrophil cytoplasmic antibodies

Case Report

A 26y old non smoker male with no significant illness in the past, presented with fever, productive cough, hemoptysis, headache, and epistaxis of 2 months duration. He was being treated as a case of pulmonary tuberculosis with standard anti tuberculosis drugs without any significant improvement in his condition. His past medical history was uneventful and was not on any medications except for the recent anti tuberculosis drugs. Clinical examinations revealed moderately build and poorly nourished young male with stable vital signs. He had digital

clubbing grade II, red eyes, (fig.1) bleeding gums with gingival hypertrophy (fig.2) & sinus tenderness. Other systemic examination was unremarkable.

There were no clinical findings in lower respiratory tract examinations. Chest radiograph showed bilateral lower zone consolidation with right upper zone cavity (fig. 3). CT scan thorax showed thick walled cavity in apical segment of right upper lobe, collapse consolidation in the lateral segment of right middle lobe as well as inferior lingular segment of left upper lobe. Another peripheral irregular air space

opacification was seen in anterolateral basal segment of right lower lobe. (figs.4-6)

CT scan of PNS showed sinusitis with destruction of nasal septum and mass filling the middle nasal cavity. (figs.7, 8). Blood investigations including complete blood count (CBC), renal function test (RFT) urine analysis (U/A), liver function test (LFT) were within normal limits. Sputum was negative for acid fast bacilli (AFB) and tuberculin skin test was non-reactive. Serology for human immunodeficiency virus (HIV) was negative for retroviral antibody. Clinical biochemistry test showed cytoplasmic antineutrophil cytoplasmic antibodies (C ANCA) =102 u/ml. Diagnosis of granulomatosis with polyangitis was suspected and to substantiate it a tissue biopsy was taken from a polypoidal nasal mass which showed presence of edema, congestion & infiltration by lymphocytes, neutrophils & eosinophils, multiple granulomas composed of epithelioid cells and multinucleated giant cells, proliferation of capillaries with plump endothelial cells, infiltration by inflammatory cells in the wall, consistent with the diagnosis of granulomatosis with polyangitis (GPA). (fig.s 9,10,11)

Patient was put on intravenous pulse methylprednisolone 1000 mg/day for 3 days followed by oral prednisolone on day four at a dose of 1mg/kg/day for six weeks and was tapered in next 4 weeks, plus Cyclophosphamide 15mg/kg/month for 6months, and he responded well to the treatment .

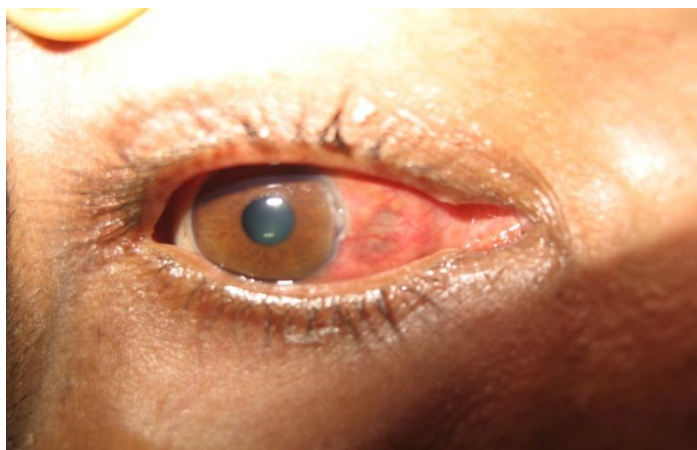


Fig. 1



Fig. 2

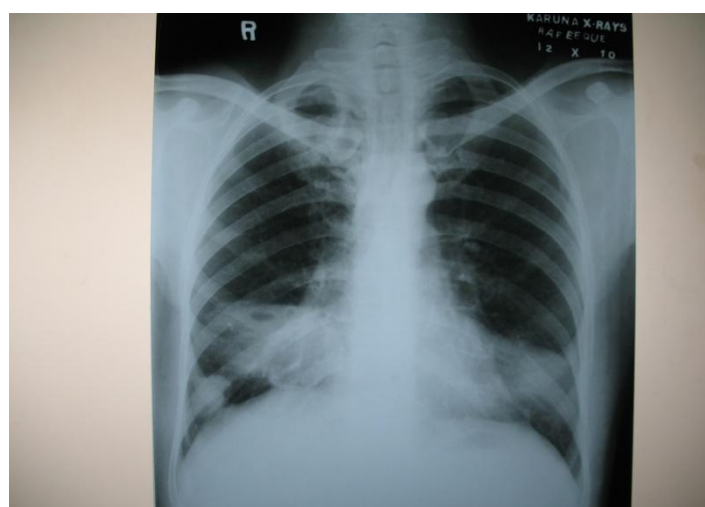


Fig. 3

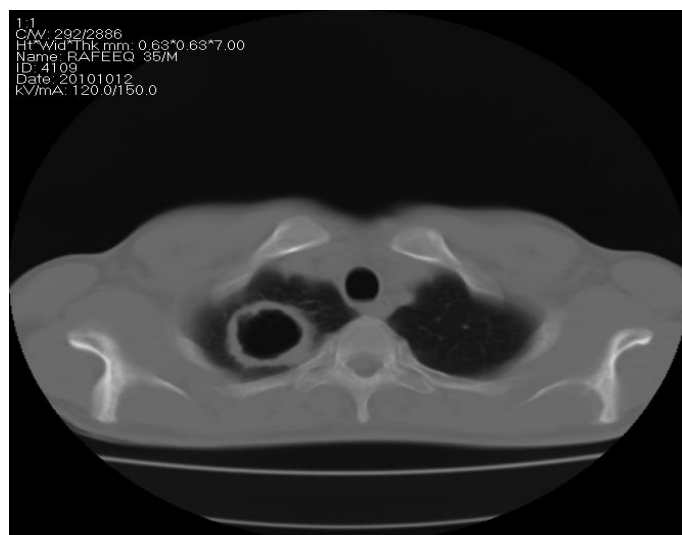


Fig.4

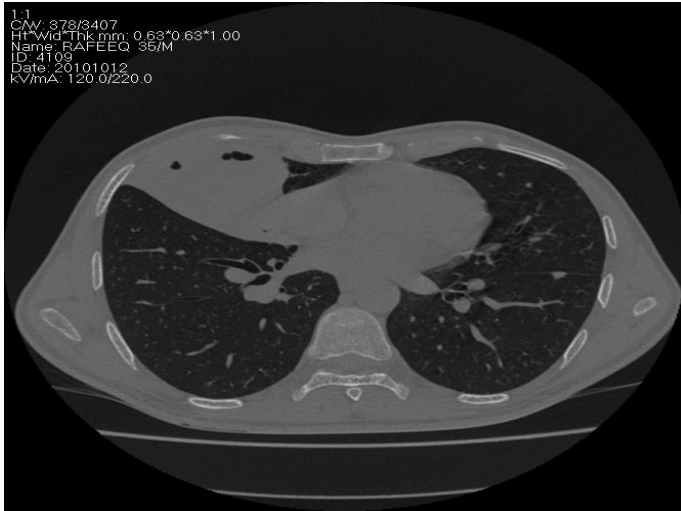


Fig. 5



Fig. 8



Fig. 6

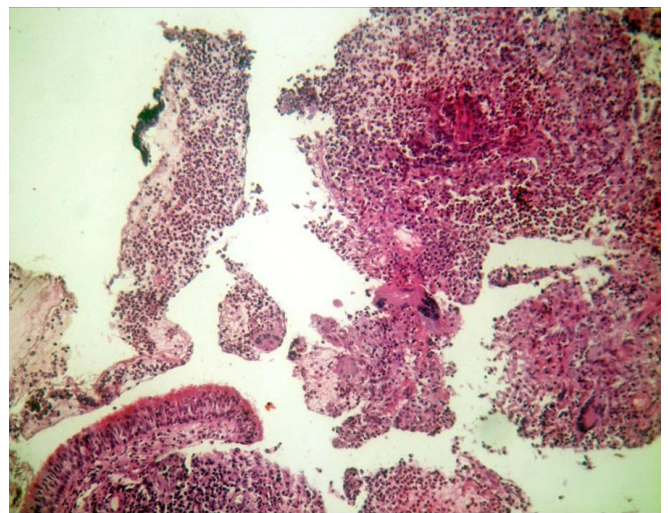


Fig. 9



Fig. 7

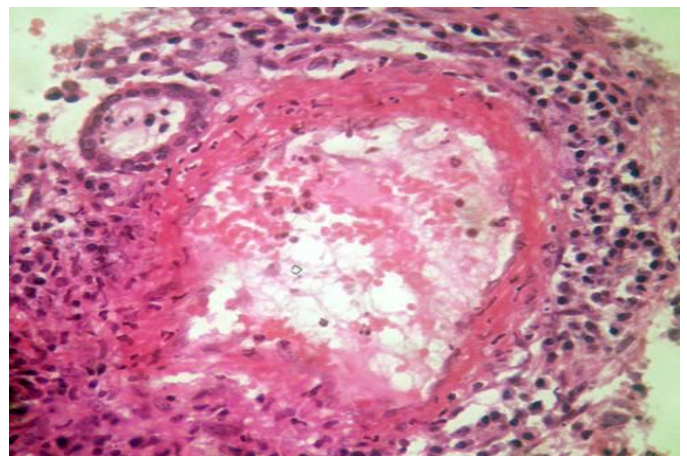


Fig. 10

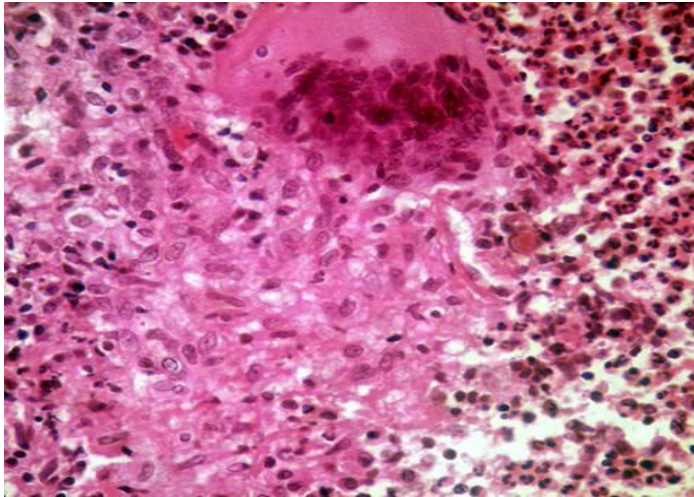


Fig.11

Discussion

In January 2011, the Boards of Directors of the American College of Rheumatology, the American Society of Nephrology, and the European League Against Rheumatism recommended that the name Wegener's granulomatosis be changed to granulomatosis with polyangiitis (Wegener's), abbreviated as GPA [1-3].

Diagnostic criteria (American college of rheumatology)

- Nasal or oral inflammation (painful or painless oral ulcers or purulent or bloody nasal discharge)
- Abnormal chest radiograph showing nodules, fixed infiltrates, or cavities
- Abnormal urinary sediment (microscopic hematuria with or without red cell casts)
- Granulomatous inflammation on biopsy of an artery or perivascular area

The presence of two or more of these four criteria yielded a sensitivity of 88 percent and a specificity of 92 percent [4]. Since the ACR did not establish specific criteria for MPA, these criteria did not discriminate GPA from either MPA or non-vasculitic diseases that can mimic GPA.

Prompt diagnosis of granulomatosis with polyangiitis (Wegener's) or microscopic

polyangiitis (GPA or MPA) is important to permit initiation of therapy that may be life-saving and organ sparing [5-8]. This may be difficult since presenting signs and symptoms are hard to distinguish from those of a patient with non-vasculitic processes such as infection or malignancy as was in our case.

Conclusion

Although a rare disease GPA should always be considered in a patient with multi organ involvement. It is important for a pulmonologist to be cautious while labeling a case with the diagnosis of smear negative pulmonary tuberculosis before ruling out the possibilities of other diseases such as GPA, and it is prudent to widen our arena of differential diagnosis even in TB endemic country like India when managing such cases.

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