

Wegener's Granulomatosis Co-Existant with Pulmonary Mycobacterium Tuberculosis (A Rare Case Report)

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Abstract: A rare case of Wegener's Granulomatosis coexistent with pulmonary tuberculosis is described with brief discussion in a 29 year old young male. Patient under follow up is keeping sound health.

Keywords: Wegener's Granulomatosis, Tuberculosis

1. Introduction

Granulomatosis with polyangiitis previously named Wegener's Granulomatosis presents as a triad of airway necrotizing granulomas, systemic vasculitis and focal necrotizing granuloma. The lungs are affected in 85-90% patients. Many entities are a part of differential diagnosis including pulmonary tuberculosis. In a country like India, where tuberculosis is very common disease, a case of Wegener's Granulomatosis presenting clinically with hemoptysis and cough, with a strong radiological support is likely to be mistaken as Tuberculosis, however coexistent lesions may exist as rarity¹. We are presenting a case which is having a dual edged sword presenting with Wegener's Granulomatosis and Tuberculosis in the same individual. The diagnosis of Wegener's Granulomatosis is based on histopathology report while that of Tuberculosis on PCR positivity in BAL. The present type of co-existent lesion bears a clinical importance as the case management widely differs in both the cases. The management profile and follow up is discussed.

2. Case Report

A 29 years old male patient presented to the Medicine wing of MGM Medical College and Hospital, Aurangabad with complaints of cough and blood stained sputum since 1 month. Initially sputum was streaked with blood and later on more quantity of blood was noted periodically. History is not suggestive of fever, weight loss, joint pain, chest pain, palpitation & breathlessness. Clinical examination revealed patient as conscious, co-operative, well oriented to time, place and person. Afebrile. Pulse was 120 beats/min regular sinus rhythm. BP was 130/80 mmHg and RR was 18 cycles/min. Oral examination revealed evidence of tonsillar congestion bilaterally. Nasal mucosa was dry with hypertrophy of inferior turbinate on left side. Systemic Examination was unrevealing. Sputum smear for AFB was found to be negative. As the patient continued to have streaks of hemoptysis, bronchoscopy undertaken revealed evidence of blood streaks, purulent secretions seen in the

right upper lobe, lower lobe and posterior segment. Bronchoscopic biopsy was undertaken from the suspicious area of bleeding and the BAL for cytology and PCR for Mycobacterium tuberculosis revealed positivity. Histopathology of the tissue showed evidence of vasculitis with perivascular granuloma with presence of eosinophil strongly favoring diagnosis of Wegener's Granulomatosis (Fig. 1, Fig. 2). Radiograph of chest revealed bilateral, peripheral, 3-5 coin shaped and fluffy shadows (Fig. 3). CT Chest shows signs of multiple nodular shadows with cavitations noted on both sides. CT PNS suggestive of minimal mucosal thickening in Right Maxillary Sinus causing partial occlusion of right infundibulum suggestive of sinusitis and mild inferior turbinate hypertrophy on left side is also noted. USG Abdomen-pelvis was unrevealing. CRP Quantitative was raised (6.60 mg/dl) (N=0.0-0.5). Immunological studies like p-ANCA was not raised (10.22 U/ml) (N-<12 U/ml) (by ELISA). c-ANCA (PR3) was significantly raised (59.66 U/ml) (N-<12 U/ml) (by ELISA) suggestive of Wegener's Granulomatosis. ANA titre was within normal limits (1:80) (by Immunofluorescence). Urine Examination, Liver Function Test, Kidney Function Test were within normal limits.

In view of dual pathology, Wegener's Granulomatosis and Tuberculosis, the patient was provided initially treatment for Tuberculosis for 2 months without chemotherapy. As per NIH protocol for Wegener's Granulomatosis, patient was given Cyclophosphamide in dose of 2mg/kg body weight/day and oral Prednisolone 1mg/kg body weight/day in tapering doses for 2 years. Chemotherapy for Wegener's Granulomatosis was initiated after 2 months of AKT therapy which was continued for 6 months. This schedule was provided in order to prevent dissemination of tuberculosis. Patient was put on Cotrimoxazole (double strength) for two months. Periodically patients X-rays and titres were carried out showing improvement clinically, serologically, radiologically & immunologically. Presently the patient is only on Chemotherapy regimen and AKT has been discontinued. It is almost over two years patient has been visiting us with no further complications of the disease or side effects of the drugs.

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3. Discussion

Wegener's granulomatosis is a rare autoimmune multisystem disorder as a result of underlying primary vasculitis involving small arteries and veins. Granulomatous inflammation of arterioles with parenchymal necrosis is a characteristic histological feature of the disease. Upper and lower respiratory tract involvement is a very classical clinical feature of Wegener's Granulomatosis as seen in the case. Though rare, Wegener's Granulomatosis can also present with ophthalmic, ear, nose, and throat affection, pulmonary, musculoskeletal, renal, nervous, cutaneous, cardiac & gastrointestinal manifestations^{2, 3}. Pulmonary involvement can be asymptomatic, insidious in onset, or severe and fulminant. Pulmonary involvement if symptomatic can present with pulmonary infiltration (71%) cough (34%), hemoptysis in (18%), chest discomfort (7%), dyspnea (8%), diffuse alveolar hemorrhages (5-45%) and atelectasis. Chronic sinusitis is the most common initial complaint in Wegener's Granulomatosis comprising in 67%, which in this case is evident from CT PNS. The case described here presents with cough and hemoptysis. CT Chest shows multiple shadows with cavitations bilaterally. X-ray Chest is also strongly suggestive of the same. In view of history and radiological findings one would think of this to be TB which was confirmed by TB PCR of BAL being positive. c-ANCA is very specific for Wegener's Granulomatosis and is being used for diagnosis and monitoring the disease. ANCA's are responsible for inflammation in Wegener's Granulomatosis. The typical ANCA's in Wegener's Granulomatosis are those which react with Proteinase 3, an enzyme prevalent in neutrophil granulocytes. In the present case c-ANCA is significantly raised (59.66 U/ml) (N=<12 U/ml) (ELISA). Non-caseating granulomatous lesion with vasculitis strongly remains consistent with the diagnosis of Wegener's Granulomatosis. Thus this confirms the co-existence of Wegener's Granulomatosis and Tuberculosis in the same individual posing it to be a great challenge in management as the management widely differs in both individual cases. Even though, Wegener's Granulomatosis would have developed dreadful complications which could prove to be fatal because of vasculitic changes in the case without the initiation of the therapy, patient was put on AKT therapy for the initial 2 months without following treatment for Wegener's Granulomatosis. The initial combined therapy would have rather flared up tuberculosis. After two months patient was provided with dual therapy for tuberculosis and Wegener's as per NIH protocol. It's been over two years patient is on chemotherapy neither with complications of the disease nor the side effects of the drugs. Under regular follow-up patient is sound.

References

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Figures & Legends to figures

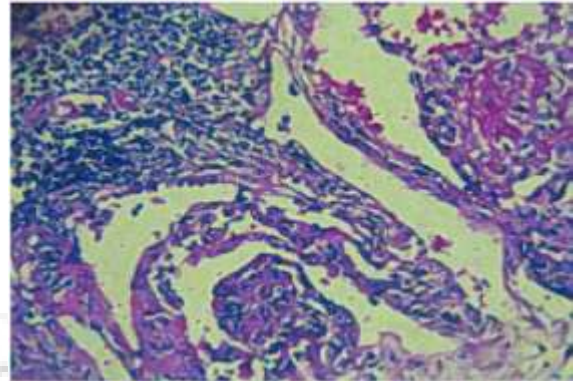


Figure 1: H&E Stain under low power Vasculitis with Perivascular Granuloma with presence of eosinophil

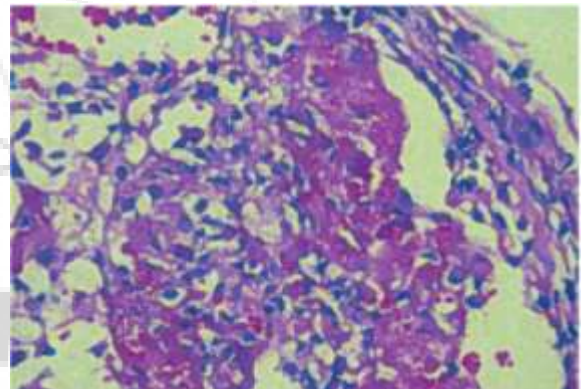


Figure 2: H&E Stain under high power- Vasculitis with Fibrinoid Degeneration



Figure 3: X-ray Chest:- B/L peripheral, 3-5 coin shaped, fluffy shadows