



WEGENER'S GRANULOMATOSIS - A CASE REPORT

Dr. C. Arshad Akeel*

MD (Gen.Med), FRCP (Glasgow), FRCP (Ireland), FRCP (Edin), FACP (USA), DABTM (USA) FCCP (USA), FICA (USA), Senior Consultant, Department of Internal Medicine, Apollo Main Hospital, Greams Road, Adjunct Professor, The Tamil Nadu Dr. MGR Medical University, Chennai *Corresponding Author

Dr. N Sadiya

MD (Pathology) , Vanagaram , Apollo Hospital, Chennai

ABSTRACT Wegener's granulomatosis (WG) is an uncommon autoimmune disease with multisystem involvement that manifests as vasculitis, granulomatosis and necrosis. While its standard form involves the upper and lower respiratory tract and kidneys, it may essentially involve any organ. We present a case in a young female patient admitted for evaluation of fever with polyarthralgia associated with conjunctival congestion, lung nodules and rashes over the left leg. This case highlights the importance of clinic-radiological correlation with histopathology and other laboratory ancillary tests for the diagnosis of this uncommon lethal condition.

KEYWORDS : Polyarthralgia, Cavitory lung nodules, Necrotizing granulomas, Wegener's granulomatosis.

INTRODUCTION:

Wegener's Granulomatosis was described in detail by Friedrich Wegener in 1936. The incidence is estimated to be 5-12 new cases /million per annum in the United States^[1]. WG is characterized by a triad of granulomatous lesions of the upper and lower respiratory tract, focal segmental glomerulonephritis and disseminated necrotizing vasculitis. The mean age at diagnosis is 40 years, but the disease can develop at an age. Other organs less commonly affected include the central and peripheral nervous system, spleen and large joints.

CASE REPORT:

A 32 year old lady was admitted with complaints of joint pain for 2 weeks associated with continuous low grade fever. She was hemodynamically stable on admission. On examination conjunctival congestion and rashes over left leg were present [Fig No 1]. She also gave a history of occasional cough with mucoid expectoration associated with intermittent haemoptysis and bleeding from the nose. No lesions in the upper respiratory tract were observed. On evaluation, laboratory parameters showed anaemia, hematuria, proteinuria, elevated ESR, CRP and hypokalemia. Her renal parameters were within normal limits. RA factor and Anti PR3 ANCA was positive. USG abdomen and Echo were normal. CT chest showed multiple nodular pleural based lesions in right middle lobe, lingula and left lower lobe with few nodules showing occasional cavitation [Fig No 2]. CT guided biopsy of lung lesion was done to rule out infectious cause. Microscopic examination showed Necrotizing Granulomatous inflammation. XPERT MTB and AFB smears were negative. Bronchoscopy showed bilateral pathology. Bronchial wash sent for AFB smear and XPERT MTB was negative. A diagnosis of Wegener's granulomatosis was rendered. The patient was treated with Pulse methylprednisolone followed by oral steroids. Inj. Cyclophosphamide 500mg was given. Patient improved symptomatically and was stable at discharge and was advised to continue oral steroids for 6 months. On follow up of patient after 4 weeks she was stable with marked resolution of the pulmonary nodules.

DISCUSSION:

Lung nodules are the most common manifestation of Wegener's granulomatosis and occur in approximately 40-70% of patients^[2] and the male to female ratio is equal. The nodules are multiple and bilateral without a zonal predilection. The size of the nodules varies from few millimeters to 10cms^[3]. Wegener's granulomatosis nodules may occur in a centrilobular distribution mimicking tuberculosis, hypersensitivity pneumonitis, or an acute viral, bacterial or fungal pneumonia. Cavitation occurs in the nodules larger than 2cms^[4].

Pathologically, WG is characterized by necrotizing granulomatous inflammation of small vessel walls, resulting in necrosis surrounded by hemorrhage, small micro abscess and granulomas within the lungs. A normocytic normochromic anaemia, leukocytosis, elevated erythrocyte sedimentation rate, positive rheumatoid factor and ANCA

(specifically PR3-ANCA) are often shown on serology. PR3-ANCA is positive in 85% of patients with multiorgan WG but this reduces to 30-40% in remission^[5]. It is frequently used in patients to assess disease activity^[2].

Other frequent but nonspecific laboratory findings include leukocytosis, thrombocytosis, an elevated erythrocyte sedimentation rate and normocytic normochromic anaemia^[6]. A tissue biopsy is essential for confirmation of diagnosis of Wegener's granulomatosis. Confirmation of diagnosis is important because therapy is often very toxic. Initial therapy consists of glucocorticoids and cyclophosphamide^[7]. Plasmapheresis is indicated when severe pulmonary hemorrhages are present^[8]. Patients with Wegener's granulomatosis have an increased risk of developing deep vein thrombosis or pulmonary embolism because of the nature of the vasculitis. In so called limited Wegener's granulomatosis, there is no kidney disease and no evidence of systemic vasculitis^[9].

Most nonfatal outcomes are related to the treatment of Wegener's granulomatosis. These include side effects from glucocorticoid therapy, increased risk of malignancy and progressive organ failure. Untreated patients have a lower survival rate of only 20% at two years. However the two year survival treated patients is about 90%

CONCLUSION:

Wegener's Granulomatosis is a rare and invariably fatal form of vasculitis, but early diagnosis and management have significant positive impact on future outcome and prognosis. High degree of suspicion is needed in TB endemic areas as mode of presentation of either disease may considerably overlap at some stage of the disease course. The study of this case emphasizes the need for careful consideration and systematic analysis of patients presenting respiratory signs and symptoms, so that the diagnosis is not delayed.



Figure 1 : Clinical photograph showing rashes in the left lower leg.

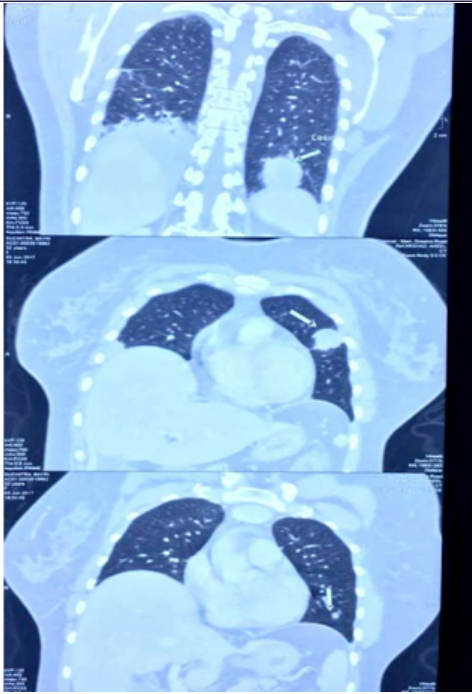


Figure 2: CT scan of chest showing pleural based nodules with cavitation.

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