

A Case Report on Limited Wegner's Granulomatosis

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Abstract: *Wegener's granulomatosis is an uncommon multi-systemic disease, characterized by necrotizing granulomatous inflammation of the upper and lower respiratory tracts and general focal necrotizing vasculitis. We described a case of 32 years old women with pansinusitis, cough without expectoration, rhinitis and joint pain. Laboratory investigations serum cytoplasmic anti-neutrophil antibody (C-ANCA) and c - reactive protein (CRP) were positive. Radiological investigation put forth a diagnosis of Wegener's granulomatosis. The patient was put on a combined therapy of prednisolone (1mg/kg) and cyclophosphamide (2mg/kg) for 2 months, which yielded positive results and provided symptomatic relief to the patient. The patient did not have renal involvement.*

Keywords: Limited Wegener's Granulomatosis, C-ANCA, c - reactive protein (CRP), Prednisolone, cyclophosphamide

1. Introduction

Wegener's Granulomatosis is an uncommon multi-systemic disease, characterized by necrotizing granulomatous inflammation of the upper and lower respiratory tracts and general focal necrotizing vasculitis. Wegener's Granulomatosis can be diagnosed if at least 2 of the 4 criteria are present namely: Nasal or oral inflammation with development of painful or painless oral ulcers or purulent or bloody nasal discharge, Abnormal chest radiograph showing the presence of nodules, fixed infiltrates, or cavities, Urinary sediment: Microhematuria (> 5 red blood cells per high power field) or red cell casts in urine sediment, Granulomatous inflammation on biopsy. Limited Wegener's Granulomatosis is a subset of Wegener's Granulomatosis lacking the renal component. The diagnosis of Wegener's Granulomatosis requires clinical acumen and correlation of a variety of laboratory, radiological, and clinical findings. Treatment regimen varies on these varieties of the disease and usually consists of steroids and immune modulators.

2. Case Report

We described a case of 32 years old woman with pansinusitis, cough without expectoration, rhinitis and joint pain. Cardiovascular system, gastrointestinal system, central nervous system and ophthalmological examination findings were normal. Liver function test and renal function test were within normal range. Sputum AFB examination was negative for tubercular bacilli. Mantoux test was positive. Chest radiography revealed multiple bilateral lateral cavities in lower zone of both lungs. HRCT revealed large areas of hypo-dense consolidation with internal irregular air pockets in apical segment of right lower lobe. She was initially treated for smear negative pulmonary tuberculosis due to bilateral nodular infiltrates in the lungs on the chest radiogram, but without expected anti-tubercular response. After 6 months of AKT, Chest radiograph revealed left sided pneumothorax for which ICDT was done. The patient was suspected for alternative diagnosis and was subjected to specific laboratory investigations to rule out any systemic illness. Antinuclear Antibody Titer (ANA) was positive, Serum Cytoplasmic Anti-Neutrophil Antibody (C-ANCA) was positive and C-Reactive Protein (CRP) was positive.

The special investigations put forth the diagnosis of Wegener's Granulomatosis not involving kidney. The patient was put on a combined therapy of prednisolone (1mg/kg) and cyclophosphamide (2mg/kg) for 2 months which yielded positive results and provided symptomatic relief to the patient. The patient did not have renal involvement.

3. Discussion

Wegener's Granulomatosis has a multi-systemic involvement with a complicated clinical sequel. The lungs are involved in 72 percent of patients and the clinical and radiographic findings indicate bilateral pulmonary nodules of varying size and definition, cavitated in half of the patients accompanied by the nodular lesion with rare involvement of the pleura. The symptoms vary from unexplained constitutional symptoms like fever and weight loss, sinusitis, oral lesions (ulcer, gingivitis), otitis media, hearing loss, epistaxis, saddle nose deformity, cough, hemoptysis, pleuritis, mild to fulminant glomerulonephritis, keratitis, conjunctivitis, scleritis, episcleritis, nasolacrimal duct obstruction, uveitis, retro-orbital pseudo tumor with proptosis, retinal vessel occlusion, and optic neuritis. ANCA is detectable in almost all cases of severe Wegener's Granulomatosis. The disease with mild to moderate manifestations gives out a confusing diagnostic paradigm. ANCA detection through immune fluorescence and ELISA gives out a precise and statistically significant outcome. Radiological investigations like X-Ray and CT scan confer high degree of specificity and sensitivity to the diagnosis by laboratory investigations. A physician must have at end to consider the possibility of Wegener's Granulomatosis in a patient with a complicated multi-systemic disease entity. Treatment regimen for Wegener's Granulomatosis depends on the severity of the disease. High dose Corticosteroid and Cyclophosphamide are necessary to induce remission. In lesser ill patients, Co-trimoxazole is effective in a dose of 960 mg twice daily. If improvement is not observed, start Prednisolone 40mg alternate days and eventually Cyclophosphamide 2mg/kg daily for those not responding to the prior treatment. Usual starting dose of Prednisolone is 1mg/kg, which may be tapered according to the condition of the patient⁶. Cyclophosphamide is started at

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2mg/kg and continued for a year after remission is achieved. If no relapse occurs, dose can be reduced to 25mg every 2 months. Pulse therapy is much more effective as it yields similar benefit at a lesser dose compared to the conventional therapy⁵. Monitoring of side effects like cystitis, bone marrow dysfunction, renal and hepato toxicity should be carefully monitored during the therapy.

4. Conclusion

Suspicion for Wegener's Granulomatosis in routine clinical practice is important for timely prediction. Continuous monitoring of treatment and side effects is very important to improve prognosis. Systemic involvement is wide spread and needs to be tackled with a cohesive and multi-pronged approach to limit its effect.

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Hypo-dense consolidation with internal irregular air pockets in apical segment of right lower lobe