A Case Report on Limited Wegner’s Granulomatosis

Dr. Ankit Vakil¹, Dr. Sumit Bochiwal², Dr. Jaydeep Padariya³

¹,²,³Resident Doctor, Dept. of Internal Medicine, Surat Municipal Institute of Medical Education and Research (SMIMER), Surat, Gujarat, India

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 cyttoplasmic 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 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 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 in filters, or cavities, Urinary sediment: Microhematuria (> 5 red blood cells per high power field) or red cell casts in urinesediment, Granulomatous inflammation on biopsy. Limited Wegener’s Granulomatosisis 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 women with pansinusitis is, 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 multi plebe 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 non-responding to the prior treatment. Usual starting dose of Prednisolone 1mg/kg, which may be titrated according to the condition of the patient. The patient did not have renal involvement.

3. Discussion

Wegener’s Granulomatosis has a multi-systemic involvement with a complicated clinicalaque. 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, nasolacrimald duct obstruction, uveitis, retro-orbital pseudo tumor with proptosis, retinal vessel occlusion, and opticneuritis. 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 encyo consider the possibility of Wegener’s Granulomatosis.

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.
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 therapy. Monitoring of side effects like cystitis, bone marrow dysfunction, renal and hepatic 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 widespread and needs to be tackled with a cohesive and multi-pronged approach to limit its effect.

References