Wegener Granulomatosis- A Case Report

Gaikwad Takale DR, Khurana R, Gaikwad S

ABSTRACT

Wegener Granulomatosis (WG) is an uncommon condition characterised by necrotising granulomatosis involving the upper and lower respiratory tract. Almost 80% of the cases eventually develop renal disease (glomerulonephritis). A case of Wegener granulomatosis in a 44 year old woman is discussed herewith emphasising on diagnosis, differentiation from Tuberculosis and treatment to avoid further complications and progress of disease.

Key Words: necrotising granulomatosis, glomerulonephritis, Wegener Granulomatosis

INTRODUCTION

Wegener Granulomatosis (WG) is an uncommon condition characterised by necrotising granulomatosis involving the upper and lower respiratory tract. Almost 80% of the cases eventually develop renal disease (glomerulonephritis). Its cause is unknown, but appears to develop as a result of an initial inflammation causing event that triggers an abnormal reaction from the immune system. The initial event may be an infection, an environmental toxin, a genetic predisposition to condition or a combination (WG is neither an infection nor a type of malignancy). The Prevalence is approximately 1 in 30000 people. Wegener granulomatosis was first described by Klinger in 1933, Wegener in 1936. It is characterised as one of the ANCA associated vasculitides distinguished clinically by its predilection for affecting the upper and lower respiratory tracts, kidneys and by the histologic presence of necrosis, granulomatous inflammation and vasculitis. Clinically WG presents by either of two types; however multisystem disease is common. Other is confined to one area of the respiratory tract.

CASE REPORT

A 47 year women presented with complaint of intermittent fever, pain in elbow and shoulder joint on both sides since one and half months, cough with expectoration since 20 days, generalized weakness, decreased appetite, gingivitis, nose pain since 15 days, blood tinged sputum and bleeding from nose since 6 to 7 days. On day 1 there was no saddling of nose, lower respiratory system examination was normal. By day 15 patients developed saddled nose. ENT surgeon’s opinion persued, examination showed nasal crusting. Earlier the patient was subjected to CT guided biopsy of lung mass which on microscopy demonstrated necrotizing granuloma and was prescribed antitubercolosis treatment, but no improvement was observed. Fresh investigation was carried out and the lab findings were: Haemoglobin 9.6, TLC 12000, neutrophils 78, eosinophils 03, lymphocytes 15, monocytes 2, platelets 475000, ESR50mm at the end of 1hr, Urine – occult blood positive, red blood cells 8-10per high power field, urinary protein 152mg/24 hour, blood urea nitrogen 8mg/dl, Serum creatinine0.6mg/dl, S. Uric acid 7.2mg/dl, Sputum- AFB negative, Bronchoscopy shows
normal with bronchoalveolar lavage-negative for Gm and Zn staining. cANCA was also positive.

Patient improved with Tab Trimethoprim-Sulphmethaxole (Septran) DS orally one tablet twice a day, Tab Prednisone 40mg OD tapered slowly, Tab Methotrexate 5mg once a week.

**Fig. 1. Photograph of patient showing saddled nose.**

**Fig. 2. Chest X ray showing bilateral multiple patchy opacities**

**Fig. 3. HRCT Thorax reported abnormal well defined mass lesion in right lower lobe and multiple heterogeneously enhancing lesions of varying sizes in bilateral lung parenchyma**

**Fig. 4. Chest X Ray showing improvement after treatment.**

**DISCUSSION**

There is a strong and specific association with antibodies directed against proteinase3, constituent of neutrophil azurophilic granules.¹ The presence of such antibodies is a strong indicator for diagnosis of WG. c ANCA has high specificity for Wegener’s granulomatosis.² Where the clinical findings are typical or consistent, and there is positive cANCA, pathologic proof may not be necessary to establish the diagnosis.³ The presence of ANCA is not required to make a diagnosis of WG by either the American College of Rheumatology (ACR) or the Chapel Hill Consensus Conference (CHCC) definitions.² Occasionally, patients with infections, neoplasms, inflammatory bowel disease, sclerosing cholangitis and other rheumatologic diseases develop ANCA, but these predominantly perinuclear ANCA or exhibit an atypical staining pattern.⁴ c- ANCA is more than 90% specific for small-vessel vasculitis in untreated WG, circulating c-ANCA are detected in more than 70% of patients.³ The clinical presentation of WG can be diverse. The list of differential diagnosis ranges from infections (fungal, bacterial & mycobacterial) to other vasculitides including Henoch-Schonlein
purpura, sarcoidosis, Behcet syndrome and malignancies. The incidence is lower (40–65%) in patients with limited disease. Mean age of onset is between 40 & 55 years and it occurs equally in both sexes.

Unexplained constitutional symptoms are often the part of initial presentation. Fever, weight loss may be reported at onset and during disease course. The upper airway disease is most common presenting feature of WG which includes sinusitis, oral ulcers or gingivitis, otitis media, hearing loss, epistaxis and saddle nose deformity. Pulmonary involvement is one of the cardinal features of WG. It occurs in 45% of patients at presentation and 87% during the course of disease. Renal disease also may be seen as the initial presentation and during the course of disease. Once present, renal disease may progress from asymptomatic and mild to fulminant glomerulonephritis within days or weeks, resulting in end-stage renal failure.

Ocular manifestations have been reported to occur in 28–50% of patients with WG, and they may be part of initial presentation in 8-16% of patients. Other unusual presentations of WG include salivary gland, cutaneous, gastrointestinal & cardiac involvement.

**Treatment:** Initial therapy for WG is oral Cyclophosphamide - corticosteroid combination therapy. This treatment has been quite effective including remission. The mean time to complete remission is 12 months, with occasional patients requiring treatment for more than 2 years before all symptoms have resolved. Response to this regimen is defined as a lessening or resolution of the inflammatory manifestations.

Other therapies for WG include methotrexate & prednisone, which is another alternative for patients with active but not immediately life-threatening disease & normal or near normal renal function.¹

In those with severe disease at presentation, pulmonary haemorrhage, or worsening disease despite immunosuppressive therapy, plasmapheresis may be indicated. Intravenous immunoglobulin G (IVIG) has therapeutic benefit in modifying the immune mediated injury associated with ANCA positive systemic vasculitis and glomerulonephritis. IVIG may provide an additional safe therapeutic option to clinicians treating patients who are not responsive to or are experiencing toxicity from conventional therapy.²

The use of aggressive immunotherapy in this disease is justified because survival in patients with untreated WG is extremely poor; up to 90% of patients die within 2 years, usually because of respiratory or renal failure. Mortality however can be significantly reduced with the introduction of a cyclophosphamide-corticosteroid therapy combination.

Our patient was treated with conventional drug Trimethoprim- Sulphamethaxol, Prednisone and Methotrexate. As patient’s renal profile was within normal limit, there was no complain of blood in urine (Occult blood in urine was advised which was positive.); Methotrexate was started orally. Patient responded to this therapy symptomatically (improved appetite, general malaise decreased, haemoptysis and epistaxis well controlled), chest roentgenogram showed resolution, no red blood cells were seen in repeat urine examination. Patient is being meticulously followed up. At present patient is in remission for more than last 8 months after stopping treatment.
AUTHOR NOTE

Deepali R Gaikwad Takale, Associate professor, (Corresponding Author);
Email: drg1818@rediffmail.com
Ritesh Khurana, Junior Resident
Sandhya Gaikwad, Junior Resident
Department of Respiratory diseases, MIMER Medical College, Talegaon Dabhade, Pune 410507

REFERENCES