



## Case Report

# Wegener's Granulomatosis- A Case Report

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### Abstract

Wegener's granulomatosis (WG) is an uncommon condition characterized by necrotizing granulomatosis of the upper and lower respiratory tract and glomerulonephritis. We describe a case of a 23-year-old man with symptoms of frequent nasal discharge with bleeding and occasional cough with blood. In this report, we wanted to emphasize that Wegener's granulomatosis, although rare, should be considered in recurrent epistaxis.

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### Introduction

Wegener's Granulomatosis (WG) is a disease of unknown etiology, which has been described for the first time by Wegener in 1936<sup>1</sup>. The prevalence of this rare disease is estimated to be 3/100000 in the United States<sup>2</sup>. WG is characterized by a triad of granulomatous lesions of the upper and lower respiratory tract, focal segmental glomerulonephritis and disseminated necrotizing vasculitis. While involvement of the upper respiratory tract and the lungs is observed in its limited form frequently seen in women, the kidneys are also involved in the common form frequently seen in men<sup>3</sup>. Direct invasion of the paranasal and paraaudal tissues by the granulomatous process, metastasis of the granulomatous process and necrotizing vasculitis cause neurological symptoms. Neurological involvement occurs in one third of the patients. It commonly manifests as peripheral neuropathy or cranial neuropathy (particularly 2, 6, and 7). ANCA is considered to be responsible for the pathogenesis together with the intervening

infections. The mortality rate is high among untreated WG cases.

### Case report

Mr. Shahin Mollah 23-year-old male farmer was admitted in RMCH with the complains of frequent purulent nasal discharge for one year, nasal bleeding for six months and occasional cough with blood for two months.

On examination patient was normotensive with mildly anaemic. Nose was disfigured, swollen and reddish. Nasal examination reveals the vestibules and septum were ulcerated and edematous. Mucosal swelling with partial obstruction noted in the right nose.

On investigation, urine R/M/E- normal, CBC- ESR-38mm in 1<sup>st</sup> hour, serum creatinine- 0.8 mg/dl, ANCA- Positive, VDRL-negative, Sputum for AFB-negative,

X- ray PNS and Chest- normal, CT scan of nose reveals soft tissue swelling is noted in the nose, more on the rt, mucosal thickening is noted within

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the left nasal cavity, Biopsy of nasal tissue revealed the soft tissue contains granulomas made of epithelioid cells, lymphocyte, many eosinophils and giant cell, vasculitic is also seen.

## Discussion

Although WG may occur at any age, the mean age of occurrence is 40 to 55 years old. The M/F ratio is 1/1. The four criteria of diagnosis defined by the American College of Rheumatology (ACR) for WG are as follows: 1) Oral or nasal ulcers, or purulent bloody flux 2) An abnormal lung radiography revealing nodules and cavities 3) An abnormal urinary sediment 4) Granulomatous inflammation in the extravascular region at biopsy. The presence of 2 or more of these criteria has a sensitivity of 88% and a specificity of 99%<sup>4,5</sup>.

As for the limited WG, the typical pathology is observed but the lungs and the kidneys are not involved. Our patient was diagnosed with WG since he had all the criteria except the kidney involvement when admitted to our clinic. The presence of nasal ulcers, the detection of a result of granulomatous inflammation at the biopsy of the soft tissue infiltrating the mucosa at that region were consistent with WG. The c-ANCA assay is being used for the diagnosis of the disease and the evaluation of the activity lately. This test with a sensitivity of 99% is 80-90% positive for the standard WG and 55-66% positive for the limited WG<sup>6</sup>. Although typical clinical findings and a positive c-ANCA result are adequate to suggest the presence of WG, biopsy is required to make a decisive diagnosis. Renal involvement is observed in 20% of the cases as the initial finding<sup>7</sup>, and identified by focal segmental glomerulonephritis. In our patient, renal USG revealed normal findings and normal findings in urine urinalyses.

The incidence of neurological involvement in WG is 50%. This includes the peripheral neuropathies in the form of polyneuropathy and mononeuritis multiplex, and multiple cranial neural involvement developing with the nasal and sinus granulomas pervading the upper cranial nerves and the pharyngeal lesions pervading the lower cranial nerves. Narrowing of the carotid siphon due to cerebral arterial involvement, cases of aneurysm

secondary to this and lateral sinus obstruction due to mastoiditis have been reported<sup>7</sup>. In untreated patients, seizures, stroke and encephalopathy may develop as the late complications<sup>8</sup>.

Our patient clinically meets the criteria of diagnosis (ACR) based on the radiology and biopsy findings. The facilitative role of positive ANCA in the confirmation of the diagnosis of WG was also positive in our patient<sup>9</sup>. Peripheral neuropathy, a neurological complication was not detected in our patient due to lack of EMG diagnostic method.

Steroids are recommended for treatment. The monitoring of sedimentation may determine the efficacy of the treatment. While the mean rate of survival was 5 months in the 1960s, now this rate has increased up to 5 years in 75% of the cases with a combination of cyclophosphamide and prednisolone administered at high doses<sup>10</sup>. In our patient, treatment was initiated with cyclophosphamide.

As a conclusion, WG should be considered in cases with biopsy and early diagnosis and treatment should be performed. If left untreated, WG is fatal, and the mean survival is 5 months in untreated patients.

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