

Wergner's Granulomatosis: A Case Study

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DOI: 10.29322/IJSRP.8.5.2018.p7722

<http://dx.doi.org/10.29322/IJSRP.8.5.2018.p7722>

Abstract- Wergner's Granulomatosis(WG) is a condition of systemic vasculitis in which the presence of circulating Anti-Neutrophil Cytoplasmic Antibody which attacks small and medium sized blood vessels. A female of age 39 was admitted with ear itching and drainage was diagnosed as WG with the help of laboratory and radiologic findings.

Index Terms- Granulomatosis, Vasculitis, Polyangitis, Vasculopathies.

I. INTRODUCTION

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis (WG) larger group of vasculitic syndromes that involves both [granulomatosis](#) and [polyangitis](#).^[2] The condition is also known as [systemic vasculitis](#) or necrotizing vasculopathies, of which feature an autoimmune attack by an abnormal type of circulating [antibody](#) termed [ANCA](#)s (antineutrophil cytoplasmic antibodies) against small and medium-size [blood vessels](#).^[1] Therefore, the signs and symptoms of GPA are highly varied and reflect which organs are supplied by the affected blood vessels. Typical signs and symptoms include [nosebleeds](#), stuffy nose and crustiness of nasal secretions, and [inflammation of the uveal layer of the eye](#). Damage to the [heart](#), [lungs](#) and [kidneys](#) can be fatal.^[2]

II. CASE REPORT

A female patient of age 39years was admitted with the complaints of left ear itching followed by drainage along with left sided facial palsy, headache and sinusitis. Her physical findings such as BP 100/60mmHg, PR 110/min, temp-101F were found abnormal. On clinical examination it was asessed that she had attic bulging along with pus on left ear and the right shows temporal perforation. While asking to the patient it was found that she had an itching sensation 1 month back followed by applied oil on the ear followed by pain and clear discharge from left ear. And CT PNS shows sphenoid sinusitis. And she had prescribed with amoxicillin-cloxacillin for 3 days thereafter persistent discharge later treated with steroid after that with ofloxacin ear drops with cefixime. There after without cure and treated with levofloxacin 500mg . CT temporal bone shows opacification left mastoid ear cells with soft tissue in middle ear cavity along with erosive changes in F.N canal and soft tissue thickening in right middle cavity. Further she develops on and off fever for 20days with dry cough. History of giddiness and

fall 2week back. She had no history of altered of altered sensorium, T2DM, trauma, ear surgery. Before admission patient was treated with antibiotics cefixime for 5 days and cefuroxime for 2days.however no pain relief was obtained.

Blood examination shows that Hb-9.2g/dl(11.5-16.5),packed cell volume 32(37-47), WBC-16,940(4000-11000), platelet count -5.36lakh(1.5-4.5) and elevated ESR-114(0-20). Liver function test shows total bilirubin 0.3mg/dl, total protein 6.3g/dl, albumin-3.1g/dl, SGPT 37U/L and GGTP 64U/L.

Vasculitis package shows that ANCA(Anti-Neutrophil Cytoplasmic Antibody) shows positive value of 1:20DIL substrate ethanol fixed human neutrophil and ANCA IgG quantitation ANTI PR3 IgG Of 93.61. Ear swab culture shows the presence of penicillin sensitive Enterococcus faecalis.

Radiologic examination chest X-ray shows necrotic mass lesion with left upper lobe with areas of necrosis and cavitations, CT image shows that thick walled fluid filled cavitory lesion in the lung cavity, lesion abducts mediastinal and chest wall pleura, wall appear hypo dense with peripheral rime of attachment, calcify nodules in right upper nodules, fibrotic strands in right middle lobe, few subcentimetric mediastinal and left hilar nodes seen. . CT temporal bone shows opacification left mastoid ear cells with soft tissue in middle ear cavity along with erosive changes in F.N canal and soft tissue thickening in right middle cavity. Perforation of the left tympanic membrane. Erosive changes are seen involving few of the air cells posteriorly around descending portion of the facial nerve canal. Minimal soft tissue thickening seen in right middle ear cavity lateral to the malleolus refraction of tympanic membrane on right side. Bronchoscopy reveals that inflamed LUL bronchi. From these data the patient was diagnosed as Wergner's Granulomatosis with lung, ear and sinus involvement. The patient was treated with inj.cyclophosphamide 500mg and inj.methylprednisolone 500mg iv twice daily as pulse therapy, T.Co-trimoxazole 400/80mg, esomeprazole 20mg twice daily and ciplox ear drops every 6hours

III. DISCUSSION

Granulomatosis with polyangiitis (GPA), formerly known as Wegener granulomatosis, is a rare multisystem autoimmune disease of unknown etiology. Its hallmark features include necrotizing granulomatous inflammation and pauci-immune vasculitis in small- and medium-sized blood vessels.^[5]

The annual incidence of GPA is estimated to be 2.1-14.4 new cases per million people in Europe. GPA is rare in [Japanese](#) and [African-American](#) populations but occurs more often in people of Northern European descent. The prevalence of

GPA in the United States is estimated to be 3 cases per 100,000 people and equally affects men and women.^[2]

The cause of GPA is unknown. [Genetics](#) have been found to play a role in GPA though the risk of inheritance appears to be low. Characteristically, granulomas form with histiocytic epithelioid cells and often with giant cells. Plasma cells, lymphocytes, neutrophils, and eosinophils are present. Inflammation affects tissues as well as vessels; vasculitis may be a small or large component of the disease. Micronecrosis, usually with neutrophils (microabscesses), occurs early. Micronecrosis progresses to macronecrosis. A central area of necrosis (called geographic necrosis) is rimmed by lymphocytes, plasma cells, macrophages, and giant cells. A zone of fibroblastic proliferation with palisading histiocytes may surround the area.^[7]

Nonspecific chronic inflammation and tissue necrosis occur in the nose. The lungs are most likely to display the full spectrum of histopathologic abnormalities, but diagnostic features are not typically identified on the small tissue samples obtained by transbronchial biopsy. In the kidneys, the most common finding is a pauci-immune crescentic focal glomerulonephritis with necrosis and thrombosis of individual loops or larger segments of the glomerulus. Vasculitic lesions and disseminated granulomas occurs only occasionally.^[4]

Before the institution of effective therapy, the mean survival of adults with untreated GPA was only 5 months, with 82% of patients dying within the first year and 90% of patients dying within the second year.^[3] Despite improvement with the use of corticosteroids, the mean survival time was increased only to 12.5 months. GPA treatment depends on the severity of the disease. Severe disease is typically treated with a combination of [immunosuppressive medications](#) such as [rituximab](#) or [cyclophosphamide](#) and high-dose [corticosteroids](#) to induce remission and [azathioprine](#), [methotrexate](#), or rituximab to keep the disease in remission. [Plasma exchange](#) is also used in severe cases with damage to the lungs, kidneys, or intestines^[3].

IV. CONCLUSION

Wegener granulomatosis (WG), also known as Granulomatosis with Polyangiitis abbreviated to GPA, typically involves the upper respiratory tract, lower respiratory tract (bronchi and lung), and kidney, with varying degrees of disseminated vasculitis. A female patient of age 39 admitted with complaints of ear drainage with pain finally diagnosed as wergner's granulomatosis with ear, sinus and lung involment. And she was treated with cyclophosphamide and prednisolone pulse therapy. Eventhough it is a rare disease but it will help to know about this condition and act as an initiator for further coming studies.

REFERENCES

- [1] Tracy L C, Diamond H, et al. Granulomatosis with Polyangiitis (Wegener Granulomatosis).
- [2] Drug and Disease. Medscape ;Dec 02, 2017. Also available as <http://reference.medscape.com/guide/rheumatology>.
- [3] <http://wikipedia/Granulomatosis with polyangiitis>.
- [4] Lynch P J, Tazelaar H M, et al. Wegener Granulomatosis (Granulomatosis with Polyangiitis): Evolving Concepts in Treatment.
- [5] Kumar A, Pandhi A, et al. Wegener's granulomatosis in India: clinical features, treatment and outcome of 25 patients. *Chest Dis Allied Sci* 2001;43:197-204.
- [6] Swain S, Ray R. Wegener's Granulomatosis of Nose: A Case Report *Indian J Otolaryngol Head Neck Surg* 2011; 63(4):402-404.
- [7] Forte V A. Granulomatosis with Polyangiitis (GPA) (Wegener Granulomatosis; Wegener's Granulomatosis). Merck Manual Professional Version. Also available in <https://www.merckmanuals.com/professional/musculoskeletal-and-connective-tissue-disorders/vasculitis/granulomatosis-with-polyangiitis-gpa>.

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