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Case Report

A case report on wegener's granulomatosis

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ABSTRACT

Wegener's Granulomatosis, also known as granulomatosis with polyangiitis, is a rare multisystem autoimmune disease with an unknown etiology that can affect persons of all ages. In most cases, the condition affects the respiratory tract and the kidneys. The initial symptoms include a runny nose (rhinorrhea), nasal crusting, and nasal blockage or congestion. More significant symptoms include nose bleeds, ulcerations of the nasal mucous membranes with subsequent bacterial infection, sinus discomfort, sinus inflammation, and hoarseness. The patient was presented with symptoms like reddish staining of the eyes, hearing loss, and numerous joint discomfort and swelling of the lower limbs. The antineutrophil cytoplasmic antibody, cytoplasmic (C-ANCA), and human leukocyte antigen- B27 (HLA-B27) were both positive, and renal biopsy revealed necrotizing and crescentic glomerulonephritis. The treatment was initiated with corticosteroids and monoclonal antibodies. The patient responded positively to the therapy, and her health appears to be improving.

Keywords: Wegener's granulomatosis, Monoclonal antibodies, Corticosteroids, Human leukocyte antigen, Glomerulonephritis

INTRODUCTION

Wegener's granulomatosis, also known as granulomatosis with polyangiitis, is a rare multisystem autoimmune disease of uncertain cause that can affect people of all ages. It is characterized by inflammation of various tissues, blood vessels, and predominantly, parts of the respiratory tract and kidneys. It is an antineutrophil cytoplasmic antibody (ANCA) associated with vasculitis that affects small to medium-sized blood vessels. A peak incidence of Wegener's granulomatosis is observed at 64-75 years of age, with a prevalence of 8 per million.[1] It occurs in all racial groups with a greater incidence in white Caucasian populations of European descent than in African Americans.[3]

HLA-DPB1 gene is the strongest genetic risk factor for developing this condition. Approximately 90% of people with Wegener's granulomatosis have an anti-neutrophil cytoplasmic antibody (ANCA) in their blood, which attacks normal human protein proteinase 3 (PR3) leading to inflammation thus, the changes in the HLA-DPB1 gene are responsible for the autoimmune response which sequentially triggers inflammation, predominantly in kidney and respiratory tract. HLA-B27 gene is a type of protein present in white blood cells and helps our immune system to attack foreign bodies, changes in this gene lead to autoimmune disorders.[1]

Wegener's Granulomatosis displays a spectrum of clinical manifestations with a different immunopathogenesis. The triad of Wegener's Granulomatosis includes granuloma formation, pauci-immune vasculitis, and glomerulonephritis. The interaction between polymorphonuclear neutrophils and endothelial cells via cell adhesion molecules is responsible for ANCA mediated vasculitis which primarily affects the nose, lungs, and kidneys. It can also affect the skin, eyes, ears, heart, and other organs. The underlying complication

associated with this condition includes hearing loss, loss of vision, skin scarring, and deep vein thrombosis.[2,3]

The current treatment for Wegener's granulomatosis is stratified based on the extent and severity of organ damage.[4] Immune suppression forms are the foundation treatment for Wegener's granulomatosis. The use of anabolic-androgenic steroids can even aggravate acute kidney injury, chronic kidney disease, and glomerular toxicity.[7] Dose tapering in steroids is of utmost importance, particularly for patients receiving long-term and high-dose treatments, to avoid serious side effects such as adrenal suppression, osteoporosis, Cushing's syndrome, psychiatric disturbances (agitation, anxiety, irritability, insomnia, restless, ness), and immune suppression. Steroid therapy is an important risk factor for the development of adverse effects, patients receiving less than 40 mg/d are at minimal risk, those receiving 40-80 mg/d are at moderate risk and patients receiving more than 80 mg/d are at high risk for developing steroid-induced psychiatric disturbances. Although reduction or cessation of corticosteroid therapy is the mainstay of treatment for steroid-induced psychiatric reactions.[8,9] For patients with active and non-severe Wegener's granulomatosis who do not have contraindications, methotrexate, and glucocorticoids can induce and maintain remission. For those with severe disease, glucocorticoids are combined either with rituximab or cyclophosphamide. Cyclophosphamide is given for 3 to 6 months and then switched to azathioprine or methotrexate for maintenance of remission. Rituximab is an eminent alternative for patients with relapsing diseases.[5,6] Here, we address a case on Wegener's granulomatosis presented as glomerulonephritis in a South Indian lady.

CASE REPORT

A 60-year-old woman with a known case of type 2 diabetes mellitus, presented with chief complaints of fever, hearing loss (since 2 months), reddish discoloration of eyes (since 6 months), multiple joint pain (since 2 weeks), swelling of lower limbs (since 10 days).

On examination, the patient's body temperature: was 98.6°F, blood pressure was 140/80 mmHg, right arm supine position, and respiratory rate was 18 cycles/min.

On the first admission, the laboratory investigations were done and its reports were as follows, serum. urea: 54 mg/dl, Sr. creatinine: 1.4 mg/dl, serum. uric acid: 4.14 mg/dl, Platelet counts were, 5,51000 cells/mcl, 7, 28000 cells/mcl, 6,86000 cells/ mcl on three consecutive days. C - reactive protein: 210.73 mg/l, and the complement C4 blood test was 40 mg/dl. She had joint pain and considering the possibility of autoimmune diseases and Wegener's disease, ANCA, C-ANCA, HLAB27, ANA, complement C4 blood test, anti-nuclear antibody (ANA) blot perinuclear anti-neutrophil cytoplasmic antibodies (P-ANCA) tests were done and the reports were as follows:

Table 1: Laboratory investigations reports.

S. No	Test	Result
1	**C-ANCA**	Positive
2	**P-ANCA**	Negative
3	**ANA**	Negative
4	**ANA BLOT**	Negative
5	**$HLAB_{27}$**	Positive

Renal biopsy report

Pauci- immune crescentic glomerulonephritis- ANCA associated.

The pattern of injury: necrotizing and crescentic glomerulonephritis.

Berden histological class: Focal.

Additional features: focal global glomerulosclerosis (0.1%), Acute tubular injury (nil), Tubular atrophy, and Interstitial fibrosis (0.1%).

Based on the chief complaints, laboratory investigations, and renal biopsy report, the diagnosis was made as, Wegener's Granulomatosis with small vessel vasculitis, Type 2 diabetes mellitus, and episcleritis.

The patient was advised for immunosuppressive therapy and admitted for rituximab treatment. The patient was treated with rituximab and tolerated it well.

Treatment

Pre-medicate with acetaminophen, an antihistamine with/without a corticosteroid.

The other medications includes, Tablet Wysolone (Prednisolone) 20 mg 3-0-0, Tablet Pantoprazole (40 mg OD), Tablet Furosemide (40 mg ½ tablet, twice daily), Tablet Glimipride (1 mg once daily), and vitamin supplements were prescribed. The patient had high-frequency sensorineural hearing loss for which a hearing aid was provided. Appropriate treatment was provided for proper eye care. Tablet Prednisolone 20 mg 3 tablets mornings (60 mg per day) were continued for the first 5 weeks, then, the dose was tapered to 5 mg per day for 1 week, within 1 week, the patient developed psychiatric disturbances (restlessness, depression, loss of interest, scared of people patenting was referred to psychiatry department and proper treatment were given. After the psychiatric treatment, the patient's cognitive behavioral pattern was found to be improved. Since the patient's kidney status is deprived, a high dose of steroids is not well tolerated, so tablets. Prednisolone 5 mg per day was continued. The patient tolerated well with the current treatment plan and the patient's condition was found to be improving, so the current treatment plan was continued throughout.

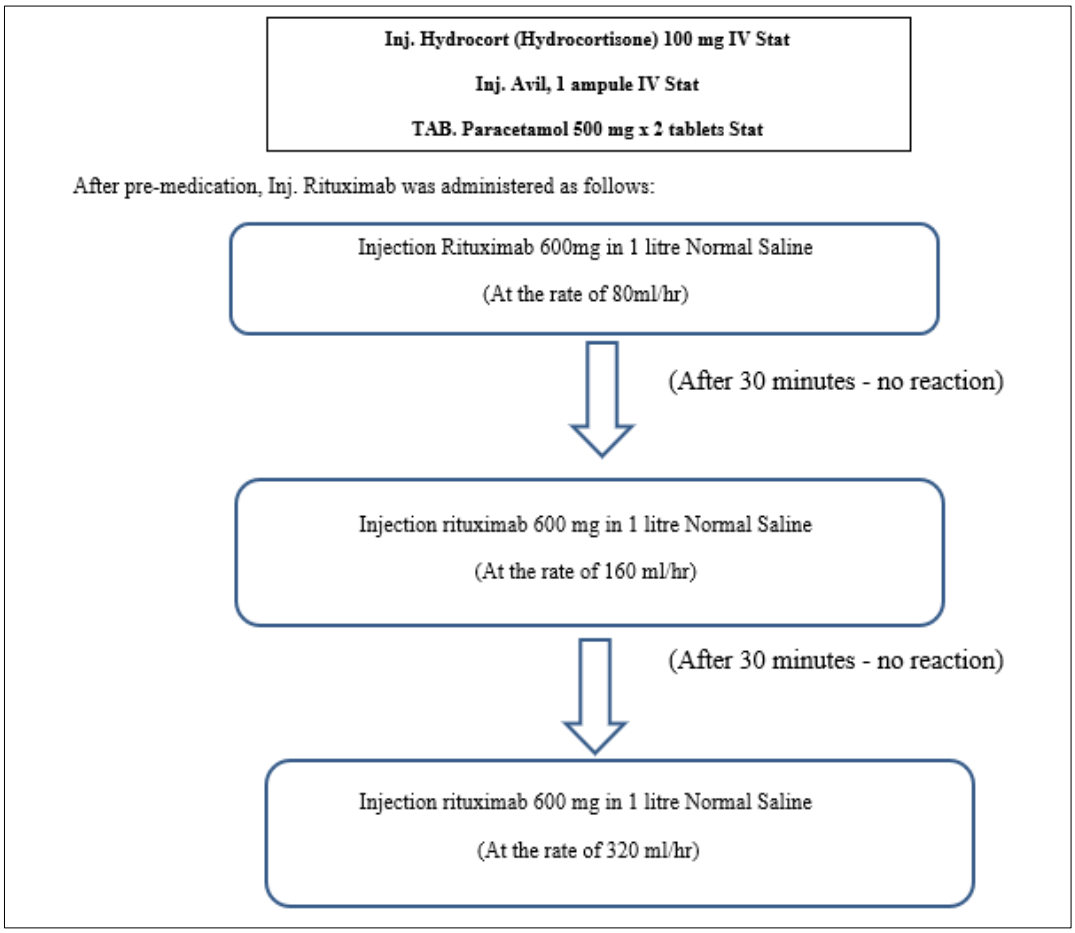

Figure 1: Treatment given.

DISCUSSION

Wegener's granulomatosis is a rare autoimmune disease of unknown etiologies, it is characterized by, granulomatous necrotizing small vessel vasculitis of the upper and lower respiratory tract and glomerulonephritis. Ocular manifestations were present in one-third of the patients.

Apart from that, half of the cases were developed with some neuro-ophthalmic complications. Depending on geographic location, the Wegener's disease is likely to be 8-10 cases per one million.[1] The clinical signs of Wegener's granulomatosis include suppurative otitis, mastoiditis, a saddle nose defect, and hearing loss. About 90% of patients with Wegener's granulomatosis have respiratory tract infections and present with pulmonary symptoms like cough, hemoptysis, dyspnea, and less commonly pleuritic chest pain and tracheal obstruction. The diagnosis of Wegener's granulomatosis is crucial as it is presented with a wide range of clinical symptoms. The available treatment options include glucocorticoids, cyclophosphamide, methotrexate, azathioprine, mycophenolate mofetil, and other cytotoxic and immunosuppressive agents.[10] In this case, the patient was presented with fever, hearing loss, reddish discoloration of the eyes, and the swelling of lower limbs. C-antineutrophil cytoplasmic antibody (C-ANCA), human leukocyte antigen B-27 (HLA-B27) tests showed positive results. The patient has been treated with injection rituximab 600 mg, injection hydrocortisone 100 mg (stat), tablet prednisolone 20 mg (3-0-0), tablet paracetamol, 650 mg (stat), injection pheniramine maleate, 12.5 mg (stat). The patient developed psychiatric symptoms while treatment, it was properly managed. The patient's condition were improved with no other further complications.

CONCLUSION

Wegener's granulomatosis is a rare disease characterized by granulomatous necrotizing of the small vessels. The peak incidence were observed between the age group of 64 to 75 years. Wegener's granulomatosis is presented with a wide range of clinical symptoms, so the diagnosis is very crucial. In Wegener granulomatosis, it is observed that C-ANCA and HLA-B27 showed a positive result. Also, necrotizing and crescentic glomerulonephritis was observed in renal biopsy. The treatment was done with monoclonal antibodies and corticosteroids. The patient's conditions were improving without any severe complications.

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REFERENCES

1. Kubaisi B, Samra KA, Foster CS. Granulomatosis with polyangiitis (Wegener's disease): An updated review of ocular disease manifestations. Intractable & rare diseases research. 2016.
2. Gross WL, Trabandt A, Csernok E. Pathogenesis of Wegener's granulomatosis. InAnnales de Medecine Interne 1998;149(5):280-6.
3. Watts RA, Scott DG. L32. ANCA vasculitis over the world. What do we learn from country differences?. Presse medicale (Paris, France: 1983). 2013;42(4):591-3.
4. Regan MJ, Hellmann DB, Stone JH. Treatment of Wegener's Granulomatosis. Rheumatic Disease Clinics. 2001;27(4):863-86.
5. Singer O, McCune WJ. Update on maintenance therapy for granulomatosis with polyangiitis and microscopic polyangiitis. Current opinion in rheumatology. 2017;29(3):248-53.
6. Langford CA. Update on the treatment of granulomatosis with polyangiitis (Wegener's). Current treatment options in cardiovascular medicine. 2012;14(2):164-76.
7. Davani-Davari D, Karimzadeh I, Khalili H. The potential effects of anabolic-androgenic steroids and growth hormone as commonly used sport supplements on the kidney: a systematic review. BMC nephrology. 2019;20(1):1-2.
8. Liu D, Ahmet A, Ward L, Krishnamoorthy P, Mandelcorn ED, Leigh R et al. A practical guide to the monitoring and management of the complications of systemic corticosteroid therapy. Allergy, Asthma & Clinical Immunology. 2013;9(1):1-25.
9. Warrington TP, Bostwick JM. Psychiatric adverse effects of corticosteroids. InMayo Clinic Proceedings. 2006;81(10):1361-7.
10. Ramsey MK, Owens D. Wegener's Granulomatosis: A review of the clinical implications, diagnosis, and treatment. Laboratory Medicine. 2006;37(2):114-6.

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