World Journal of Biology Pharmacy and Health Sciences
eISSN: 2582-5542
Cross Ref DOI: 10.30574/wjbphs
Journal homepage: https://wjbphs.com/

(RESEARCH ARTICLE)

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Clinical presentation and favored medicines of GPA in tertiary care centers of central Kerala

Sneha Elisa Bency [1,*], Sunil Antony [2], Saranya R K [2] and Shibin Biju [1]

[1] *Department of Pharmacy Practice, Nazareth College of Pharmacy, Othera, Thiruvalla, Kerala, India.*
[2] *Department of General Medicine, Believers Church Medical College Hospital, Thiruvalla, Kerala, India.*

World Journal of Biology Pharmacy and Health Sciences, 2026, 27(01), 194–198

Publication history: Received on 13 June 2026; revised on 25 July 2026; accepted on 28 July 2026

Article DOI: https://doi.org/10.30574/wjbphs.2026.27.1.0409

Abstract

Granulomatosis with polyangiitis is a systemic disease occurs as result of inflammation of the respiratory tract and necrotizing vasculitis of small and medium vessels. This case series illustrates the clinical symptoms and favored medications used to treat patients with granulomatosis and polyangiitis in a central Kerala tertiary care setting. The information for this case series were gathered from the inpatient department of a tertiary care teaching hospital between 2022 and 2025. Certain diagnostic procedures were also performed ;it includes blood counts, radio imaging, inflammatory markers PANCA.

Keywords: Granulomatosis with polyangiitis, Anti-neutrophil Cytoplasmic Antibody (ANCA)-Associated Vasculitis, Proteinase, Myeloperoxidase, Inj. Rituximab, Cough with expectoration.

1. Introduction

Granulomatosis with polyangiitis (GPA; formerly called as Wegener's granulomatosis) is an uncommon necrotizing vasculitis that combines extravascular and perivascular granulomatosis with vascular wall inflammation [1]. The current nomenclature classification places it in the same group as anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV). Granulomatosis with polyangiitis occurs in 5–10 cases per million people annually, with the same prevalence in males and females. Granulomatosis with polyangiitis (GPA) was quite uncommon in young adults and children. The seventh decade of life, between the ages of 65 and 70, is when the stated peak incidence of Granulomatosis with polyangiitis occurs [2]. The pathophysiology of Granulomatosis with polyangiitis is intricate and multifaceted. When paired with environmental triggers like viruses or Staphylococcus aureus infections, genetic variables like Human Leukocyte Antigen -DP Beta1 (HLA-DPB1) variations increase vulnerability. These impair immunological tolerance by encouraging the formation of autoantibodies against neutrophil proteins such as (Proteinase) PR3 or (Myeloperoxidase) MPO. The loss of immunological T and B cell tolerance to neutrophilic proteins, specifically (Proteinase) PR3 or (Myeloperoxidase) MPO, is the pathogenic characteristic. Neutrophils are activated by ANCA binding, which results in neutrophil extracellular traps (NETs), endothelium adhesion, degranulation, and the generation of reactive oxygen species, all of which prolong inflammation [3]. Tissue damage is exacerbated by poor NET clearance brought on by reduced DNAse I activity. Risk variables include age, genetic background, environmental factors, inflammation, and infection coexist with this process. Tracheo- or bronchomalacia can occasionally result from Granulomatosis with polyangiitis (GPA) affecting the airways [4]. This condition could be brought by stenosis or cartilage involvement. The trachea or bronchi are at risk of dynamic collapse during expiration or inspiration due to tracheomalacia or bronchomalacia. Patients may had vague symptoms such as coughing, hemoptysis, or dyspnea. Additionally, because of inadequate secretion clearance, patients experience recurring infections and bronchiectasis [4].

* Corresponding author: Sneha Elisa Bency. elisabency.2@gmail.com

Aim

The purpose of this study was to determine the preferred medications for Granulomatosis with polyangiitis, as well as to evaluate the clinical presentation of Granulomatosis with polyangiitis in tertiary care in central Kerala.

2. Materials and methodology

This case series have been collected from inpatient department between the years 2022 and 2025 at a tertiary care teaching hospital For the final study, only cases with suitable and reliable evidence of clinical presentation and favored medications for granulomatous with polyangiitis were taken into account. As a result, three cases that satisfied the criteria were selected for this case series. Every participant gave their informed consent in a language they could understand, ensuring ethical compliance understanding by patients.

2.1. Case series

2.1.1. CASE 1

A 56 year old female patient with no prior co morbidities presented with complaints of asymmetric pain with swelling bilateral small and large joints associated with early morning stiffness for 3-4 hours, gradual onset of progressive shortness of breath, not associated illness (fever, cough, palpitation or hemoptysis). Also complaints of stabbing type chest pain increased on inspiration. Her chest Xray showed blunting of left costophrenic angle. Her HRCT revealed multifocal, consolidated opacities involving the right middle lobe and left lower lobe with predominantly peribronchial thickening, subtle ill defined centrilobular interstitial pattern and was treated with inj piptaz 4. 5 gram for 3 days. Her 2D Echo showed grade 1 LV dysfunction. She was started on immunosuppression with wysolone and antibiotic cover. Her P-ANCA results came out to be positive. She was diagnosed with granulomatosis with polyangiitis and was decided to start Inj. Rituximab 1000 mg, along with antitubercular therapy, (T. Rifampicin 450 mg +Isoniazide 300 mg +pyrazinamide 1500 mg +ethambutol 800 mg), T. Septran DS, steroids in tapering dose(T. Omnacortil 60 mg).

2.1.2. CASE 2

A 60 year old female patient presented with complaints of cough with expectoration since one month following which she had taken medicines. She also had complaints of fever since two weeks. It was acute in onset aggravated during night associated with chills. She had past history of systemic hypertension. Her routine investigations were done. Hb was mildly reduced, CRP elevated, potassium level mildly reduced, ESR elevated, iron studies showed mildly reduced total iron, TIBC, TSAT. Her HRCT was done and showed ill -defined areas of ground glass opacities with smooth inter and intra lobular septal thickening involving left upper and lower lobes. Left pleural effusion, fibro atelectasis with air bronchograms and volume loss involving right middle lobe with compensatory hyperinflation of right upper and lower lobes. Bronchoscopy was also performed it revealed thin frothy secretion all segments in bronchial tree. Her ANCA testing was performed. Following pulmonology consultation was sought as she was diagnosed with granulomatosis polyangiitis. She was advised to take Inj. Rituximab 1 g once in six month. During hospital stay, she was managed with intravenous antibiotics (InJ. Levofloxacin), antipyretics, antihypertensives, fluids and other supportive measures

2.1.3. CASE 3

A 79 year old female patient was admitted under rheumatology department with granulomatosis with polyangiitis, osteoporosis. Presented with complaints of cough with expectoration for 1 month, low grade fever and worsening pedal edema. She had medical history of systemic hypertension, hypothyroidism, dyslipidemia, coronary artery disease. A HRCT was done showed multiple patchy consolidations with adjacent interstitial septal thickening in bilateral lung fields predominantly in peripheral subpleural distribution, Multiple centrilobular nodules, few enlarged mediastinal lymph nodes and inflammatory markers were elevated ESR 90, CRP 81. Thus she was admitted for further therapeutic management. She initially visited with biopsy proven leukocytoclastic vasculitis, ANA positivity. She had a flare of skin lesions while tapering steroids, then PANCA positivity also contributed to the diagnosis of small vessel vasculitis. She was started with Inj Rituximab 1 gram following. Her BAL study showed normal cytology but had significant growth of Pseudomonas putida following which Inj. Meropenem 1gram thrice daily was given. Patient was continued on mupirocin ointment, and Bactrim ds on alternative days to standard remission maintenance. Inj. Dexamthasone 4mg, T. Calcium Carbonate, T. Nexpro RD, T. Nebivolol, T. Cilnidipine, T. Thyronorm, T. Rosuvastain were also given. Following immunotherapy patient had a good clearance of x ray findings, cough and sputum production.

2.1.4. CASE 4

32 year old male patient admitted with complaints of joint pain and lower limb swelling. It was acute in onset, asymmetric involving both large and small joints. He had been taking anti-inflammatory medications. He noticed the appearance of raised, non itching macular rashes, initially seen over the lower limbs with asymmetric distribution. Then he develops shortness of breath, intermittent cough which was persistent and progressive in nature. He had history TB-Lymph node at 6 months of age. Routine blood investigations was done. Haemoglobin drop was noted. Total count was raised, increased inflammatory markers were present. Renal function test and Liver function test were deranged. Peripheral smear showed moderate anemia-Normocytic normochromic blood picture and neutrophilic leukocytosis with moderate eosinophilia. Due to Hemoglobin drop, one unit of PRBC transfusion was done. He was diagnosed with leukocytoclastic vasculitis. Patient was started on Intravenous antibiotics. His C-ANCA was done and turned out to be positive. Due to the persistent joint pain and was advised to send complements, antiphospholipid antibodies, S. Galctomannan, viral markers and to start patient on pulse methyl prednisolone therapy. Antibiotics were stepped down and patient was given Inj. FCM

500 mg for the replenishment of iron stores. Inj. Rituximab was given due to the persistent symptoms. His urea and creatinine level were raising following SLED was carried out and patient developed acute onset of breathlessness and desaturation subsequently he was intubated. Chest x ray showed bilateral infiltrates. Total counts showed raising trend. Ultrasound of bilateral lower limbs were performed and showed Deep Vein Thrombosis of left CFV and left calf veins. Patient underwent Mechanical Thrombectomy in view of thrombus formation. RDP was also transfused as he develops thrombocytopenia. Post procedure patient was started on Noradrenaline, one unit of PRBC transfusion was done, two units of albumin was transfused in view of low albumin level. ET Mini BAL Culture of patient was sent and showed Klebsiella pneumonia. Patient had persistent fever spikes. His antibiotics were continued. He shows good tolerance to medications. Following Hemodialysis was done on alternative days. During dialysis he developed tachycardia and tachypnea and was started on NIV tapered to oxygen two liters. During dialysis one unit of PRBC transfusion was done in view of low hemoglobin level. He was started on Inj. Ceftazidime avibactam 1. 2 gm once daily, Inj. Aztreonam 2 gram once daily. Following HRCT was performed and showed multifocal predominantly central/peri-broncho vascular air space opacities and tiny nodules in bilateral lungs. Trace possible left pleural effusion. Sub centimetric mediastinal lymph nodes. Rheumatology consultation was done to decide to start Inj. Cyclophosphamide and advised to do urine culture and sensitivity which showed normal flora. He was also started on Inj. Heparin but it stopped due to bleeding from right side of thigh. Subsequently bilateral lower limb venous doppler was done showed no signs of DVT. Inj. Cyclophosphamide 450 mg in 500 ml NS was started. He developed generalized tonic clonic seizures which subsided by itself within two minutes and was started on Inj. Levipil, Nitroglycerin infusion and other hypertensive measures. CT Brain was done showed focal hypodensity in right parieto-occipital-temporal region and right frontal region, which represent acute infarcts. Nitroglycerin infusion was stopped when patient condition became stable. He continues Inj. Cyclophophamide. Following his condition gets improved.

Case	**Case 1**	**Case 2**	**Case 3**	**Case 4**
AGE/ GENDER	60 year, Female	56 year, Female	79 year, Female	32 year, Male
SYMPTOMS	Cough with expectoration, fever	Pain with swelling of joints, shortness of breath, stabbing type chest pain	Cough with expectoration, low grade fever, worsening pedal edema	Joint pain and lower limb swelling. shortness of breath, intermittent cough
PAST HISTORY	Systemic Hypertension, Iron Deficiency Anemia, Hypokalemia, Prediabetes.	No comorbidities	Hypertension, Hypothyroidism, Dyslipidemia, Coronary Artery Disease.	TB-Lymph node
MANAGEMENT	Inj. Rituximab 1g	Inj. Rituximab 1 g	Inj. Rituximab 1g	Inj. Rituximab 1g, Inj. Cyclophosphamide 450 mg

3. Discussion

In this study we examined clinical presentation and therapeutic management of granulomatosis with polyangiitis. Common symptoms include sinusitis, otitis media, cough, chest pain, hemoptysis, joint pain, hearing loss, nasal bone deformation, dyspnoea etc. The diagnosis of GPA may be delayed for a number of reasons.

Constitutional symptoms and joint complaints were often reported in addition to specific findings for the afflicted organ in GPA. It may happen weeks or months prior to organ involvement. According to Abdou et al. , 46% of patients received a diagnosis one to six months after the onset of symptoms, compared to just 22% who had a diagnosis within the first month. They revealed that 15% of patients had a diagnosis delay between six and twelve months, while 18% had a delay longer than twelve months. [4]

A combination of clinical observations, imaging, laboratory results (erythrocyte sedimentation rate, c-reactive protein, complete blood count, renal and urine parameters), serological markers (ANCA testing), and histological findings when biopsy can be done ; were used to diagnose GPA [5].

Immunosuppressive medications were used in a variety of combinations as part of the therapeutic management of GPA. It can be divided into two stages: the maintenance phase and the induction phase. Cyclophosphamide, glucocorticoids, azathioprine, rituximab, methotrexate, and, if necessary, plasmapheresis were frequently utilized medications [6.]

Abbreviations

- GPA: Granulomatosis with polyangiitis
- ANCA: Anti-neutrophil Cytoplasmic Antibody
- HLA-DPB1: Human Leukocyte Antigen -DP Beta1
- MPO: Myeloperoxidase
- HRCT: High Resolution Computed Tomography
- DVT: Deep vein thrombosis
- ET-MINI-BAL: Endotracheal Mini-Bronchoalveolar Lavage
- PRBC**:** Packed Red Blood Cells
- SLED Sustained Low-Efficiency Dialysis
- Inj. FCM**:** Injection Ferric Carboxymaltose
- CFV: Common Femoral Vein
- RDP: Random Donor Platelets
- ESR: Erythrocyte Sedimentation Rate
- CRP: C-Reactive Protein
- NIV: Non-invasive ventilation
- HRCT: High-Resolution Computed Tomography
- TSAT: Transferrin Saturation
- TIBC: Total Iron-Binding Capacity

4. Conclusion

Patients with GPA who were admitted to this examination showed symptoms like low grade fever, shortness of breath, cough and pedal edema. Among these patients, injection rituximab was the preferred medication for treating GPA. The tests utilized to diagnose GPA included biopsy, PANCA, radiographic imaging, blood counts, and inflammatory markers.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

The authors certify that they have obtained consent from the participants in this study and their details will be concealed with due effort.

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