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Case Report

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Challenging Management of Granulomatosis with Polyangiitis in a Patient with Multiorgan Involvement and Persistent Disease Activity Despite CD-19 Depletion

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Abstract

Background: Odontogenic infections are treated with an extended spectrum penicillin especially ampicillin. But, these pathogens acquires resistance due to wide spread usage, remains as a major challenge with the newer generation of antibiotics. So, natural products, from the ancient times, used to cure various ailments and emerges as alternatives to prevent the development of resistant species.

Aim: To evaluate the antimicrobial efficacy of neem, aloe vera and turmeric on *Streptococcus pyogenes(S.pyogenes)*, *Streptococcus oralis(S. oralis)* and *Staphylococcus aureus(S.aureus)*.

Methodology: Soxhlet extraction of *Azadirachta indica* (neem), *Curcuma longa* (turmeric) and direct extraction of *Aloe barbadensis* (aloe vera) aloe vera was done from the leaves. Petri plates containing 20 ml nutrient agar medium with 24 hr culture of *S. aureus*, *S. pyogenes* and *S. oralis* were subjected to concentrations of Neem, Turmeric and Aloe vera hydroalcoholic extracts (1000 µg/ml, 500 µg/ml, 200 µg/ml, and 100 µg/ml) and Ampicillin (positive control). These plates were incubated at 37°C for 24 hours. Antibacterial activity was assayed by measuring the diameter of the inhibition zone. The values were calculated using Graph Pad Prism 6.0 software.

Results: Ampicillin showed the maximum zone of inhibition on *S.pyogenes* and *S. oralis*, whereas *Neem* at 1000 ug/ml concentration showed a greater zone of inhibition against *S.pyogenes* and *S. aureus*, and *Aloe vera* showed maximum zone of inhibition on S. oralis.

Keywords: Granulomatosis with Polyangiitis (GPA), Rituximab, Iatrogenic Hypogammaglobulinemia, Antineutrophil Cytoplasmic Antibody (ANCA), Intravenous Immunoglobulin (IVIG)

Introduction

Immunity is crucial for maintaining homeostasis and safeguarding the body against pathogens; however, dysfunction within the immune system can lead to severe health consequences, potentially resulting in life-threatening conditions. These dysfunctions are frequently categorized as autoimmune disorders [1]. which can emerge from a variety of factors affecting different organs. When autoimmune processes involve blood vessels, the condition is termed vasculitis [2]. A significant subset of vasculitis includes conditions associated with anti-neutrophil cytoplasmic antibodies (ANCA), such as Wegener's granulomatosis, now recognized as granulomatosis with

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polyangiitis (GPA) [3]. The estimated annual worldwide incidence of GPA ranges from 10 to 20 cases per million individuals, with geographical variations noted [4]. GPA primarily affects small to medium-sized vessels [5]. It is characterized by a triad of symptoms: upper respiratory tract involvement, pulmonary nodules or hemorrhage, and glomerulonephritis [6]. However, the spectrum of symptoms associated with GPA is extensive; not all patients will exhibit the classic triad, and variations in symptoms and organ involvement are common. Patient present with seasonal variations sometimes [7]. While various diagnostic protocols and tests are employed to assess GPA and monitor disease progression, results can be inconsistent. Management strategies for GPA aim to control the disease and prevent progression [8]. Management is there yet treatment responses can vary significantly among individuals. Some patients may present with rare symptoms or unique responses to therapy highlighting the need for careful documentation and individualized treatment plans to optimize patient care [9].

Case Presentation

A 41-year-old female with allergies to latex, penicillin, sulfa drugs, sulfamethoxazole, and trimethoprim (TABLE 1). She is a non-smoker with a significant medical history that includes longstanding granulomatosis with polyangiitis (GPA) diagnosed at age 13. Her GPA has been severe, characterized by multiple organ involvement, including sinusitis with saddle nose deformity, pulmonary nodules, hearing loss, scleritis/episcleritis, nephritis, gingival hypertrophy, possible cerebrovascular accident (CVA), neuropathy, and cutaneous manifestations. Additionally, she has untreated atrial fibrillation and coronary artery disease, along with iatrogenic hypogammaglobulinemia likely secondary to previous rituximab infusions. At the time of evaluation, she was on linaclotide 72 micrograms capsule, oxycodone and acetaminophen 7.5 mg-325 mg, duloxetine 30 mg capsule, budesonide and formoterol 160 micrograms-4.5 micrograms per actuation aerosol inhaler, mycophenolate mofetil 500 mg tablet, prednisone 20 mg tablet, ergocalciferol 1,250 micrograms (50,000 units) capsule, atovaquone 750 mg per 5 mL oral suspension, cyclophosphamide 500 mg/mL intravenous solution, and mesne 100 mg/mL intravenous solution (Table 2).

Table 1: Allergies

INGREDIENT	REACTION (SEVERITY)
LATEX	Pruritic rash
PENICILLIN	Pruritic rash
SULA (SULFONAMIDE ANTIBIOTICS)	Pruritic rash
SULFAMETHOXAZOLE	Pruritic rash
TRIMETHOPRIM	Pruritic rash

At her first visit in November 2023, she had been off all immunosuppressive therapy for an undetermined duration. Previously, she received steroids, cyclophosphamide (two doses), and rituximab, which led to hypogammaglobulinemia requiring IV immunoglobulin (IVIG) treatment every four weeks. During this visit, laboratory studies were inconclusive regarding ANCA disease activity, and the primary concern was an infection due to a left lower lobe (LLL) cavitary lung lesion identified on a CT scan in July 2023. A biopsy of this lesion revealed scant benign bronchial mucosa, negative for granuloma or malignancy, although respiratory cultures grew Staphylococcus aureus, for which she was treated with antibiotics. In August 2023, she underwent sinus surgery, which noted purulent material and resulted in an unrevealing biopsy of a sinus polyp. Following this, she experienced significant headaches, leading to multiple emergency department visits, where she was given intermittent steroids. By late December 2023, an ophthalmology evaluation revealed isolated sixth nerve palsy, prompting plans for MRI of the brain and further neurology evaluation. However, she deteriorated and was hospitalized in January 2024, where worsening cavitary lung lesions were noted, and MRI indicated significant changes in the sinuses.

During this hospitalization, she received intravenous antibiotics, and a pigtail catheter was placed due to the lung lesions, but cultures were negative. A positive Streptococcus pneumoniae antigen led to a switch in antibiotics to IV Rocephin. She was discharged on February 8, 2024, on oral steroids and IV antibiotics, but returned to the emergency department five days later with new skin lesions, diagnosed as leukocytoclastic vasculitis, and continued progression of her lung lesions. A bronchoscopy revealed diffusely inflamed mucosa and old bloody secretions, but cultures were negative. High-dose steroids were initiated, and upon consultation, I recommended transfer for inpatient rheumatology care and resuming her IVIG therapy. She was given pulse-dose steroids and transitioned to oral prednisone. Unfortunately, she did not follow up as planned due to difficulties related to her leg wounds and a lack of primary care.She returned to the hospital on March 14, 2024, with worsening leg wounds, which were cultured and found to grow Staphylococcus aureus. Imaging showed evolving cavitary lung lesions, prompting another recommendation for higher-level care. After surgical debridement, she was discharged on March 21, 2024, on oral steroids, IV Rocephin, and prophylaxis for Pneumocystis jirovecii pneumonia (PJP). Following readmissions in April for hypotension and further infections, she developed osteomyelitis of the right fifth metatarsal, leading to a transition to IV Meropenem. As her condition progressed, she was evaluated for her eye disease, which continued to worsen. I recommended further immunosuppressive therapy, leading to the administration of rituximab starting in May 2024.

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Table 2: Medication (Prior to Last Visit)

MEDICATIONS	DOSE	FORM	FREQUENCY	START DATE
Linzess (Linaclotide)	72 mcg	capsule	One every day on an empty stomach at least 30 minutes before 1st meal of the day	03-07-2024
Percocet (Oxycodone/paracetamol)	7.5 mg-325 mg	Tablet	One tablet every 6 hours as needed	03-07-2024
Cymbalta (Duloxetine) delayed release	30 mg	capsule	Once per day	03-07-2024
Symbicort (Budesonide / Formoterol)	160 mcg-4,5 mcg/ actuation HFA	Aerosol inhaler	inhale 2 puff by inhalation, 2 times every day in the morning and evening as needed	03-07-2024
Mycophenolate mofetil	500 mg	Tablets	2 tablets twice daily for 07/03/2024 2 weeks then increase to 3 tablets twice daily	03-07-2024
Prednisone	20 mg	Tablet	4 tablets daily	
Vitamin D2 1,250 mcg	50,000 unit	Capsule	One per week	03-07-2024
Atovaquone	750 mg/5 mL	oral suspension	10 milliliter once every day with meal	03-07-2024
Cyclophosphamide	500 mg/ml	Intravenous solution	Infuse 500mg every 2 weeks for 6 doses	08-07-2024
Mesna (sodium 2-mercapto ethane sulfonate)	100mg/mL	Intravenous solution	Infuse 100mg before and after cyclophosphamide infusion	08-07-2024

mcg (microgram): It is one-millionth of a gram, used for measuring tiny amounts in pharmaceuticals and supplements.
mg (milligram): A milligram is one-thousandth of a gram, commonly used for drug dosages and nutritional content.
mL (milliliter): A milliliter is one-thousandth of a liter, used to measure small volumes of liquids in various fields.
HFA (hydrofluoroalkane): Hydrofluoroalkanes are eco-friendlier propellants used in inhalers and aerosol products.

At her follow-up visit in July 2024, lab studies indicated elevated CRP/ESR, positive C-ANCA and PR3, and new proteinuria, prompting the initiation of cyclophosphamide while continuing steroids and discontinuing mycophenolate mofetil (see Table 3). On July 17, 2024, the patient presented with increased fatigue, prolonged sleep, low mood, and significant functional limitations. She reported difficulty kneeling and challenges with activities of daily living, stating she could only walk approximately 10 blocks. Additionally, she experienced obstacles with climbing stairs and performing routine tasks, such as putting on socks and shoes. Vital signs revealed elevated blood pressure (see Table 4), and her pain level was assessed as 7 out of 10 using the numeric pain intensity scale. Systemic review indicated symptoms of fatigue, blurred vision, vision loss, and a rash. Physical examination findings included a saddle nose deformity, redness and proptosis of the right eye, and healing skin lesions on the lower extremities, which were covered by clean dressings. Auscultation revealed diffuse wheezing, and the patient ambulated with the assistance of a walker. Notably, there was a deformity at the distal interphalangeal joint of the right second digit. Laboratory tests performed on the same day included an antineutrophil cytoplasmic antibody (ANCA) screen, which reflexed positive to myeloperoxidase (MPO) and proteinase 3 (PR-3) antibodies; a qualitative beta-hCG; C-reactive protein (CRP); complete blood count (CBC) with platelet and differential; comprehensive metabolic panel (CMP); erythrocyte sedimentation rate (ESR); protein/creatinine ratio from random urine; and microscopic urinalysis (see table 5). The patient's medication regimen was adjusted to include intravenous cyclophosphamide 500 mg administered once, with pre-infusion medications comprising acetaminophen 1000 mg orally, diphenhydramine 25 mg intravenously, Mesna 100 mg intravenously pre-infusion and as needed, and methylprednisolone (Solumedrol) 80 mg intravenously pre-infusion. Ondansetron 8 mg was also prescribed as needed. Mycophenolate mofetil (MMF) was discontinued, and the patient had completed four doses of rituximab at a dosage of 375 mg/m². Despite the depletion of CD19 count, disease progression continued, necessitating the administration of intravenous cyclophosphamide (table 6).

Table 3: LABs collected on 3/7/24

LABS	RESULTS	NORMAL VALUES	UNIT
Hemoglobin	11.3	11.5-15.5	Gm/dL
Hematocrit	39.9	35.2-46.4	%
Platelet counts	418	137-397	K/cumm
Total leukocytic count	10.9	3.8-11.5	K/uL
Erythrocyte Sedimentation Rate (ESR)	28	<26	mm/hr
Albumin/Globulin ratio	1.9	1.1-2.5	Mg/dl
Albumin	4.1	3.5-5.2	g/dl
Alkaline phosphatase	98	35-121	IU/L
Serum glutamic pyruvic transaminase	14	<5-40	IU/L
Bilirubin, Total	0.3	<0.2-1.2	mg/dL
blood urea nitrogen (BUN)	21	Jun-20	Mg/dl
Calcium	9.7	8.6-10.4	Mg/dl
Chloride	106	97-108	Mmol/L
Carbon dioxide	28	22-32	Mmol/L
Creatinine	0.5	0.5-1	Mg/dl
Glucose	188	65-99	Mg/dl
Potassium	4.5	3.5-5.3	Mmol/L
Protein	6.3	6.0-8.3	g/dl
Sodium	147	135-145	Mmol/L
Creatine kinase	16	20-180	U/L
C-Reactive protein	5.01	<0.5	Mg/dl
HIV 1/2 Ab Screen w/p24Ag	Non-reactive	Non-reactive	
Vitamin-D (25-hydroxy)	9.8	30-100	Ng/dl
Hemoglobin A1C	5.7	>6.5= Diabetic	%
		5.78 – 6.49= Pre-Diabetic	
		<5.7= non-diabetic	
Serum IgG	439	700-1600	Mg/dl
Thyroid stimulating hormone (TSH)	0.24	0.43-5.2	mU/L
Free thyroxine 4 (Ft4)	1.5	0.8-1.7	ng/dL
Complement C3	195	90-180	mg/dL
Complement C4	29	Oct-40	mg/dL
Hepatitis B core antibodies (HBcAb)	Non-reactive	Non-reactive	
Hepatitis B surface antibodies (HBsAb)	113	<10 = non-reactive	
		>10 = reactive	
Hepatitis B surface antigen (HBsAg)	Non-reactive	Non-reactive	
Hepatitis C antibodies IgG	Non-reactive	Non-reactive	
Aldolase	4.4	1.2-7.6	U/L
% CD19	0	Jun-23	%
% CD3	69	62-87	%
% CD4	43	32-64	%
% CD8	26	15-46	%
% Natural killer cells	30	Apr-26	%

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Absolute CD19 Cells	0	91-610	cells/uL
Absolute CD3 Cells	256	570-2400	cells/uL
Absolute CD4 Cells	157	430-1800	cells/uL
Absolute CD8 Cells	94	210-1200	cells/uL
Absolute natural killer Cells	109	78-470	cells/uL
CD4:CD8 ratio	1.6	0.8-3.9	Ratio
c-ANCA	Detected	Not detected	
ANCA titer	01:40		Titer
Myeloperoxidase autoantibodies	0	0-19	AU/mL
Serine proteinase-3 antibodies	191	0-19	AU/mL
Qunatiferon Mitogen minus NIL	6.8		IU/mL
Qunatiferon NIL	0.02		IU/mL
Qunatiferon Plus TB1 minus NIL	0.008	<0.346	IU/mL
Qunatiferon plus TB2 minus NIL	0.009	<0.346	IU/mL
Qunatiferon TB Gold Plus	Negative	Negative	Negative
Creatinine, urine sample	75.7		Mg/dl
Protein, urine sample	78.2		Mg/dl
Protein/Creatinine ratio	1.03		ratio

mg/dL (milligrams per deciliter): A measure of concentration used for substances like glucose and cholesterol in blood.

ng/dL (nanograms per deciliter): A small concentration unit often used for hormones and drugs in serum.

IU/mL (international units per milliliter): A standardized measure for the biological activity of vitamins and hormones.

AU/mL (arbitrary units per milliliter): A relative measurement used in diagnostics based on standard reference samples.

Cells/μL (cells per microliter): A count of cells, such as red or white blood cells, in a microliter of blood.

CD (cluster of differentiation): A classification system for identifying immune cell surface markers.

mU/L (milliunits per liter): A unit for expressing the concentration of active substances like hormones in a liquid.

mmol/L (millimoles per liter): A unit measuring the concentration of solutes in a solution, commonly used for blood metabolites

Table 4: vital signs recorded on 17/7/2024

COMPONENT	PATIENT'S VALUE	NORMAL RANGE
Blood pressure (mm Hg)	140/100 mmHg	120/80
Pulse (pulse per min)	70/min	60-100
Respiratory rate (breathes per min)	18/min	Dec-20
Temperature (F)	97.0 F	97-99
Weight (lb)	157	Variable
Weight (kilogram)	71.2	Variable
Height (foot)	5	Variable
Height (centimeter)	172.2	Variable
Oxygen saturation (%)	97	>92
Body Mass Index (kg/m2)	23.8	18.5-24.9

mmHg (millimeters of mercury): A unit of pressure often used to measure blood pressure and atmospheric pressure.

lb (pound): A unit of weight in the imperial system, commonly used to quantify mass in the United States.

F (Fahrenheit): A temperature scale where water freezes at 32°F and boils at 212°F, primarily used in the U.S.

kg/m² (kilograms per meter squared): A unit of measurement for density or body mass index (BMI), indicating mass per unit area.

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Table 5: LABs collected on 17/7/24

LABS	RESULTS	NORMAL VALUES	UNIT
Hemoglobin	10.8	11.5-15.5	Gm/dL
Hematocrit	38.9	35.2-46.4	%
Platelet counts	416	137-397	K/cumm
Total leukocytic count	14	3.8-11.5	K/uL
Erythrocyte Sedimentation Rate (ESR)	12	<26	mm/hr
Albumin/Globulin ratio	1.9	1.1-2.5	Mg/dl
Albumin	3.9	3.5-5.2	g/dl
Alkaline phosphatase	100	35-121	IU/L
Serum glutamic pyruvic transaminase	11	<5-40	IU/L
Bilirubin, Total	0.2	<0.2-1.2	mg/dL
blood urea nitrogen (BUN)	20	Jun-20	Mg/dl
Calcium	9.8	8.6-10.4	Mg/dl
Chloride	106	97-108	Mmol/L
Carbon dioxide	27	22-32	Mmol/L
Creatinine	0.64	0.5-1	Mg/dl
Glucose	105	65-99	Mg/dl
Potassium	4	3.5-5.3	Mmol/L
Protein	6	6.0-8.3	g/dl
Sodium	144	135-145	Mmol/L
C-Reactive protein	2	<0.5	Mg/dl
c-ANCA	Detected	Not detected	
ANCA titer	01:20		Titer
Myeloperoxidase autoantibodies	0	0-19	AU/mL
Serine proteinase-3 antibodies	132	0-19	AU/mL
Beta-hCG qualitative, urine	Negative	Negative	
Creatinine, urine sample	78		Mg/dl
Protein, urine sample	44.8		Mg/dl
Protein/Creatinine ratio	0.57		ratio
Urine bacteria	1+	None seen	/HPF
Urine Red Blood Cells	03-May	0-2	/HPF
Urine White blood cells	06-Oct	0-5	/HPF

mg/dL (milligrams per deciliter): Measures concentration in blood, commonly for glucose and cholesterol.

K/cumm (thousands per cubic millimeter): Counts cells in blood, expressed in thousands per cubic millimeter.

K/µL (thousands per microliter): Cell concentration measurement, often used in hematology.

IU/mL (international units per milliliter): Standardized measure for biological activity of substances.

AU/mL (arbitrary units per milliliter): Relative unit based on reference standards in diagnostics.

mmol/L (millimoles per liter): Indicates concentration of solutes, used for blood metabolites.

/HPF (per high power field): Indicates the number of cells or particles in a microscopy field.

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Table 6: Medications (Added, Continued or Stop Date)

MEDICATIONS	DOSE	FORM	FREQUENCY	START DATE
Linzess (Linaclotide)	72 mcg	capsule	One every day on an empty stomach at least 30 minutes before 1st meal of the day	03-07-2024
Percocet (Oxycodone/paracetamol)	7.5 mg-325 mg	Tablet	One tablet every 6 hours as needed	03-07-2024
Cymbalta (Duloxetine) delayed release	30 mg	capsule	Once per day	03-07-2024
Symbicort (Budesonide / Formoterol)	160 mcg-4,5 mcg/actuation HFA	Aerosol inhaler	inhale 2 puff by inhalation, 2 times every day in the morning and evening as needed	03-07-2024
Mycophenolate mofetil	500 mg	Tablets	2 tablets twice daily for 2 weeks then increase to 3 tablets twice daily	03-07-2024
Prednisone	20 mg	Tablet	4 tablets daily	
Vitamin D2 1,250 mcg	50,000 unit	Capsule	One per week	03-07-2024
Atovaquone	750 mg/5 mL	oral suspension	10 milliliter once every day with meal	03-07-2024
Cyclophosphamide	500 mg/ml	Intravenous solution	Infuse 500mg every 2 weeks for 6 doses	08-07-2024
Mesna	100mg/mL	Intravenous solution	Infuse 100mg before and after cyclophosphamide infusion	08-07-2024
Lisinopril (angiotensin-converting enzyme inhibitors)	20mg	Tablet	Once per day	17-07-2024
Tavneos (Avacopan)	10mg	capsule	One per week	17/72024

Table 7: Condition Descriptions

CONDITIONS	ONSET DATE
Wegener's granulomatosis with renal impairment	12-04-2021
Immunodeficiency, unspecified	15-11-2023
Age-related osteoporosis without pathological fracture	15-11-2023
Age-related osteoporosis without pathological fracture	29-11-2023

A CBC was planned for the next 10-14 days, and the patient continued on prednisone 60 mg daily, with plans for a dose reduction in 1-2 weeks. Her last immunoglobulin G (IgG) level was recorded at 439 on July 2, 2024, and she was rescheduled for intravenous immunoglobulin (IVIG) infusions as soon as possible, noting her high risk of steroid-related complications. The treatment for glucocorticoid-induced osteoporosis was also considered for the future. To manage hypertension, lisinopril 20 mg daily was added to her medication regimen,with advice for home blood pressure monitoring. A follow-up appointment was scheduled in four weeks, and the patient was counseled regarding the potential development of Pneumocystis jirovecii pneumonia (PCP). Patient condition is stated in Table 7.

Discussion

This case underscores the complexities involved in managing granulomatosis with polyangiitis (GPA), particularly in a patient exhibiting extensive multi-organ involvement and significant comorbid conditions. GPA, which is classified as ANCA-associated vasculitis, presents a diverse array of clinical symptoms, as demonstrated by this patient's severe manifestations across multiple systems, including respiratory, renal, and ocular [10]. The patient's long-standing diagnosis of GPA, established at an early age, highlights the critical need for early detection and ongoing management of this chronic illness. Despite prior treatments, including cyclophosphamide and rituximab, the recent pause in her immunosuppressive therapy may have played a role in exacerbating her disease and contributing to infection risks, complicating her clinical picture[11][12]. The persistent cavitary lung lesions and subsequent bacterial infections

illustrate the inherent dangers of both the condition and the therapies employed.

Additionally, the presence of untreated comorbidities, such as atrial fibrillation and coronary artery disease, introduces further complications and risks in managing GPA. The patient's hypogammaglobulinemia, likely a consequence of earlier rituximab treatment, raises concerns regarding her immune function, increasing her vulnerability to infections and necessitating regular intravenous immunoglobulin (IVIG) therapy. This scenario emphasizes the importance of a multidisciplinary approach when addressing the needs of patients with complex autoimmune disorders. The diverse symptomatology observed in GPA patients can lead to diagnostic hurdles and delays in appropriate treatment. In this instance, the patient experienced significant neurological symptoms, including isolated sixth cranial nerve palsy, necessitating further imaging studies. Such neurological issues can complicate GPA management and require immediate attention. The treatment strategy implemented for this patient included high-dose corticosteroids and intravenous antibiotics, reflecting the need for proactive management of acute exacerbations. However, the variability in treatment responses and the emergence of leukocytoclastic vasculitis suggest that individual patient responses can differ greatly. This highlights the importance of continuous monitoring and the adjustment of therapeutic approaches based on the patient's changing clinical condition. Furthermore, the patient's psychosocial issues, including fatigue and limitations in daily functioning, are noteworthy and should be integrated into the overall treatment strategy. Supportive measures, such as counseling and rehabilitation, can be vital in improving the patient's quality of life and functional abilities.

Conclusion

This case illustrates the multifaceted nature of managing GPA, emphasizing the importance of a tailored approach that addresses both the disease and its complications. Continuous monitoring, interdisciplinary collaboration, and individualized treatment plans are essential to optimize outcomes for patients with GPA and similar autoimmune conditions. Future research should focus on identifying biomarkers for disease activity and response to therapy, which could further refine management strategies and improve prognostic accuracy.

Patient Consent

A written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest

The authors declare that they have no conflicts of interest.

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