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(CASE REPORT)

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Granulomatosis with Polyangiitis Presenting as Aortic Valve Incompetence: Case Report and a Brief Review of the Literature

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Abstract

Granulomatosis with polyangiitis (GPA) is an antineutrophil cytoplasmic antibody (ANCA)–associated small-vessel vasculitis that predominantly affects the respiratory tract and kidneys, with cardiac involvement uncommon and often underrecognized. When present, cardiac manifestations are associated with increased morbidity and mortality and may include myocarditis, pericarditis, conduction abnormalities, coronary arteritis, and intracardiac masses. The nonspecific nature of cardiac symptoms often delays diagnosis, underscoring the importance of heightened clinical suspicion.

We report a case of GPA complicated by clinically significant cardiac involvement, presenting with progressive cardiopulmonary symptoms and electrocardiographic abnormalities. Laboratory evaluation demonstrated antineutrophil cytoplasmic antibody positivity consistent with GPA, and multimodality cardiac imaging revealed myocardial involvement. Definitive diagnosis was established through histopathologic confirmation of necrotizing granulomatous inflammation, supporting vasculitic cardiac involvement. Cardiac magnetic resonance imaging played a critical role in delineating the extent of myocardial disease, consistent with prior studies demonstrating its utility in detecting overt and subclinical cardiac manifestations of ANCA-associated vasculitis.

The patient was treated with immunosuppressive therapy in accordance with established vasculitis management strategies, resulting in clinical stabilization. This case highlights the diagnostic challenge posed by cardiac GPA and reinforces the importance of integrating advanced cardiac imaging with serologic and tissue-based diagnosis to guide timely therapy. Increased awareness of cardiac involvement in GPA is essential to facilitate early recognition, individualized treatment, and prevention of irreversible cardiac injury.

Keywords: Granulomatosis with polyangiitis; Wegener granulomatosis; Antineutrophil cytoplasmic antibody; Transthoracic echocardiography

1. Introduction

Granulomatosis with polyangiitis (GPA) is a rare systemic necrotizing vasculitis characterized by granulomatous inflammation involving small- to medium-sized vessels. It typically affects the respiratory tract and kidneys, with presentations such as chronic sinonasal disease, pulmonary nodules, and even rapidly progressive glomerulonephritis.

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[1,2] While ANCA, specifically proteinase 3-ANCA (PR3-ANCA), supports the diagnosis, GPA may present with a wide array of symptoms, including atypical manifestations, contributing to diagnostic delay. [3]

Cardiac involvement in GPA is uncommon, often subclinical, yet has important prognostic implications. Reported cardiac manifestations include coronary arteritis, conduction system abnormalities, myocarditis, and pericarditis. [4] In GPA, valvular involvement is atypical and rare, with aortic valve disease being especially uncommon and an underreported manifestation. Aortic valve incompetence in GPA may be caused by chronic granulomatous inflammation of the valvular apparatus or the adjacent aortic root. It can clinically resemble more common etiologies, such as congenital valvular disease or infective endocarditis. [5,6] This overlap presents a diagnostic dilemma, specifically in patients without classical systemic vasculitis symptoms at presentation.

The ideal approach to the diagnosis and management of GPA-associated valvular disease remains uncertain due to its rarity and the lack of prospective data. Decisions regarding immunosuppressive therapy, timing of surgical intervention, and long-term outcomes are largely guided by individual case reports and small case series, underscoring a gap in the existing literature. [7] We report this case to highlight GPA presenting as aortic valve incompetence and to emphasize the importance of considering vasculitis in the differential diagnosis of unexplained valvular heart disease.

2. Case Presentation

2.1. Clinical Presentation

A 62-year-old woman presented to the emergency department because she passed out while doing simple housework. She reported a three-month history of progressive dyspnea on exertion and orthopnea. Her fatigue worsened, and her exercise tolerance decreased, from walking several blocks to becoming short of breath after one flight of stairs. Over the preceding six weeks, she developed nocturnal paroxysmal dyspnea and bilateral lower extremity edema. She denied chest pain but noted occasional palpitations.

The patient had initially attributed her symptoms to deconditioning and aging, managing them with rest and over-the-counter diuretics. There was a vague history of intermittent sinus congestion over the past year, which she had self-treated with antihistamines, and one episode of epistaxis three months prior that resolved spontaneously.

2.2. Physical Examination, History, Investigations, and Differential Diagnosis

Physical examination revealed blood pressure of 145/50 mmHg with a widened pulse pressure. Cardiac auscultation identified a grade 3/6 high-pitched, blowing diastolic murmur best heard at the left sternal border, with an Austin Flint murmur at the apex. Bilateral basilar crackles were present, and there was 2+ pitting edema of the lower extremities. No saddle nose deformity or cutaneous lesions were noted. Personal history was significant for hypertension managed with an ACE inhibitor. Family history was noncontributory for valvular or autoimmune diseases.

Laboratory studies showed mild normocytic anemia (hemoglobin 10.2 g/dL), elevated ESR (78 mm/hr), and CRP (45 mg/L). Serum creatinine was mildly elevated to 1.4 mg/dL. Urinalysis demonstrated microscopic hematuria with dysmorphic red blood cells and mild proteinuria. Transthoracic echocardiography (TTF) revealed severe aortic regurgitation with a dilated left ventricle and reduced ejection fraction (40%). The aortic valve appeared thickened with irregular leaflets. Chest radiography showed cardiomegaly and mild pulmonary vascular congestion.

Differential diagnosis included: rheumatic heart disease, infective endocarditis (though blood cultures were negative and no fever was documented), bicuspid aortic valve with secondary degeneration, and connective tissue disease-related valve pathology.

2.3. Multidisciplinary Discussion, Surgical Intervention, and Diagnostic Revelation

The multidisciplinary cardiac team reviewed the case and concluded that the patient required urgent aortic valve replacement given the severity of regurgitation, symptomatic heart failure, and ventricular dysfunction. Despite the elevated inflammatory markers and renal findings, these were attributed to cardiorenal syndrome and chronic heart failure. The absence of fever and negative blood cultures made active endocarditis unlikely.

The patient underwent successful aortic valve replacement with a bioprosthetic valve. Intraoperatively, the native valve demonstrated thickened, friable leaflets with focal areas of destruction but no vegetations. The surgical specimen was sent for comprehensive pathological examination. Histopathological analysis revealed the classic triad of GPA: geographic necrosis, granulomatous inflammation with multinucleated giant cells, and necrotizing vasculitis affecting

small and medium-sized vessels within the valve tissue. There was transmural inflammation with fibrinoid necrosis of the vessel walls. (Figure 1 A, B, C) Special studies did not reveal any microorganisms.

This unexpected finding prompted immediate rheumatologic consultation and additional testing. Serum c-ANCA (proteinase-3 antibodies) returned strongly positive at 185 U/mL. A later review of the chest CT found small nodules in the upper lung lobes that had been missed in the first evaluation. High-resolution CT of the sinuses demonstrated mucosal thickening and bone erosion consistent with chronic granulomatous sinusitis.

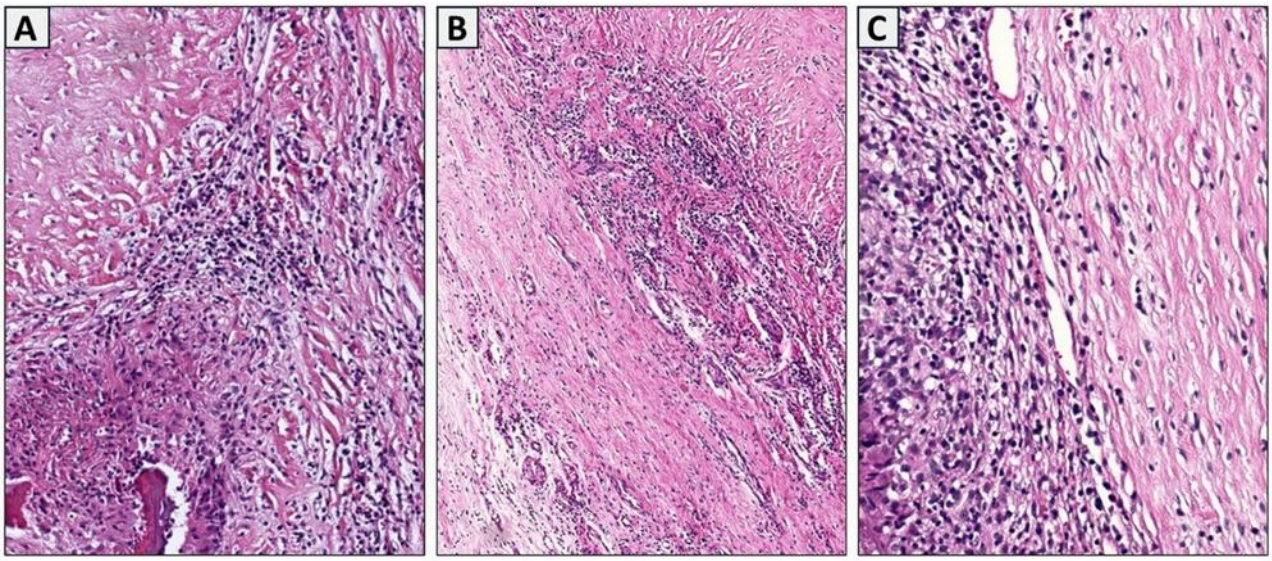
At this point in the evaluation, the differential diagnosis was considerably refined. While the pathology confirmed GPA, the team considered other ANCA-associated vasculitides, including microscopic polyangiitis (though granulomas favored GPA) and eosinophilic granulomatosis with polyangiitis (EGPA, though eosinophilia was absent). The constellation of upper respiratory involvement, pulmonary nodules, glomerulonephritis, and PR3-ANCA positivity confirmed the diagnosis of GPA.

2.4. Treatment, Outcome, and Follow-up

The multidisciplinary team, including rheumatology and nephrology, convened to establish a treatment plan for a newly diagnosed GPA in a post-cardiac surgery patient. Because multiple organs were affected, including the heart, kidneys, lungs, and sino-nasal region, the patient was diagnosed with a severe, widespread condition that necessitates strong immunosuppressive treatment. Induction therapy started with 1000 mg of intravenous methylprednisolone each day for three days, followed by oral prednisone at 1 mg/kg/day, which was tapered down gradually. A weekly dose of rituximab at 375 mg/m^2 was administered over 4 weeks. This option was chosen over cyclophosphamide for comparable efficacy and greater perioperative safety. Prophylaxis against *Pneumocystis jirovecii* with trimethoprim-sulfamethoxazole was initiated.

The patient's postoperative course was complicated by a delayed pericardial effusion requiring drainage, but she experienced no rejection episodes or infectious complications. Her renal function stabilized with creatinine returning to 1.1 mg/dL by three months. Repeat urinalysis showed resolution of hematuria. Maintenance therapy consisted of rituximab infusions every six months and low-dose prednisone (5 mg daily), which was eventually discontinued after 18 months. Serial ANCA titers decreased progressively, becoming undetectable at one year.

At four-year follow-up, the patient remained in complete remission with no evidence of disease activity. Her bioprosthetic valve functioned normally on echocardiography, with preserved left ventricular function (ejection fraction 55%). Renal function remained stable, and surveillance CT imaging showed resolution of pulmonary nodules. She maintained good functional capacity (NYHA Class I) and quality of life on rituximab maintenance therapy alone.



1A: Intermediate power view showing medium-sized vessel of the valve with granulomatous inflammation, necrosis, and surrounding sclerosis (H&E X40); 1B: Low power view showing small vessels of the valve tissue with geographic necrosis and granulomatous inflammation (H&E X20); 1C: High power view showing chronic inflammatory cells infiltrating the vessel wall (H&E X60).

Figure 1 Microscopic examination of the replaced aortic valve with Granulomatosis with Polyangiitis Presenting (GPA)

3. Discussion

3.1. Background (History, epidemiology, risk factors, and WHO classification)

Granulomatosis with polyangiitis (GPA), formerly known as Wegener granulomatosis, is a rare systemic necrotizing vasculitis characterized by granulomatous inflammation and involvement of small- to medium-sized blood vessels. [1] The disease was first described in the early 20th century by Friedrich Wegener, who recognized its classic combination of upper respiratory tract disease, pulmonary involvement, and rapidly progressive glomerulonephritis. [8] GPA is now classified by the World Health Organization as one of the ANCA–associated vasculitides and is officially categorized as one of the three main ANCA-associated vasculitides (AAV). [1] [9]

Epidemiologically, GPA has an estimated annual incidence of approximately 10–20 cases per million people and most commonly affects individuals between 40 and 65 years of age, with no strong sex predisposition. [10] It is more frequently reported in individuals of Northern European descent. [10] The majority of patients demonstrate circulating cytoplasmic ANCA (c-ANCA), typically directed against proteinase-3 (PR3), which plays a key role in disease pathogenesis. [11]

Risk factors for GPA are not fully understood but are believed to involve a combination of genetic susceptibility, environmental exposures, and immune dysregulation. Chronic nasal carriage of *Staphylococcus aureus* and prior upper respiratory tract inflammation have been implicated as potential environmental contributors. [11]

GPA classically involves the upper and lower respiratory tracts and kidneys; however, cardiac involvement is uncommon and often underrecognized. When present, cardiac manifestations may include pericarditis, myocarditis, coronary arteritis, and, rarely, valvular disease. [12] Valvular involvement is typically inflammatory rather than degenerative and may lead to regurgitant lesions, most commonly affecting the aortic valve. [12] GPA is considered a malignant systemic inflammatory disease if untreated; however, with modern immunosuppressive therapy, long-term remission and improved survival are achievable. [12]

3.2. Pathogenesis, Pathophysiology

GPA is a systemic autoimmune disease characterized by necrotizing granulomatous inflammation and vasculitis affecting small and medium-sized vessels. The pathogenesis is intricately linked to ANCAs, particularly those targeting proteinase-3 (PR3-ANCA). These autoantibodies activate neutrophils, leading to degranulation and the release of pro-inflammatory mediators and reactive oxygen species. This cascade results in endothelial damage, vascular inflammation, and granuloma formation. [3] In the context of cardiac involvement, this inflammatory process can directly affect the heart valves. [13] The granulomatous inflammation infiltrates the valvular tissue, causing thickening, friability, and structural damage, which ultimately impairs valve function. This direct inflammatory assault on the aortic valve, as seen in this case, leads to progressive incompetence, often mimicking infective endocarditis due to the destructive nature of the vasculitic process. [14] The healing process can result in a thick, fibrous membrane that encases the valve, distorting its structure and preventing it from closing properly. [5] The heart compensates by dilating and thickening its muscular wall (eccentric hypertrophy) to accommodate the extra blood. [6,15] A patient can remain stable and asymptomatic for years. However, if the inflammation continues or the volume overload becomes too great, these compensatory mechanisms fail, leading to a decline in the heart's pumping function and symptoms of heart failure such as shortness of breath, fatigue, and chest pain. [5]

3.3. Comparative Analysis of our Case with Existing Literature. (Clinical-radiology-pathology, lab, diagnosis, management and outcome)

3.3.1. Clinical Presentation

GPA normally presents with upper airway inflammation, pulmonary nodules, and pauci- immune glomerulonephritis. [16,17] Even so, recent case reports describe patients presenting primarily with cardiac pathology prior to the presentation of systemic disease. [18,19] In most instances, the age of valvular GPA presentation is typically 50–70 years, which differs from the traditional consensus that vasculitides affect younger individuals. [15,18] Our patient fits this emerging demographic profile. The presenting symptoms, including progressive dyspnea, orthopnea, and syncope, reflect severe aortic regurgitation rather than vasculitis itself. Similar heart-failure presentations have been described in recent literature, including cases associated with conduction abnormalities, aortitis, and neurologic involvement. [18,19] However, unlike several published cases, there was no heart block or neurological manifestation. Instead, sinonasal symptoms were noted first, prior to the cardiac presentation. This is an important observation because mild sinus disease and epistaxis are among the earliest manifestations of GPA. [16] A key diagnostic challenge highlighted

across reports is that GPA frequently manifests as infective endocarditis or degenerative valve disease. [19] Culture-negative valvular destruction combined with systemic inflammation is increasingly recognized as a major clinical hint. For this reason, rapidly progressive aortic regurgitation with inflammatory markers should prompt consideration of vasculitis even when usual systemic features appear minimal.

3.3.2. Diagnostic Workup

Diagnosing cardiac GPA is complex. Echocardiography demonstrates leaflet thickening, regurgitation, or intracardiac masses. [18, 20] In our case, severe aortic regurgitation with thickened valve leaflets was consistent with inflammatory valvulitis. Recent studies emphasize the expanding role of cardiac MRI and FDG-PET CT, which can identify myocardial inflammation, aortic root vasculitis, or occult disease in asymptomatic patients. [15, 20] These features have enhanced the detection of cardiac involvement; however, diagnosis is still often made after surgical intervention. Serologic testing supports but does not establish the diagnosis. A positive Proteinase-3 ANCA test is often found in most GPA cases, but seronegativity does not exclude disease. [16,17] Even so, biopsy confirmation remains the gold standard. [21] The present case reinforces the need for definitive diagnosis followed by histopathologic evaluation of the removed valve. Subsequently, this could reveal necrotizing granulomatous vasculitis. Importantly, published cases demonstrate variability in tissue findings. Some cases reveal active vasculitis while others show only post-inflammatory fibrosis. [18] The difference likely reflects timing relative to treatment and disease activity. Several recent case series describe repeated negative blood cultures prior to recognition of GPA. [19] Nonbacterial thrombotic endocarditis and rheumatic disease need to be considered. From the existing literature, inexplicable valvular damage, coupled with systemic inflammation, is a precedent for early ANCA testing and multidisciplinary evaluation. [18]

3.3.3. Management

Management requires synchronized treatment of systemic vasculitis and irreversible structural heart disease. Current recommendations for organ-threatening disease include capitalizing on high-dose glucocorticoids combined with either cyclophosphamide or rituximab. [16,17] Rituximab is preferred due to equivalent remission rates and improved safety. [22] In this case, using pulse corticosteroids followed by rituximab aligns with both EULAR and American College of Rheumatology guidelines. [16,17] Current trials demonstrate non-inferiority of rituximab to cyclophosphamide and possible superiority in relapsing disease. [22] New therapies such as avacopan, a complement C5a receptor inhibitor, may also reduce glucocorticoid exposure while preserving remission. [23] According to the existing literature, immunosuppression cannot reverse established structural valve damage. Several studies describe persistent severe regurgitation despite inflammatory control, dictating valve replacement. [18, 20] The present case supports early surgical intervention once hemodynamic compromise occurs rather than delaying surgery while expecting a medical response. Furthermore, maintenance therapy remains controversial. Rituximab administered twice a year (every 6–12 months), reduces relapse risk, mostly in PR3- ANCA disease. [22] However, long-term therapy could trigger infection and hypogammaglobulinemia risk, which calls for the use of individualized regimens. [22]

3.3.4. Outcome and Follow-up

Modern immunosuppressive therapy has noticeably improved GPA outcomes. Untreated GPA is associated with mortality exceeding 50% and remission rates exceeding 80%. [16,22] In addition, cardiac involvement remains linked to amplified instances of relapses and cardiovascular complications. [15] The favorable long-term outcome in this case, which is sustained remission and preserved ventricular function, compares well with recent studies that show recovery of ventricular function after combined surgical and immunologic treatment. [18] However, literature emphasizes lifelong follow-up since relapse occurs in up to 30–50% of patients within five years. [22] Chronic cardiovascular scrutiny is also suggested due to the increased risk of heart failure and vascular events. [15]

4. What have we learned from this case?

From the existing literature, several consistent principles emerge. GPA should be considered in cases of quickly progressive, culture-negative aortic regurgitation. [15,16,18,19,22] Histopathology, as a gold standard, plays a significant role as a diagnostic strategy even with advancements in imaging. [15,16,18,19,22] Importantly, effective GPA management combines immunosuppression and timely surgery. Long-term follow-up is essential, but early diagnosis is crucial and requires cardiological, rheumatological, nephrological, pathological, and cardiothoracic surgery input. [15,16,18,19,22] With multidisciplinary involvement, diagnostic accuracy and management are achieved, which together improves the patient survival rate.

5. Conclusion

This case report details a rare and diagnostically challenging presentation of Granulomatosis with Polyangiitis manifesting as severe aortic valve incompetence. The primary rationale for reporting this case is to enhance clinical awareness regarding the diverse and sometimes misleading cardiac manifestations of GPA. It serves as a significant reminder that systemic autoimmune diseases can directly impact cardiac structures, necessitating a broad differential diagnosis in patients with unexplained valvular pathology. The value to the medical community lies in emphasizing the importance of integrating clinical, serological, and histopathological findings for accurate diagnosis, particularly when initial presentations are atypical. Early recognition and initiation of targeted immunosuppressive therapy, guided by a multidisciplinary approach, are paramount for improving patient outcomes and preventing disease progression in this complex systemic vasculitis.

Compliance with ethical standards

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Disclosure of conflict of interest

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- Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might be interested in the submitted work.

Statement of ethical approval

Ethical review and approval were not required for this study involving human participants. The paper has been sufficiently anonymized to maintain the patient's confidentiality.

Statement of informed consent

This study was conducted as a retrospective review of archival pathology material collected during routine clinical care. All data were fully de-identified prior to analysis. No patient contact or intervention was involved, and informed consent was not required in accordance with institutional policies.

Data access statement

All relevant data are included in the paper.

Author contributions

All authors contributed equally to producing this manuscript.

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