International Journal of Biomedicine 16(4) (2026) 513-516
http://dx.doi.org/10.21103/Article16(4)_CR1

CASE REPORT

INTERNATIONAL JOURNAL OF BIOMEDICINE

Diagnosis of ANCA-Negative Eosinophilic Granulomatosis with Polyangiitis with Cardiac Involvement and Progressive Neuropathy via Cardiac Magnetic Resonance Imaging and Bone Marrow Biopsy

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Abstract

Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare disorder that may present with cardiac involvement. EGPA diagnosis is a challenge. Our patient is a 57-year-old male with a history of adult-onset asthma who presented with hypoxemic respiratory failure and volume overload. Echocardiography displayed a reduced left ventricular ejection fraction (LVEF) of 42%. The patient didn't improve despite therapy for heart failure. The tests revealed eosinophilia and elevated serum IgE, but ANCA was negative. CMR and bone marrow biopsy findings confirmed secondary/reactive eosinophilia consistent with EGPA. The patient received induction, immunosuppressive, and maintenance therapies, and he improved. Therefore, the rapid and correct diagnosis of EGPA with cardiac involvement helps the patient improve and avoid poor prognosis and severe consequences. **(International Journal of Biomedicine. 2026;16(4):513-516.)**

Keywords: eosinophilic granulomatosis • polyangiitis • cardiac involvement • bone marrow biopsy

For citation: Shuayl MA. Diagnosis of ANCA-Negative Eosinophilic Granulomatosis with Polyangiitis with Cardiac Involvement and Progressive Neuropathy via Cardiac Magnetic Resonance Imaging and Bone Marrow Biopsy. International Journal of Biomedicine. 2026;16(4):513-516. doi:10.21103/ Article16(4)_CR1

Introduction

Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare disorder characterized by late-onset asthma, blood and tissue eosinophilia, and small- to medium-vessel vasculitis,[1] with an annual incidence of 1-3 per 1000000 individuals.[2] EGPA has been described as progressing through three phases: the prodromal, the eosinophilic, and the vasculitic.[3] EGPA is one of the systemic vasculitides that affect the heart; the rate of cardiac involvement ranges from 1% to 29%.[3] Cardiac involvement causes mortality in almost 50%, and it is a poor prognostic factor.[4] However, diagnosing cardiac involvement in EGPA is challenging.[5] Here, we present a 57-year-old patient with ANCA-EGPA diagnosed by cardiac magnetic resonance (CMR) imaging and bone marrow biopsy (BMP).

Case Presentation

History and physical examination

A 57-year-old male was admitted to the emergency department with acute-onset hypoxemic respiratory failure and clinical signs of severe volume overload. His medical history revealed type 2 diabetes mellitus, systemic hypertension, dyslipidemia, and developed adult-onset asthma at 41 years of age, which has been relatively well-controlled with inhaled therapies, but he didn't experience asthmatic manifestations during childhood, and he had no known history of pre-existing ischemic or non-ischemic cardiomyopathy. He had respiratory distress with bilateral pulmonary rales, peripheral pitting edema, and elevated jugular venous pressure.

Laboratory tests and imaging

The tests revealed a markedly elevated B-type natriuretic peptide (BNP) level of 617 pg/mL. Electrocardiogram (ECG) showed normal sinus rhythm with no acute ischemic changes or conduction delays. Transthoracic echocardiography (TTE) showed a normal-sized left ventricle with mildly decreased systolic function (LVEF 42%) and global hypokinesia, and a mildly dilated right atrium. High-flow oxygen therapy and aggressive intravenous loop diuresis were used to stabilize the patient. A diagnostic coronary angiography was performed to rule out macrovascular coronary artery disease, which demonstrated non-significant atheromatous plaques in the mid-left anterior descending (LAD) and right coronary arteries (RCA). Based on optimized Guideline-Directed Medical Therapy (GDMT), our patient was diagnosed with

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heart failure and reduced ejection fraction (HFrEF) and was prescribed sacubitril/valsartan 24/26 mg orally twice daily, empagliflozin 10 mg, and bisoprolol 2.5 mg orally once daily.

Outpatient assessment

Our patient was referred to the specialized Heart Failure Clinic (HFC); there was no active chest pain, orthopnea, paroxysmal nocturnal dyspnea (PND), or ankle swelling. There was no nausea, vomiting, gastroesophageal reflux disease (GERD), oral ulcers, bowel habit changes, generalized weakness, arthralgias, or inflammatory eye pain. However, he was still highly restricted from a functional standpoint with severe persistent dyspnea in agreement with New York Heart Association (NYHA) Functional Class III. He was complaining of a progressive, debilitating numbness in both hands and feet, which compromised his fine motor skills and balance. Laboratory tests, including CBC, kidney function, total serum IgE, and inflammatory markers, were performed. The tests were normal except for eosinophils (%), absolute eosinophil count, and total serum IgE, which were elevated at 14%, 1.4×10^9/L, and 563 IU/mL, respectively. Hepatitis B and C and HIV were negative, with an entirely negative autoimmune serology panel including anti-neutrophil cytoplasmic antibodies (ANCA) and anti-nuclear antibodies (ANA). During subsequent follow-up visits, spironolactone 12.5 mg daily was added to achieve full GDMT. The dyspnea and peripheral neuropathy failed to improve.

Further imaging and biopsy

The patient underwent CMR imaging to investigate the cause of non-ischemic cardiomyopathy and evaluate for suspected myocardial eosinophilic infiltration. The CMR displayed a moderately dilated left ventricle with a calculated LVEF of 46%. There was no evidence of acute myocardial edema. Late gadolinium enhancement (LGE) imaging showed non-ischemic mid-myocardial enhancement involving the basal septum, with a small, localized subendocardial region of enhancement in the mid-to-apical inferior portions. The patient underwent BMP, which displayed isolated eosinophilic hyperplasia with normal maturation and no excess of blasts, confirming secondary/reactive eosinophilia consistent with EGPA.

Management

Aggressive induction therapy was initiated in line with current international guidelines for severe EGPA. The treatment included systemic glucocorticoids, specifically prednisone, initiated at a high dose of 50 mg orally daily to achieve immediate suppression of eosinophilic activation and vascular inflammation, then gradually tapered over several months from the initial induction dose to a minimal chronic maintenance dose of 5 mg daily. He received intravenous cyclophosphamide pulse therapy for a total of three doses and azathioprine at a maintenance dose of 150 mg orally daily. Standard HFrEF therapy was continued, including immunosuppression, to support reverse ventricular remodeling.

Outcomes and follow-up

The response to targeted immunosuppression was quick and favorable. The numbness was resolved totally, with restoration of manual dexterity and balance. The dyspnea improved significantly to NYHA Class I. Follow-up echocardiography and clinical parameters revealed stable cardiac metrics (LVEF at 46%) without recurrence of systemic volume overload or asthmatic exacerbations. The patient remains adherent to azathioprine and a low-dose prednisone regimen under close multidisciplinary surveillance by cardiology and rheumatology.

Discussion

In EGPA with an asthma history, eosinophilia, and both eosinophilic and vasculitic organ damage, the suspicion of EGPA is straightforward. However, some cases lack evidence of vasculitis or ANCA, and there is debate over whether such patients can be recognized as having EGPA.[6] Our patient had asthma, but he displayed negative serology for ANCA, which made EGPA diagnosis hard. The patient was diagnosed as HFrEF based on GDMT; however, the treatment wasn't effective.

Laboratory investigations revealed elevated eosinophils (%), absolute eosinophil count, and total serum IgE. Active EGPA is characterized by eosinophilia > 10%, which correlates with disease activity.[7,8] Additionally, a non-specific rise in IgE levels was discovered in 75% of patients.[9] Therefore, our patient was suspected to have EGPA. However, the cardiac manifestations couldn't be explained as he lacked a history of cardiovascular problems.

ANCA is positive in 30-40% of patients with EGPA. ANCA-negative type is associated with organ damage related to Eosinophilic inflammation[1] and is more prone to develop cardiac involvement.[4] The rate of cardiac involvement in EGPA ranges from 16% to 29%.[3] Clinically evident cardiac manifestations are uncommon despite the wide range of cardiac manifestations, including cardiomyopathy, heart failure, and myocarditis.[10] The patient was not suspected of having EGPA with cardiac involvement, as the signs of cardiac involvement include chest pain, palpitations, arrhythmia, hypotension, and cardiogenic shock.[5] However, the patient didn't experience any of the previous signs.

Cardiac involvement is associated with a poor prognosis; therefore, all patients should undergo basic cardiological examinations, including ECG, troponin, BNP, and echocardiography.[1] Our patient's BNP was significantly elevated, and the initial ECG showed normal sinus rhythm with no acute ischemic changes or conduction delays. Therefore, a more advanced imaging modality was needed to identify the cause of non-ischemic cardiomyopathy in our patient.

Recently, CMR imaging has been considered the gold standard for cardiomyopathy; it is a safe, non-invasive tool for assessing cardiac involvement in ANCA-associated vasculitis.[11] LGE by CMR is characterized by high specificity and sensitivity in detecting fibrosis and cardiac inflammation.[12] The signs of cardiac involvement in EGPA based on CMR

include subendocardial LGE, myocardial edema, and left ventricular dysfunction.[1] Cardiac involvement in EGPA can affect the coronary arteries, endocardium, myocardium, pericardium, and valvular structures.[5]

In our patient, the CMR revealed a moderately dilated left ventricle with an LVEF of 46%, and LGE showed non-ischemic mid-myocardial enhancement involving the basal septum, with a small, localized region of subendocardial enhancement.

Additionally, our patient underwent BMP, as EGPA shares features of vasculitis and hypereosinophilic syndrome (HES),[13] which causes heterogeneity in clinical symptoms, making diagnosis a challenging process.[1] The similarities in clinical presentations made the exclusion of HES and, in some cases, the identification of overlapping HES possible only with BMP or trepanobiopsy and genetic testing.[14] The BMP of our patient revealed marked, isolated eosinophilic hyperplasia with normal maturation and no excess of blasts, confirming secondary/reactive eosinophilia consistent with EGPA. Cardiac involvement in EGPA is strongly linked with eosinophilia.[5]

Therefore, our patient was diagnosed with EGPA based on CMR and BMP. Our patient was diagnosed as an EGPA case with cardiac involvement represented as heart failure with left ventricular dysfunction (46%). A previous meta-analysis revealed that the most common echocardiographic finding was systolic left ventricular dysfunction (83.9%), whereas clinical heart failure was the most common clinical presentation (51.6%).[3]

In the vasculitic phase, manifestations including those related to the nervous system can be observed, and it is often preceded by general manifestations such as muscle pain, fever, and weakness.[1] Additionally, our patient experienced numbness in the hands and feet, which wasn't improved after the initial therapy. However, the patient didn't experience fever, weakness, or muscle pain.

The management of EGPA depends on disease severity and consists of induction and maintenance phases; the former aims to achieve disease remission, whereas the latter aims to prevent relapse.[3] The cornerstone therapy in EGPA is systemic corticosteroids with a recommended initial dose of 1mg/kg/day of prednisolone followed by gradual reduction to the minimal effective dose.[15] Cyclophosphamide is recommended as an immunosuppressant for those who don't respond well to corticosteroids or have poor prognostic factors as defined by the five-factor score, which predicts disease severity and prognosis.[4] Maintenance therapy is preferably azathioprine at a dose of 2-3mg/kg/day.[15] Our patient received induction, immunosuppression, and maintenance therapies, and he improved; these improvements included reduced dyspnea and stable cardiac metrics, with no recurrence of systemic volume overload or asthma exacerbation.

Conclusion

The diagnosis of EGPA with cardiac involvement and neuropathy is challenging, especially with the absence of ANAC. CMR and BMP served as effective non-invasive diagnostic tools that helped correctly diagnose the patient; consequently, he was prescribed appropriate treatment, improved, and a poor prognosis associated with EGPA cardiac involvement was avoided. Therefore, ANCA-negative asthma patients with eosinophilia and cardiac manifestations should undergo CMR and BMP to rapidly confirm the diagnosis of EGPA with cardiac involvement to establish early treatment and avoid poor prognosis and severe consequences.

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