

Case Report

Eosinophilic granulomatosis with polyangiitis: easy to miss at an early stage; easy to halt progression if caught early; a success story presented as a case reportSatharasinghe DS^{1*}, Atukorale H², Siribaddana S³, Sarathchandra C³, Pilapitiya S³¹Postgraduate Institute of Medicine, University of Colombo, Colombo, Sri Lanka²Teaching Hospital, Anuradhapura, Sri Lanka³Faculty of Medicine and Allied Sciences, Rajarata University of Sri Lanka*Corresponding author,  <https://orcid.org/0000-0002-4831-0556>**Abstract**

Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare multisystemic small-vessel vasculitic disease. We report a case of non-severe eosinophilic granulomatosis with polyangiitis who was treated early in its course and remained in remission.

A 57-year-old lady presented with new-onset episodic wheezing for six months. This was associated with rhinitis, sinusitis and chronic urticaria for the same duration. Examination revealed tender skin nodules. Investigations revealed elevated inflammatory markers with positive Myeloperoxidase (MPO) antibodies. She was diagnosed with the early stage of eosinophilic granulomatosis with polyangiitis. She was treated with oral prednisolone and mycophenolate mofetil as induction therapy and treatment is maintained with mycophenolate mofetil. She remains in remission to date.

This case highlights the importance of early detection and treatment of Antineutrophil cytoplasmic antibodies (ANCA)-associated-vasculitis and explores the potential of mycophenolate mofetil as a therapeutic agent.

Keywords: ANCA associated vasculitis, Eosinophilic granulomatosis with polyangiitis, Mycophenolate mofetil**Copyright:** © 2020 Satharasinghe DS et al.  This is an open-access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.**Funding:** None**Competing interest:** None**Received:** 01 October 2020**Published:** 31 December 2020✉ **Correspondence:** sachintha.satharasinghe@gmail.com

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Introduction

Antineutrophil cytoplasmic antibodies ANCA associated vasculitis (AAV) is a rare disease entity mainly seen in the elderly (1) with high morbidity and

population per year, and this entity is more common in Asians and Caucasians (2).

Case presentation

A previously well 57-year-old lady presented with new-onset episodic wheezing for six months associated with rhinitis and sinusitis. She had no history of allergies, childhood asthma, or a family history of asthma.

For the same duration, she had developed episodes of generalized urticaria. Lesions spontaneously disappeared in a few hours after the onset. There was no mucosal involvement and no angioedema. Bilateral hand and feet numbness was developing over a few weeks. She also had a mild intermittent fever, fatigue, malaise, and weight loss of 6 kg over six months.

There was no joint involvement. She did not have chronic cough and a history or contact history of tuberculosis. There was no history of epistaxis, haemoptysis, nasal crusting, haematuria, frothy urine, red or painful eyes, or hearing impairment. Comorbidities included recently diagnosed diabetes mellitus and hypertension.

The examination was unremarkable except for generalized urticarial rash. These lesions disappeared within six to eight hours. Tender skin nodules were noted on the forearm and dorsal foot.

Her white blood cell count was 5.1×10^9 cells/L with 4.8% eosinophils, her haemoglobin level was 9.8 g/dL with an MCV of 80 fL, and the platelet count was 221×10^9 cells/L. The blood picture was suggestive of early mixed deficiency anaemia. ESR was 108 mm/1st hour. CRP was 15 mg/dL. Serum electrolytes, serum calcium, urinalysis, and renal function tests were normal. Liver function tests were unremarkable. The chest x-ray was normal. Mantoux and sputum for acid-fast bacilli (AFB) were negative. The skeletal survey, urine Bence Jones protein (BJP), and serum protein electrophoresis were normal. Ultrasound scan of abdomen and pelvis was normal.

Her antinuclear antibodies (ANA) was negative. In her Antineutrophil cytoplasmic antibodies (ANCA) profile, antibodies against Myeloperoxidase (MPO) antigen was positive, and antibodies against the PR3 antigen were negative. Nasal mucosal biopsy showed normal respiratory epithelium with seromucous glands and a few stromal chronic inflammatory cells. There was no necrotizing vasculitis or granuloma formation with eosinophilic infiltration.

She was referred to the consultant rheumatologist and started treatment for early-stage eosinophilic granulomatosis with polyangiitis with oral prednisolone 40 mg daily and mycophenolate mofetil 250 mg twice daily. Her baseline Birmingham Vasculitis Activity Score (BVAS) was 7 out of 33.

Her wheezing and rash settled within days. After one month of prednisolone and mycophenolate mofetil, prednisolone was started to being tailed off. By six months, her BVAS was improved to 2 out of 33. She's in remission up to now. She is continuing maintenance therapy with mycophenolate mofetil 250 mg twice a day.

Discussion

The American College of Rheumatology criteria for the diagnosis of eosinophilic granulomatosis with polyangiitis (3) is used widely and has a sensitivity of 85%. Our patient does not meet the American College of Rheumatology (ACR) criteria for eosinophilic granulomatosis with polyangiitis due to several factors including, being in the prodromal phase of eosinophilic granulomatosis with polyangiitis where the eosinophilic phase and the vasculitic phase develop later, and some of the disease manifestations might have been masked by the short courses of steroids she had received up to now.

The disease is identified as severe when there are manifestations that threaten the function of vital organs, such as diffuse alveolar haemorrhage, glomerulonephritis, mononeuritis multiplex, sensorineural deafness, scleritis, or gangrene (4). It is good practice to routinely perform urinalysis in people presenting with persistent systemic symptoms and in those with specific features of vasculitis (scleritis, chronic dyspnoea, cough, haemoptysis, foot drop) because these individuals have a high probability of multi-system disease (6).

Severe AAV is generally fatal if left untreated. Moreover, often, after months to years of non-severe manifestations, patients with the non-severe disease develop the severe disease (5). This highlights the importance of early recognition and treatment of the disease. Patients with haemoptysis plus other features of AAV warrant same-day hospital assessment to evaluate for pulmonary haemorrhages (6).

A limited number of studies have compared the treatment options of non-severe EGPA. According to 'MYCYC' trial, Mycophenolic acid (MMF) alone is not proven to be non-inferior to intravenous

cyclophosphamide as induction therapy (7). Our patient treated with glucocorticoid and mycophenolate mofetil as induction and treatment maintenance with mycophenolate mofetil had no relapse by the ninth month follow up. Further studies are required to assess the role of steroid, mycophenolate mofetil combination as induction therapy and as maintenance therapy in non-severe EGPA.

Conclusion

ANCA associated vasculitis has to be considered early in people with chronic systemic symptoms and evidence of renal, pulmonary, ear, nose, and throat, ophthalmic, or peripheral nerve disease. This is strengthened by data supporting a higher prevalence of vasculitis in Asian populations.

Early initiation of treatment possibly can regress or prevent the progression of disease into a severe state. Further studies are needed to explore the potential role of MMF in the management of non-severe Eosinophilic granulomatosis with polyangiitis.

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