DERMATOLOGY-VENEROLOGY

INDEX COPERNICUS INTERNATIONAL

ICV value for 2024 = 82.51

Case Report | International Journal of Medical Case Reports

Eosinophilic Granulomatosis with Polyangiitis Revealed by Cutaneous and Neurologic Manifestations in a Young Woman

S. Jebbouje[1], F. Hali[1], F. Marnissi[2], S. Chiheb[1]

[1] Department of Dermatology-Venerology, Chu Ibn Rochd, Casablanca,Morocco.
[2] Departement Of Anatomopathology, Chu Ibn Rochd, Casablanca, Morocco.

ISSN 2455-0574

ABSTRACT

Background:
Eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as Churg-Strauss syndrome, is a rare immune-mediated vasculitis classified under anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides. It typically involves small- to medium-sized vessels and manifests as a triad of asthma, peripheral eosinophilia, and necrotizing vasculitis with extravascular eosinophilic granulomas. Due to its heterogeneous presentation, diagnosis is challenging and relies on clinical, laboratory, radiological, and histopathological correlation.

Case report:
We present the case of a 37-year-old woman with a history of asthma who developed painful cutaneous lesions, sensory-motor neuropathy, and respiratory complaints. Laboratory investigations revealed marked leukocytosis with eosinophilia, elevated inflammatory markers, and positive ANA with negative ANCA. Skin biopsy confirmed necrotizing vasculitis with eosinophilic infiltration. Imaging demonstrated bilateral ground-glass opacities and peribronchovascular nodules, while bronchoalveolar lavage showed eosinophilic predominance. Electroneurography indicated early polyneuropathy. Based on asthma, eosinophilia, pulmonary infiltrates, neuropathy, and histological findings, EGPA was diagnosed. The patient was treated with high-dose prednisone and cyclophosphamide pulses, leading to rapid hematological and clinical improvement, including resolution of cutaneous lesions and weight recovery, although neuropathic symptoms persisted.

Conclusion:
This case illustrates the multisystemic involvement of EGPA and highlights the diagnostic significance of cutaneous findings in the appropriate clinical context. EGPA should be suspected in patients with adult-onset asthma, unexplained eosinophilia, and systemic manifestations, even in the absence of ANCA positivity. Early recognition and initiation of immunosuppressive therapy are crucial to achieve remission and prevent irreversible organ damage.

Keywords: Asthma, Eosinophilia, Eosinophilic granulomatosis with polyangiitis, Immunosuppressive therapy, Vasculitis

International Journal Of Medical Case Reports (ISSN 2455-0574) is an indexed medical journal indexed in Index Copernicus

Access this Journal Online
Quick Response Code
Website: www.ijomcr.net
Email:ijomcr@gmail.com

INTRODUCTION:

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly Churg-Strauss syndrome, is an extremely rare immune-mediated vasculitis affecting multiple tissues and organs simultaneously. It belongs to the group of anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides (AAV) and occurs with systemic vasculitis involving small- and medium-sized vessels. It is classically described as a triad of asthma, hyper-eosinophilia and necrotizing vasculitis with extravascular eosinophilic granulomas.[1] The global incidence of this disease is 2.5 cases per 100,000 adults per year.[2] Vasculitis is labeled as an EGPA when four or more of these conditions are satisfied. ACR criteria: Presence of four or more of the following: 1) peripheral eosinophilia (more than 10%), 2) asthma, 3) pulmonary infiltrates, 4) paranasal abnormalities, 5) neuropathy, and 6) extravascular eosinophilia on biopsy.[3,4] Herein we report a case of EGPA with Pulmonary, Cutaneous and Neurological manifestations.

S. Jebbouje[1]
Department of dermatology-venerology, chu ibn rochd, Casablanca, Morocco.
email : salihajebbouj@gmail.com

CASE REPORT

A 37-year-old woman with a three-year history of treated asthma presented with a six-week history of a painful, infiltrated plaque on the left thigh, numbness in her left hand and leg, and debilitating joint pain affecting her knee and ankle. She had experienced recurrent respiratory infections and unintentional weight loss (Fig 1).

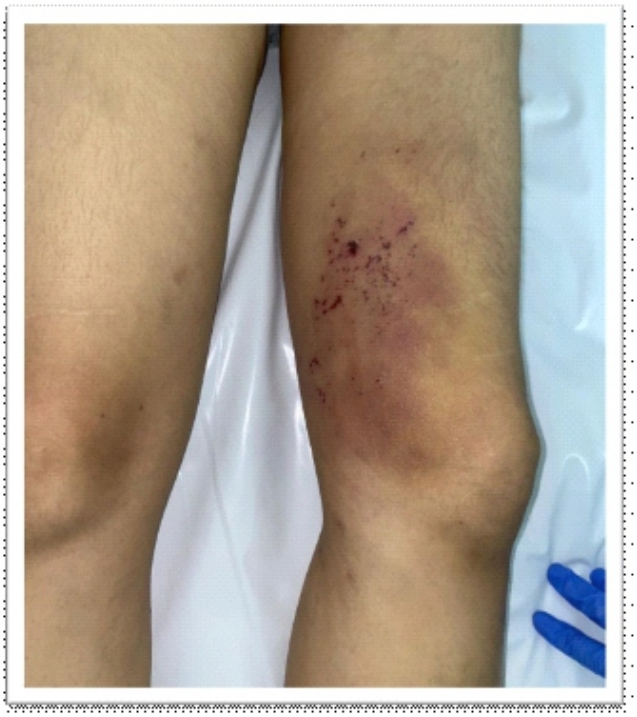

Figure 1: painful infiltrated plaque affecting the surface of the left thigh

Laboratory tests revealed leukocytosis ($30.7\times10^3/\mu L$) with marked eosinophilia ($19.1\times10^3/\mu L$), elevated CRP (114 mg/L), and raised ESR (39 mm/h). Liver enzymes and GGT were mildly elevated; renal function was normal. Autoimmune screening showed a positive ANA (speckled, 1:80), with negative dsDNA, pANCA, and cANCA.

Skin biopsy demonstrated necrotizing vasculitis in the dermal vessels, significant eosinophilic infiltration, and fibrinoid necrosis—findings consistent with EGPA. Chest CT revealed bilateral ground-glass opacities, peribronchovascular nodules, and limited peritoneal fluid; BAL analysis confirmed eosinophilic predominance (17.4%). Electroneurography indicated early length-dependent sensory-motor polyneuropathy. Echocardiography remained within normal limits.

Based on the combination of asthma, eosinophilia, neuropathy, pulmonary involvement, skin vasculitis, and biopsy findings, EGPA was diagnosed. Treatment with prednisone (1 mg/kg/day) and cyclophosphamide pulses was initiated. One week into treatment, eosinophil counts dropped significantly. After eight weeks, the skin lesions had resolved, the patient had gained weight, and her overall condition improved, though neuropathic symptoms persisted.

DISCUSSION

EGPA is to date an incompletely characterized disease due to the heterogeneous presentation and low incidence. It is a type of ANCA-associated vasculitis that predominantly affects small- and medium-sized vessels of many organs simultaneously.[5] The exact pathogenesis of EGPA is not known. Some studies reported that the immunopathology is complex, having features of small vessel vasculitis often overlapping with eosinophilic inflammation. In addition, Th1, Th-2, and Th-17-mediated immunities are described.[6] A landmark genetic study by Lyons et al. demonstrates that seropositive and seronegative EGPA cases correspond to two distinct genetic signatures and can be considered separate disease entities. MPO-negative disease which represents our case, has molecular features similar to asthma and may derive from a pre-existing barrier dysfunction and hyper-eosinophilia.[7,8]

The clinical picture of EGPA is characterized by a history of chronic asthma, rhinosinusitis, and peripheral eosinophilia. Clinical signs, routine laboratory tests, radiological examinations and biopsy results help construct a complete picture of the disease in such patients. Multiple organs and tissues get affected as well as small- and medium-sized vessels, hence, organ-specific evaluation techniques should be used to determine the extent of the disease and it is important for the sake of the treatment.

The best-known lab hallmark of the disease is peripheral blood eosinophilia. Other lab findings may also be present but are non-specific. X-ray and CT chest are beneficial in determining respiratory involvement. Electroneurography is a standard for diagnosing peripheral neuropathy associated with EGPA. Eosinophilia is also observed in skin lesions on biopsy. Recently, the American College of Rheumatology (ACR) and the European Alliance of Associations for Rheumatology have published a new set of EGPA diagnostic criteria having a sensitivity of 85% and a specificity of 99%.[9] It is meant to be used when a diagnosis of small-vessel or medium-vessel vasculitis has been made. For EGPA categorization, a total score of 6 or above is required.

The treatment of EGPA is carried out with corticosteroids and immunosuppressants depending on the disease stage and disease activity. Corticosteroids are beneficial for controlling eosinophilia, inflammation, and symptoms of asthma. Immunosuppressive drugs deal with the autoimmune factors, which play an etiological role in the progression of the disease. Prednisone, methylprednisolone, methotrexate, azathioprine and cyclophosphamide are the most commonly prescribed drugs. Recent studies have revealed that new regimens, mepolizumab (IL-5 monoclonal antibody) and omalizumab are successful in controlling refractory EGPA.[10]

Pharmacological treatment is based on early recognition, irrespective of the etiology and mechanism of the disease. Patients with EGPA are able to achieve disease remission with corticosteroids. It responds well to treatment but is also characterized by a high remission rate and a lingering persistence of difficult-to-control asthma and systemic manifestations affecting the quality of life.Our observation represented a typical picture of EGPA, from clinical features, paraclinical involvements, and therapeutic interventions to the evolution of the disease.[11]

CONCLUSION

EGPA should be considered in patients with adult-onset asthma when unexplained eosinophilia and multisystem symptoms emerge, even in the absence of ANCA. This case underlines that cutaneous manifestations can be subtle yet diagnostically important. Prompt recognition and immunosuppressive therapy can lead to remission of severe symptoms and halt progression of vasculitis.

Conflict Of Interest : None

Source of Funding: Nil

REFERENCES:

1. Medvedev I, Podzolkov VI, Ishina TI, Pisarev MV. Case report: A 64-year-old man with 10-year history of eosinophilic granulomatosis with polyangiitis with bronchiectasis and severe Klebsiella pneumonia. Am J Case Rep. 2021;22:e931150.
2. Greco A, Rizzo MI, De Virgilio A, et al. Churg-Strauss syndrome. Autoimmun Rev. 2015;14(4):341-348.
3. Spencer TL, Tracy CL. Eosinophilic Granulomatosis With Polyangiitis (Churg-Strauss Syndrome): Practice Essentials, Pathophysiology, Etiology [Internet]. Medscape; 2022 [cited 2025 Jul 30]. Available from: https://emedicine.medscape.com/article/333492-overview
4. Ullah S, Ali M, Kyaw P, Nayeemuddin F, Sreedhar R, Gao J. P-ANCA negative eosinophilic granulomatosis with polyangiitis. Respir Med Case Rep. 2019;27:100830.
5. Comarmond C, Pagnoux C, Khellaf M, et al. Eosinophilic granulomatosis with polyangiitis (Churg-Strauss): Clinical characteristics and long-term follow-up of the 383 patients enrolled in the French Vasculitis Study Group cohort. Arthritis Rheum. 2013;65(1):270-281.
6. Furuta S, Iwamoto T, Nakajima H. Update on eosinophilic granulomatosis with polyangiitis. Allergol Int. 2019;68(4):430-436. doi:10.1016/j.alit.2019.06.004.
7. Lyons PA, Peters JE, Alberici F, et al. Genome-wide association study of eosinophilic granulomatosis with polyangiitis reveals genomic loci stratified by ANCA status. Nat Commun. 2019;10:5120. doi:10.1038/s41467-019-12515-9.
8. Koike H, Nishi R, Ohyama K, et al. ANCA-associated vasculitic neuropathies: A review. Neurol Ther. 2022;11(1):21-38. doi:10.1007/s40120-021-00315-7.
9. Robson JC, Grayson PC, Ponte C, et al. 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for granulomatosis with polyangiitis. Arthritis Rheumatol. 2022;74(3):393-399.
10. Pabst S, Tiyerili V, Grohé C. Apparent response to anti-IgE therapy in two patients with refractory "forme fruste" of Churg-Strauss syndrome. Thorax. 2008;63(8):747-748.
11. Kahn JE, Grandpeix-Guyodo C, Marroun I, et al. Sustained response to mepolizumab in refractory Churg-Strauss syndrome. J Allergy Clin Immunol. 2010;125(2):267-270.

*****

Author Contribution:- SJ,FH: Conceptualized, supervised, revised, and edited the manuscript. FM,SC: Acquisition of data. Wrote the original draft, revised, and edited the manuscript.
Received : 10-07-2025 Revised: 15-08-2025 Accepted : 20-09-2025