Case Report

Eosinophilic Granulomatosis with Polyangiitis

Abstract

Eosinophilic granulomatosis with polyangiitis also known as Churg–Strauss syndrome or allergic granulomatosis and angiitis is an extremely rare systemic necrotizing vasculitis that affects small-to-medium-sized vessels. Here, we report a case of a 58-year-old man presenting with painful swelling of bilateral lower limbs with multiple well-defined erosions on lower legs and dorsum of the foot with a few of them showing brownish crusts and with slight watery discharge and a few with raw areas. The patient is a known case of bronchial asthma. Histopathology of foot and face lesion shows inflammatory cell infiltrate predominantly comprising of eosinophils, small- and medium-sized blood vessels are thickened. P-antineutrophil cytoplasmic antibody positivity was seen. Thus, on the basis of positive findings which satisfy the criteria for eosinophilic granulomatosis with polyangiitis, we diagnosed this as a case of eosinophilic granulomatosis with polyangiitis, and for rarity of this case, we would like to report it.

Keywords: *Churg–Strauss syndrome, eosinophilia, eosinophilic granulomatosis with polyangiitis, P-antineutrophil cytoplasmic antibody*

Ratnakar Shukla, Ashwini Kandikatla[1], Nitin J Nadkarni[2], Anant Patil[3]

Department of Dermatology, All India Institute of Medical Sciences, Gorakhpur, Uttar Pradesh, [1]Department of Dermatology, HBT Medical College and Dr. R N Cooper Municipal General Hospital, Departments of [2]Dermatology and [3]Pharmacology, Dr. DY Patil Medical College, Navi Mumbai, Maharashtra, India

Introduction

Churg–Strauss syndrome (CSS), a multisystem disorder, was first described by Jacob Churg and Lotte Strauss in 1951.[1] It is a rare autoimmune disseminated systemic condition with inflammation of small- and medium-sized vessels in a known case of bronchial asthma and tissue eosinophilia. It manifests between 7 and 74 years of age with no gender or ethnic predilection.[2] The incidence is 2.5 cases per 100,000 adults per year. The exact cause is unknown, but various trigger factors including allergens, infections, vaccinations, and drugs are known. It is a Th-2-mediated disease with upregulation of IL-4, 5, and 13. Clinical features are seen in three stages (not always distinguishable); prodromal stage, eosinophilic stage, and vasculitis phase.[3,4]

CSS is also known as eosinophilic granulomatosis with polyangiitis or allergic granulomatosis and angiitis. Necrotizing vasculitis affects small-to-medium-sized vessels and is characterized by excess circulating tissue eosinophils, vasculitis affecting all major organ system and extravascular granuloma. The entity is classified along with Wegener's granulomatosis and microscopic polyangiitis as a small vessel vasculitis. Exact etiopathogenesis is unknown, but autoimmunity is evident with the presence of hypergammaglobulinemia, increased levels of IgE, and antineutrophil cytoplasmic antibody (ANCA).

The disease evolves in three stages: the first stage or prodromal stage is of asthma or allergic rhinitis, the second or transitional stage of peripheral and tissue eosinophilia, and the third or critical stage of life-threatening vasculitis. The treatment is individualized based on the prognostic factors. A combination of high-dose corticosteroids and cyclophosphamide is the gold standard treatment for severe cases. Biologics such as rituximab or mepolizumab seem to be a promising alternative to conventional therapy. We report a case of CSS with cardiac involvement.

Case Report

A 58-year-old male patient presented with complaints of ulcers on lower limbs and skin-colored raised lesions on forehead with periorbital edema for 15 days. The lesions were associated with pain and intermittent rise of temperature. After admission, the patient developed chest pain, radiating to shoulder and back, which on further investigation was diagnosed as myocardial

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Address for correspondence:
Dr. Ratnakar Shukla, Department of Dermatology, All India Institute of Medical Sciences, Gorakhpur, Uttar Pradesh, India. E-mail: ratnakarshukla88@gmail.com

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infarction. The patient was also a known case of bronchial asthma.

Cutaneous examination revealed multiple, well-defined purpura on the lower half of the shin with 2 well-defined ulcers near the ankle. The ulcers were 5 cm × 3 cm in diameter with a sloping edge, nonindurated base with chocolate-colored crusts associated with serous fluid discharge [Figure 1a]. Multiple, well-defined, skin-colored nodules were seen on the forehead associated with periorbital edema [Figure 1b]. On general examination, vitals were stable. Systemic examination showed decreased chest expansion. Blood investigations reported positive p-ANCA with eosinophilia (51.6%), high absolute eosinophil count (12.9%), and raised erythrocyte sedimentation rate. Chest radiograph was suggestive of infiltrative foci [Figure 1c]. Histopathology examination showed fibrinoid degeneration of vessels along with perivascular inflammatory infiltrate predominantly comprising of eosinophils and neutrophils. Periadnexal, perivascular, and perineural eosinophilic infiltrate was seen [Figures 2 and 3]. These features were suggestive of eosinophilic vasculitis.

The patient was started on tablet prednisolone (1 mg/kg). Electrocardiography on 3rd day of admission for chest pain showed anterior wall myocardial infarction with raised creatine kinase-MB (52.8 ng/ml), troponin-I (1.49 ng/ml), and brain natriuretic peptide (416 pg/ml). The patient was successfully thrombolysed after being shifted to the intensive care unit**.** On the basis of clinical presentation and investigations, he was confirmed to have eosinophilic granulomatosis with polyangiitis.

Discussion

Eosinophilic granulomatosis with polyangiitis is seen in three stages (not always distinguishable): prodromal stage is characterized by asthma, allergic rhinosinusitis, and symptoms such as myalgia, arthralgia, malaise, and fever[3,4] Eosinophilic stage is characterized by peripheral and tissue (M/C Lung) eosinophilia, manifesting as Loeffler's syndrome, eosinophilic pneumonia, gastroenteritis, or myocarditis[3,4] Vasculitis stage is characterized by life-threatening organ involvement. Skin involvement in this stage can be seen in the form of palpable purpura or maculopapular erythematous eruption resembling erythema multiforme.[3,4] Systemic features in the form of gastrointestinal, neurologic, cardiac, and renal involvement may also be seen. Cardiac involvement occurs in approximately 15%–60% of the patients. Any cardiac structure can be involved and present with myocarditis, heart failure, pericarditis, arrhythmia, coronary arteritis, valvulopathy, and intracavitary cardiac thrombosis, cardiac involvement may be detected in nearly 40% of the patients.[5] Our patient had cardiac involvement in the form of myocardial infarction. All patients should be studied not only with a detailed history of cardiac symptoms and electrocardiograph but also with echocardiography and if abnormalities are detected, a cardiac magnetic

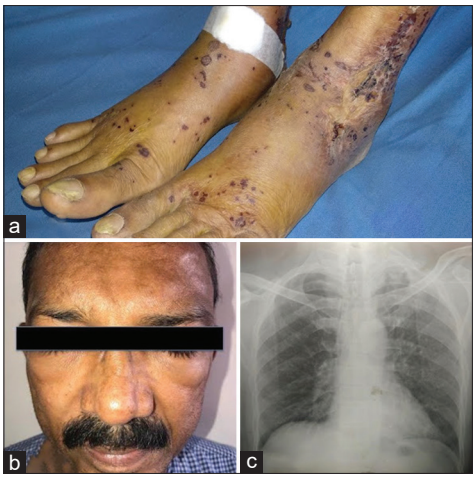

Figure 1: (a) Multiple well-defined purpuric lesions and ulcers on lower limb and dorsum of the foot (b) Erythematous nodules on the forehead and periorbital edema (c) Chest X-ray showing bilateral pulmonary infiltrates

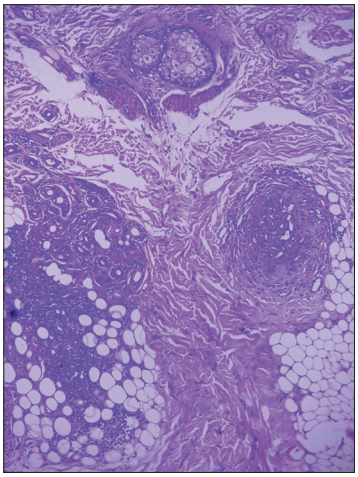

Figure 2: Necrotizing vasculitis of small to medium vessels, eosinophilic infiltration, and extravascular malformed granuloma (×10)

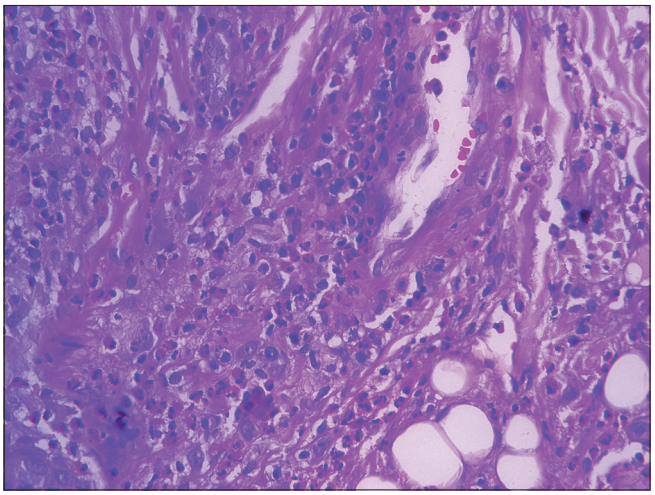

Figure 3: Dermis showing dense eosinophilic infiltrate (×40)

resonance imaging should be performed.[5] Infarction and arrhythmias carry a poor prognosis, responsible for 50% of the deaths.[1,5] Thus, early diagnosis and therapy prevent the progression of cardiac disease.[5] Laboratory abnormalities include eosinophilia −>10%, >1500 cells/µl, high IgE, raised acute phase reactants, and p-ANCA positivity (70%)[1,4] Our patient showed the presence of p-ANCA. The main histopathologic finding is leukocytoclastic vasculitis along with eosinophilic infiltration, extensive necrosis, and interstitial/perivascular necrotizing granulomas.[1]

Primary therapy is systemic glucocorticoid and other immunosuppressants such as cyclophosphamide, azathioprine, and mycophenolate mofetil (MMF) can be used. Other agents which can be used includes- intravenous immunoglobulin, interferon-α, mepolizumab, omalizumab, etc.[3] The duration of treatment is prolonged with the induction of remission followed by maintenance lasting for 12–18 months or longer (azathioprine being the preferred agent with steroids).[3] Optimal therapy for cardiac involvement is based on high-dose corticosteroids plus cyclophosphamide for myocardial inflammation.[5]

Conclusion

Eosinophilic granulomatosis with polyangiitis also known as CSS is rare systemic necrotizing vasculitis. In our case, on clinical examination patient presented with painful swelling of bilateral lower limbs along with multiple well-defined erosions on lower legs and dorsum of foot, associated with swelling of face and periorbital area, histopathology was done to confirm diagnosis which shows inflammatory cell infiltrate predominantly comprising of eosinophils with thickened small and medium sized blood vessel. On the basis of clinical features and investigations, he was diagnosed as eosinophilic granulomatosis with polyangiitis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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