HYPEREOSINOPHILIA PRESENTING AS DIGITAL ISCHEMIA AND CEREBRAL VASCULITIS

Rezgui Amel*, Karmani Monia, Ben Hassine Imen, Ben Fredj Fatma, Mzabi Anis and Laouani Chedia

Internal Medicine Department, CHU Sahloul, Sousse, Tunisia.

Corresponding Author Dr. Rezgui Amel
Internal Medicine Department, CHU Sahloul, Sousse, Tunisia.

ARTICLE

ABSTRACT

Hypereosinophilic syndrome is characterized by an overproduction of eosinophils that leads to organ damage. Although most cases of this syndrome frequently affect the heart, lungs and gastrointestinal tract, there are a few reported cases of vascular involvement. We report here a case with hypereosinophilia, peripheral artery occlusion, digital ischemia, cutaneous and cerebral vasculitis. A 47-year-old Tunisian man presented to us with purpura on lower extremity, swelling and pain on upper extremity and cyanosis of his fingers. Arterial Doppler revealed occlusion of radial and ulnar arteries. The biopsy specimen showed perivascular and periadnexal dense eosinophilic infiltration in dermis and subcutaneous adipose tissue. Laboratory investigations revealed a persistent hypereosinophilia. He was prescribed prednisolone 80 mg daily with nifedipine and Iloprost. His skin lesion, pain and digital ischemia were improved and the eosinophil count was dramatically decreased. After discharge, eosinophil count gradually increased again. Dysarthria and left hemiparesis occurred. The MRI showed lesions of cerebral vasculitis. We prescribed prednisolone, cyclophosphamide and clopidogrel with good results.

KEYWORDS hypereosinophilia, artery occlusion, vasculitis.

INTRODUCTION

Hypereosinophilia is defined by the presence of more than 500 eosinophils per microliter of blood. It occurs in helminthic infestation, allergic reactions, collagen vascular disease, drugs and lymphoma.[1, 2] Hypereosinophilic syndrome (HES), a subcategory of idiopathic eosinophilia, is defined by the presence of a peripheral blood eosinophil count of 1.5 x 10^9/L, or greater for at least 6 months, exclusion of both secondary and clonal eosinophilia, evidence of organ involvement, and absence of phenotypically abnormal and/or clonal T lymphocytes.[3] Although most cases of HES frequently affect the heart, lungs and gastrointestinal tract, there are a few reported cases of vascular involvement. We report here a case with hypereosinophilia, peripheral artery occlusion, digital ischemia, cutaneous and cerebral vasculitis.

OBSERVATION

A 47-year-old Tunisian man presented with upper extremity swelling and pain. He was prescribed antalgic with the impression of having arthralgia. But, this swelling progressed to his both hands and pain was developed. Bluish discoloration then appeared at both hands with necrosis at some areas. Purpura appeared on lower extremity. Laboratory investigations revealed a persistent hypereosinophilia (2000-5000/ mm3). Doppler-duplex arterial examination of the upper extremities showed an occlusion of the radial artery down to the palmar arcade on the right arm, and an occlusion of the cubital and radial artery on the left arm. Upper extremity CT angiography showed that multiple arteries were occlusive (figure 1).

![Figure 1: Occlusion of the radial (A) and cubital (B) artery on the left arm CT angiography.](image)

A biopsy specimen from his leg foot showed perivascular and periadnexal dense eosinophilic infiltration in dermis and subcutaneous adipose tissue. A biopsy of the obliterated left radial artery did not show any distinct vasculitis, but there was a thrombus. Bone marrow finding showed reactive eosinophilia. We also performed microscopic stool exam, ELISA for parasitic antibody, skin prick test for common allergens and bronchial provocation test. The patient had no evidence of connective tissue disease or other known causes of eosinophilia. He was prescribed oral prednisolone 60 mg.
Heparin, clopidogrel 75 mg and Ilprost. The skin lesion, pain and digital ischemia were improved and the eosinophil count was dramatically decreased. One year later after discharge, eosinophil count gradually increased up to 4,565/ L again. Six months later, dysarthria and left hemiparesis occurred. The MRI findings was suggestive of cerebral vasculitis (Figure 2).

We judged that the disease had not remitted with new manifestation. We prescribed prednisolone 60 mg, and cyclophosphamide with good results. We noted a partial regression of the motor deficit and a decreasing in eosinophil count. However, further follow-up is still needed to evaluate whether symptoms and signs change.

**DISCUSSION**

This case has several important characteristics and many differences from typical idiopathic hypereosinophilic syndrome. We have illustrated here several forms of eosinophilic vasculitis; the digital necrosis, the cutaneous vasculitis, the vascular occlusion on arms and finally the cerebral vasculitis.

Some authors suggested that tumor necrosis factor (TNF) might play a part in the mechanism. There was also a description of the deleterious effect of eosinophils on the arterial wall and their role in promoting local development of vasculitis and thrombosis. Although the mechanism of eosinophil-induced hypercoagulability is not entirely clear, contributing factors probably include initiation of the clotting cascade by tissue factor (released during eosinophil degranulation), inhibition of vascular endothelial thrombomodulin (a potent anticoagulant) and activation of platelets. Therefore, the major goal of therapy is to debunk the blood and tissue eosinophil burden.

The digital necrosis and the peripheral artery occlusion has been described in the literature. But this case involved the relatively medium-sized vessels. This is a very rare feature distinctive from previous reported eosinophilic vasculitis. Also, there is no evidence of multi-organ involvement in this case, and this differs from typical idiopathic hypereosinophilic syndrome. The cerebral vasculitis makes also the originality of this case presentation. In fact, HES might be rarely accompanied by eosinophil-mediated tissue injury of central nervous system or peripheral nerve. It is suggested that eosinophils can directly injure nerve tissue, but nerve ischaemia due to eosinophilic infiltration of blood vessel walls, vasculitis, might be the predominant mechanism.

The most common cutaneous manifestations described in HES are either pruritic, erythematous nodules or papules or angio-edematous and urticarial plaques, but rarely a purpuric lesions as had presented our patient. In previous reports, cases with cutaneous vasculitis including eosinophilic infiltration involving small dermal vessel were described.

Most cases of eosinophilic vasculitis are characterized as a glucocorticoid-responsive disorder. Yet in this case, although the eosinophil count was decreased after glucocorticoid therapy, vasculitis was recurrent and progressive. So, the normalization of eosinophils with corticosteroids is not always a sufficient element to prevent the progression of vascular lesions in the HES. Eosinophilic vasculitis can constitute a therapeutic challenge given the resistance to current immunosuppressive regimens.

**REFERENCES**
