Eosinophilia in Rheumatologic/Vascular Disorders

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KEYWORDS

- Eosinophilic granulomatosis with polyangiitis (Churg-Strauss)
- Diffuse fasciitis with eosinophilia • Eosinophilic fasciitis
- Immunoglobulin G4-related disease • Eosinophilic myositis
- Eosinophilia-myalgia syndrome

KEY POINTS

- Rheumatologic and vascular conditions associated with peripheral or tissue eosinophilia are rare and can manifest with single-organ or systemic disease.
- The pathophysiology of rheumatologic and vascular conditions associated with peripheral or tissue eosinophilia remains largely unknown.
- A limited number of randomized controlled therapeutic trials have been performed in rheumatologic and vascular conditions associated with peripheral or tissue eosinophilia so that treatment is largely based on open-label and cohort studies.

INTRODUCTION

Eosinophilia in blood or tissue can be seen in many rheumatologic and vascular conditions (**Box 1**). Although this occurs as an uncommon but described feature in several diseases, for some entities eosinophilia seems to have a significant clinical and pathophysiologic role. When eosinophilia is seen in rheumatic or vascular conditions where this is not generally considered to be an integral part of the clinical picture, care should be taken to consider other causes of eosinophilia. These other causes can particularly include medications; allergic diseases; infections, especially parasitic

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Box 1
Rheumatologic or vascular conditions associated with tissue and blood eosinophilia

Common

EGPA (Churg-Strauss)

IgG4-related disease

DFE

Eosinophilia myalgia syndrome

EM

Less common

GPA (Wegener)

Polymyositis

Dermatomyositis

Inclusion body myositis

Rheumatoid arthritis

Systemic sclerosis

Sjögren syndrome

Systemic lupus erythematosus

Behçet disease

diseases; hematologic conditions; or the possibility of an alternative diagnosis. This article examines 5 rheumatologic and vascular conditions wherein peripheral blood or tissue eosinophilia is a prominent feature: eosinophilic granulomatosis with polyangiitis (Churg-Strauss) (EGPA), immunoglobulin G4 (IgG4)-related disease (IgG4RD), diffuse fasciitis with eosinophilia (DFE), eosinophilia-myalgia syndrome (EMS), and eosinophilic myositis (EM).

EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (CHURG-STRAUSS)

EGPA is a form of primary systemic vasculitis defined by the presence of eosinophil-rich and necrotizing granulomatous inflammation often involving the respiratory tract, necrotizing vasculitis of the small to medium vessels, and an association with asthma and eosinophilia.[1] EGPA carries a wide spectrum of clinical manifestations and can be life-threatening. Although EGPA is considered within the family of antineutrophil cytoplasmic antibody (ANCA) -associated vasculitides, ANCA are found in a minority of patients and cannot confirm or exclude the diagnosis.

Epidemiology

Epidemiologic studies in EGPA have been difficult to perform. The incidence of EGPA has varied between 0 and 3.7 per million person-years with a prevalence of 2 to 22.3 million inhabitants.[2,3] In studies of patients with asthma, the incidence of EGPA was reported to be 34.6 to 67 per million person-years.[4,5] The mean age of diagnosis of EGPA is between 40 and 50 years with an equal occurrence in men and women.

Risk Factors

Two studies found an association between HLA and EGPA. HLA-DRB1*04 and HLA-DRB1*7 seemed to enhance disease risk, whereas HLA-DRB1*03 and HLA-DRB1*13