Eosinophilia Everywhere

Key Points:
• Eosinophilia is most often caused by helminthic infections, allergic reactions, or hematologic/neoplastic disorders
• The hypereosinophilic syndrome is defined as otherwise unexplained eosinophilia of > 6 months duration with evidence of end-organ dysfunction
• Loffler’s syndrome is transient pulmonary infiltrates associated with eosinophilia; Loffler’s endocarditis causes a restrictive cardiomyopathy caused by endomyocardial fibrosis and associated with peripheral eosinophilia

1. What is the differential diagnosis of eosinophilia?
• Allergic: allergic rhinitis, asthma, medication-related
• Infectious:
  -- Parasitic: mainly helminthic infections (Strongyloides, hookworm, Toxocara)
  -- Fungal: aspergillus, coccidiodiomycoses
  -- HIV: usually not just caused by HIV but related to relative lymphopenia, medication reaction, adrenal insufficiency, or eosinophilic folliculitis
• Hematologic/Neoplastic: hypereosinophilic syndrome (see below), systemic mastocytosis, eosinophilic leukemia, lymphoma
• Collagen-Vascular: Churg-Strauss is fairly unusual among the vasculitides for causing eosinophilia
• Miscellaneous: adrenal insufficiency, atheroemboli, congenital immunodeficiencies

2. What is the hypereosinophilic syndrome?
• Definition: blood eosinophilia > 1500/µl for > 6 months
  exclusion of other causes of eosinophilia
  signs and symptoms of endorgan dysfunction
• Male: female = 9:1; most commonly occurs in 4th decade
• Symptoms include fatigue, cough, SOB, myalgias, angioedema, rash, and fever; however, manifestations of the disease can range from nearly asymptomatic and subacute to fulminant
• Organ systems involved include cardiovascular (see below under Loffler’s endocarditis -- endomyocarditis progressing to a fibrotic, restrictive cardiomyopathy), CNS (thromboemboli, encephalopathy, peripheral neuropathy), skin (angioedema, urticaria, nonspecific nodules or papules), pulmonary (frequently with a negative CXR)
• May be differentiated from eosinophilic leukemia by absence of immature eos, < 10% blasts in marrow
• Rx with steroids, cytotoxic agents; prognosis depends on extent of end-organ involvement

3. Loffler’s syndrome vs. Loffler’s endocarditis?
• Loffler’s endocarditis = a.k.a. endocarditis parietalis fibroplastica; endomyocardial fibrosis associated with peripheral eosinophilia, causing a restrictive cardiomyopathy.
  -- Most patients have the hypereosinophilic syndrome but some may have other causes of eosinophilia, as the eosinophils are thought to be responsible for the endomyocardial damage
• Loffler’s syndrome = transient pulmonary infiltrates and peripheral eosinophilia, classically caused by helminthic pulmonary infections (Ascaris, hookworm, or Strongyloides) but may also be caused by crack use, certain medications, nickel exposure, blood transfusion
• Neither are to be confused with Lofgren’s syndrome, which is an acute presentation of sarcoidosis associated with erythema nodosum, migratory polyarthralgias, hilar adenopathy, and fever; seen primarily in women

References
On hypereosinophilic syndrome: UpToDate 10.2 has an excellent review (also a good review on eosinophilia).