PULMONARY EOSINOPHILIC Syndromes

Diagnostic Approach

Characteristics

- Increased respiratory symptoms with increased eosinophils in peripheral blood, lung tissue, sputum or BAL fluid
- Pulmonary infiltrates
- Possible signs or symptoms of systemic disease

STEP 1

Basic evaluation

• DETAILED HISTORY AND PHYSICAL EXAM
• ASK ABOUT EXPOSURES: Medications, dietary supplements, recreational drugs, toxins, workplace, travel
• SEARCH FOR EXTRAPULMONARY DISEASE: Nasal/sinus, ocular, skin, cardiac, gastrointestinal, neurologic, renal, vascular
Tailor according to history and physical:
  • IMAGING: CXR, chest CT scan, sinus CT scan, echocardiogram
  • LABORATORY: CBC with differential, ESR, CRP, ANCA, vitamin B12, electrolytes, LFTs, total protein, urinalysis
  Serum tryptase level, serum Ig's (include IgE)
  Bone marrow biopsy with reticulin and tryptase stains

STEP 2

Exclude major secondary causes of pulmonary eosinophilia

DISORDERS TO EXCLUDE

- Drug or toxin reaction
- Parasitic/helminthic infection
- Nonparasitic infection (TB, Coccidioides)
- Leukemia/lymphoma

EVALUATIONS AND TESTS TO CONSIDER

- Drug and dietary supplement history
- Discontinuation of potential causative agents
- Travel history
- Stools for ova and parasites
- Serologies for parasitic infection (based on travel history)
- TB skin test, IGRA, M. tuberculosis cultures
- Coccidioides serologies, fungal cultures
- Bone marrow aspirate/biopsy with cytogenetics, immunophenotyping
- RT-PCR analysis of PBMCs for BCR-ABL fusion gene
- Abdomen/chest CT, gallium scan, PET scan
- c-KIT mutation analysis, serum tryptase level
- CT/MR scan
- HIV
  - HIV serology
- Primary allergic disorders
  - Allergen skin testing, Aspergillus precipitins, specific RASTs
- Autoimmune disorders
  - Auto-antibodies (ANA & ANCA), specialized clotting studies
- Systemic mastocytosis
  - c-KIT mutation analysis, serum tryptase level
- Solid tumors
  - CT/MR scan

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Exclude other diseases associated with eosinophilia

LUNG DISEASES
- Asthma/eosinophilic bronchitis
- Allergic bronchopulmonary aspergillosis
- Bronchocentric granulomatosis
- Cryptogenic organizing pneumonia
- Hypersensitivity pneumonitis
- Idiopathic pulmonary fibrosis
- Pulmonary Langerhans cell histiocytosis
- Postradiation pneumonia

SYSTEMIC DISEASES
- Rheumatoid arthritis
- Sarcoidosis
- Sjögren syndrome

Distinguish between the primary pulmonary eosinophilic disorders

<table>
<thead>
<tr>
<th>KEY FEATURES</th>
<th>ACUTE EOSINOPHILIC PNEUMONIA</th>
<th>CHRONIC EOSINOPHILIC PNEUMONIA</th>
<th>HYPEREOSINOPHILIC SYNDROME</th>
<th>EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS*</th>
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<tbody>
<tr>
<td>Onset</td>
<td>Acute (days)</td>
<td>Indolent (weeks/months)</td>
<td>Indolent (months/years)</td>
<td>Indolent (months/years)</td>
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<tr>
<td>Pulmonary infiltrates</td>
<td>Diffuse</td>
<td>Peripheral</td>
<td>Patchy</td>
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<tr>
<td>Fulminant respiratory failure</td>
<td>Frequently Occurs</td>
<td>Rarely Occurs</td>
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<td>Asthma/allergy history</td>
<td>Rarely Occurs</td>
<td>Commonly Occurs</td>
<td>Rarely Occurs</td>
<td>Frequently Occurs</td>
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<td>Smoking history</td>
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<td>Vasculitis</td>
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<td>ANCA</td>
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<td>Commonly Occurs</td>
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<td>Cardiac involvement</td>
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<td>Occasionally Occurs</td>
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<td>Commonly Occurs</td>
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<tr>
<td>Neurologic involvement</td>
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<td>Frequently Occurs</td>
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<td>Requirement for therapies other than corticosteroids</td>
<td>Rarely Occurs</td>
<td>Rarely Occurs</td>
<td>Frequently Occurs</td>
<td>Commonly Occurs</td>
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</tbody>
</table>

* Eosinophilic granulomatosis with polyangiitis (EGPA) was previously called Churg-Strauss syndrome.
• BAL, transbronchial or open-lung biopsy may be necessary, depending on clinical and radiographic findings.
• Additional testing may be needed to evaluate for extrapulmonary disease.

Abbreviations:
BAL, bronchoalveolar lavage; ESR, erythrocyte sedimentation rate; CRP, C-reactive protein; ANCA, antineutrophil cytoplasmic antibody; Ig, immunoglobulin; LFT, liver function test; TB, tuberculosis; IGRA, interferon gamma release assay; RT-PCR, reverse transcriptase-polymerase chain reaction; PBMC, peripheral blood mononuclear cell; PET, positron emission tomography; RAST, radioallergosorbent test.