Allergy and Immunology Review Corner: Chapter 49 of *Middleton's Allergy Principles and Practice,* Seventh Edition, edited by N. Franklin Adkinson, et al.

Chapter 49 – Eosinophilia and Eosinophil-Related Disorders

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1. Which protein is encoded by a molecular defect on chromosome 4 in some cases of Hypereosinophilic Syndrome?
A. FIP1L2/PDGFRA
B. FIP1L1/PDGFRA
C. FIP1L2/PDGFRB
D. FIP1L1/PDGFRB

2. Which parasite is the most common cause of Loffler's syndrome worldwide?
A. *Ascaris lumbricoides*
B. *Wucheria bancrofti*
C. *Strongyloides stercoralis*
D. *Paragonimus* lung flukes

3. Churg- Strauss Syndrome affects which type of vessel(s):
A. Small- sized arteries only
B. Medium-sized arteries only
C. Large-sized arteries only
D. Small- and medium- sized arteries and veins

4. Which is the second most common organ system involved in HES?
A. Pulmonary
B. Cutaneous
C. Cardiovascular
D. Hematologic

5. What is the most common symptom in histiocytosis X?
A. Non- productive cough
B. Cough productive of dark- colored secretions
C. Fever
D. Chest pain

6. Which of the following medications causes pulmonary pleural effusions with eosinophilia?
A. Aztreonam
B. Cimetidine
C. Dantrolene
D. Rifampin

7. Which of the following cause eosinophilia?
A. Giardia lamblia
B. Isospora belli

C. Entamoeba histolytica
D. Plasmodium knowlesi

8. Which of the following diseases presents as large subcutaneous masses on the head and neck of Oriental males?
A. Kimura's disease
B. Shulman's Syndrome
C. Well's Syndrome
D. Recurrent cutaneous necrotizing eosinophilic vasculitis

9. Which of the following has acute onset of erythema, swelling, and induration of the extremities often presenting after exercise?
A. Kimura's disease
B. Shulman's Syndrome
C. Well's Syndrome
D. Recurrent cutaneous necrotizing eosinophilic vasculitis

10. Which of the following is marked by recurrent swelling of the extremities and eosinophilic infiltrate of the dermis?
A. Kimura's disease
B. Shulman's Syndrome
C. Well's Syndrome
D. Recurrent cutaneous necrotizing eosinophilic vasculitis

Answers

1. B, page 864

In some of the myeloproliferative HESs, a molecular defect has been identified as a chromosome 4 deletion that yields fusion gene encoding FIP1L1/PDGFRA (platelet derived growth factor alpha- protein) protein that constitutively express receptor kinase activity.

2. A, page 869

Ascaris lumbricoides is the most common cause of Loffler's syndrome worldwide.

3. D, page 873

According to the diagnostic criteria established by Churg and Strauss, necrotizing vasculitis of small- and medium- sized arteries and veins occurs in Churg -Strauss Syndrome.

4. C, page 865

Table 49.4 lists the frequency of organ involvement in HES. The cardiovascular system is the second most common organ system involved. In 58% of HES cases, the cardiovascular system is involved.

5. A, page 871

Eosinophilic granuloma of the lung (histiocytosis X) is associated with pulmonary interstitial fibrosis. The most common symptom is non- productive cough. CT scan of the lungs shows cysts and nodules.

6. C, page 862
Dantrolene causes pleural effusions and eosinophilia. The other medications listed can cause interstitial nephritis.

7. B, page 863
Unlike infections with helminthic parasites, infections with single-celled protozoan parasites do not elicit blood eosinophilia. *Isospora belli* and *Dientamoeba fragilis* are the only two protozoa that cause eosinophilia.

8. A, page 868
Kimura's disease is an angiolymphoid hyperplasia presenting with eosinophilia and large subcutaneous masses on the head and neck of Orientall males.

9. B, page 868
Shulman's syndrome is an eosinophilic fasciitis presenting as acute onset of erythema, swelling, and induration of the extremities often presenting after exercise. The dermis and epidermis are normal with pathology in the tissue, fascia, and muscle. Tissue eosinophils are increased in half to two-thirds of patients.

10. C, page 868
Well's syndrome is a recurrent cellulitis with eosinophilic infiltrate in the dermis. It does not respond to antibiotics but resolves spontaneously leaving a granulomatous infiltrate. Half of patients have blood eosinophilia.

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Chapter 50: Eosinophilic Gastroenteropathies – Pages 879-891

Prepared by Eiman Al- Selahi, Childrens' Hospital of Winnipeg, and Christopher R. Martin, MD, Walter Reed Army Medical Center

1. What percentage of patients with an EGID has an immediate family member afflicted with the disorder?
A. 10%
B. 25%
C. 33%
D. 50%

2. A good response to imatinib would be expected in HES patients that have a fusion gene producing what novel kinase?
A. FIPL1-PDGFRA
B. FIPL2-PDGFRA
C. FIPL1-PDGFRB

D. FIPL1-TNF-α

3. Which symptom of EE is more common in older teens?
A. Vomiting
B. Weight Loss
C. Halitosis
D. Food Impaction

4. Which is true of eosinophilic gastritis?
A. History of food allergy is required for diagnosis
B. Presence of peripheral eosinophilia is required for diagnosis
C. Aggregates of lymphocytes and eosinophils are present in micronodules
D. Serosal form of gastritis makes up the majority of cases

5. What percent of EE patients have symptom improvement with dietary elimination of allergic foods based on skin testing?
A. 10%
B. 25%
C. 50%
D. 75%

6. Before using systemic immunosuppression for EGID, it is especially important to rule out infection with what specific organism?
A. Ancylostoma caninum
B. Strongyloids stercoralis
C. Helicobacter pylori
D. Pinworm

7. What is the number of Eosinophils required for the diagnosis of HES?
A. 1600/mm3
B. 1700/mm3
C. 1500/mm3
D. 1400/mm3

8. Which treatment modalities is not used in EGID
A. Elemental diet
B. Allergen avoidance
C. Anti IL-4
D. Oral suspension of budesonide

9. All are histological types of primary eosinophilic gastroenteritis EXCEPT
A. Subepithelial
B. Mucosal
C. Muscularis
D. Serosal

10. Concerns regarding untreated EE include all of the following except?
A. Progressive esophageal scaring and dysfunction
B. Concern of developing Barrett's Esophagus
C. Increased risk of developing other forms of EGID
D. Risk of Hodgkin's lymphoma

Answers
1. A, page 879
Evidence is developing that EGID has a large genetic component in addition to environmental factors.

2. A, page 883

3. D, page 884
Abdominal pain, dysphagia, and food impaction become more common as a child ages.

4. C, page 887
Muscularis form is more common and food allergy and eosinophilia is not required for the diagnosis

5. D, page 885
The vast majority of patients with EE improve when food triggers are identified and eliminated from their diet. The amount of improvement however is variable.

6. B, page 882
Strongyloids stercoralis should be ruled out because this infection can become life threatening in the setting of immunosuression.

7. C, page 883
Persistent eosinophilia of at least 1500/mm3

8. C, page 885

9. A, page 886

10. D, page 886