



Syndromes that Masquerade as Infections

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Disclosures of Financial Relationships with Relevant Commercial Interests

- None



With Appreciation to
Dr. Bennett Lorber

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Fungi	5%
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Internal Medicine and Non-Infectious Syndromes	18%
Total	100%

MARCH 2026

SEE ENLARGED SLIDE FOLLOWING ENTIRE PRESENTATION

General Internal Medicine-1

INTERNAL MEDICINE AND NON-INFECTIOUS SYNDROMES (18% of exam)	Diagnosis/Testing	Treatment/Core Decisions	Risk Assessment/Prognosis/Epidemiology	Pathophysiology/Basic Science
GENERAL INTERNAL MEDICINE				
Malignancies	HIGH	MEDIUM	MEDIUM	LOW
Hemophagocytic lymphohistiocytosis (Hemophagocytic syndrome)	LF MEDIUM	MEDIUM	MEDIUM	LOW
Noninfectious inflammatory disorders (e.g., vasculitis, granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis, aortitis)	HIGH	MEDIUM	MEDIUM	MEDIUM
Dermatologic disorders	HIGH	MEDIUM	MEDIUM	LOW

SEE ENLARGED SLIDE FOLLOWING ENTIRE PRESENTATION

General Internal Medicine-2

INTERNAL MEDICINE AND NON-INFECTIOUS SYNDROMES continued... (18% of exam)	Diagnosis/ Testing	Treatment/ Care Decisions	Risk Assessment/ Prognosis/ Epidemiology	Pathophysiology/ Basic Science
GENERAL INTERNAL MEDICINE continued...				
Hematologic disorders	MEDIUM	MEDIUM	MEDIUM	MEDIUM
Noninfectious central nervous system disease	MEDIUM	MEDIUM	MEDIUM	LOW
Bites, stings, and toxins	MEDIUM	MEDIUM	MEDIUM	LOW
Drug fever	HIGH	HIGH	MEDIUM	LOW
Ethical and legal decision making	Not Applicable	MEDIUM	MEDIUM	Not Applicable

SEE ENLARGED SLIDE FOLLOWING ENTIRE PRESENTATION

IM and Non-Infectious Syndromes

- Surgical Infections
- Critical Care Medicine
 - SIRS
 - VAP
 - ARDS/eosinophilic pneumonia
 - Hyperthermia/hypothermia

Mimics

- Many conditions masquerade as infections
 - Fever almost universally present
 - Sometimes focal abnormality
 - Cellulitis vs. stasis dermatitis
 - Viral vs. Organizing Pneumonia
 - Lymphadenitis vs. Lymphoma
 - ID likely to be consulted!



vs.



Test Taking Tips

- Emphasis on high-importance content (70% of Q)
- For non-infectious syndromes, this is heavily skewed toward **diagnosis**
 - Limited emphasis on treatment, epidemiology or prognosis
 - No questions on pathophysiology

INTERNAL MEDICINE AND NON-INFECTIOUS SYNDROMES (18% of exam)	Diagnosis/ Testing	Treatment/ Care Decisions	Risk Assessment/ Prognosis/ Epidemiology	Pathophysiology/ Basic Science
GENERAL INTERNAL MEDICINE				
Malignancies	HIGH	MEDIUM	MEDIUM	LOW
Hemophagocytic lymphohistiocytosis (hemophagocytic syndrome)	LP	MEDIUM	MEDIUM	LOW
Noninfectious inflammatory disorders (e.g., vasculitis, granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis, sarcoid)	HIGH	MEDIUM	MEDIUM	MEDIUM
Dermatologic disorders	HIGH	MEDIUM	MEDIUM	LOW

SEE ENLARGED SLIDE FOLLOWING ENTIRE PRESENTATION

Test Taking Tip

- Just as for infections, look for “buzzwords” and “hooks” in red font in the lecture
- For infections:
 - If I say, “skinned rabbit”, you say...

Test Taking Tip

- For infections:
 - If I say, “rabbit”, you say.....



(Pulmonary) TULAREMIA

Test Taking Tip

If I say, “chitterlings” (aka chitlins, aka hog intestines)

You say.....



Test Taking Tip

If I say, “chitterlings”

You say.....



YERSINIA (gastroenteritis)

Test Taking Tip

If I say, "Bull's-eye rash"

You say.....



Test Taking Tip

If I say, "Bull's-eye rash"

You say.....



Lyme disease
(or **Erythema migrans** or **STARI**)

My Approach to Mimics

- Think like an Internist
- The key is recognition, not treatment
- This talk will emphasize illustrative cases
- Goal is to cover lots of non-infectious diseases rather than in-depth discussion using **buzzwords** for easy recognition!

Cases



Question #1

- 26-year-old man presents with a 1-month h/o fever, night sweats and 10 lb. weight loss
- This started with a sore throat, but RADT was negative for Strep, and pharyngitis has resolved
- He lives in Indiana with his wife and 2 yo son, who are healthy. They have 2 cats.

Question #1

- Exam:
 - Vitals:
 - T=**38.4°C**, HR=118 bpm
 - No tonsillar erythema or exudate
 - No lymphadenopathy
 - No rash
 - **Palpable spleen tip**
- Labs
 - CBC
 - WBC=**2.7**, plt=**53**
 - Normal H/H
 - Normal Cr
 - AST/ALT=**120/200**
 - Alk phos=**494**, bili=**1.9**
 - Ferritin=**35,148** mg/ml

Question #1

What is the most likely cause of this presentation?

- A. Lymphoma
- B. Rheumatic fever
- C. Acute EBV infection
- D. Bartonella endocarditis
- E. Histoplasmosis

Question #1

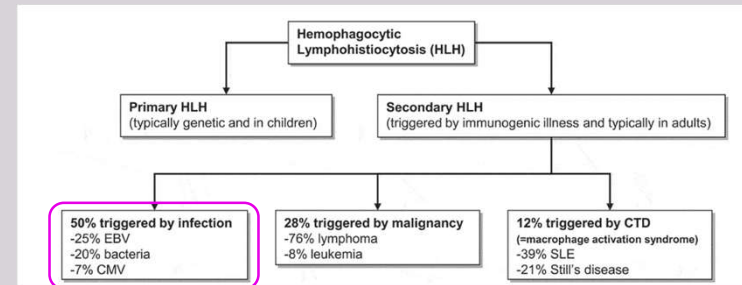
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- A. Lymphoma
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- D. Bartonella endocarditis
- E. Histoplasmosis

Hemophagocytic Lymphohistiocytosis

- AKA HLH
- Immune activation syndrome
 - Primary (Peds): Familial due to genetic mutation
 - Secondary (Adult or peds):
 - Infections (**EBV** or other herpes group viruses, HIV, histoplasmosis, Ehrlichia, COVID-19 etc.)
 - Malignancy (lymphoma, leukemia)

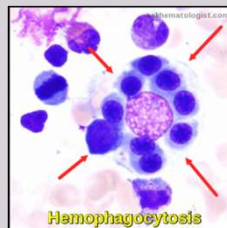
HLH & Infection



SEE ENLARGED SLIDE FOLLOWING ENTIRE PRESENTATION

HLH-2024: Diagnostic Criteria

- At least **5/7** of the following:
 - Fever ($T > 38.5^{\circ}\text{C}$)
 - Splenomegaly
 - Cytopenias (2/3 lineages)
 - Hypertriglyceridemia ($> 3\text{mmol/L}$) and/or hypofibrinogenemia
 - **Ferritin $> 500\text{ mcg/mL}$**
 - Elevated soluble IL-2 receptor (aka sCD25)
- NK cell function now a functional criteria



HLH Pearls

- Antecedent **mono that doesn't get better**
- **Cytopenias**
- **Elevated ferritin**
- Treating an underlying infection is often curative

Question #2

- A 39-year-old woman is admitted for fever of 3 weeks associated with diffuse arthralgias involving the knees, wrists and ankles
- A severe sore throat was present during the first week of the illness but has resolved

Question #2

Physical Exam

- T=104.2° F
- Tender cervical LAN appreciated
- Spleen tip is palpable
- Both knees are swollen & painful
- A rash is present on the trunk and extremities, most prominently under the breasts and in her underwear waistband



Question #2

- Labs:
 - Ferritin **3600** ng/ml (nl 40-200)
 - WBC **32,200** (89% neutrophils)
 - **AST and ALT 3x** normal
 - **ESR and CRP 5x** normal
 - ANA and RF negative
 - Throat and blood cultures are negative
- On afternoon rounds with the attending, the fever has resolved with Tylenol, and the rash is no longer present

Question #2

Which of the following would support the diagnosis?

- A. Elevated eosinophils
- B. Koebner phenomenon
- C. Positive hepatitis B antigen
- D. Elevated LDH
- E. Elevated ASO titer

Question #2

Which of the following would support the diagnosis?

- A. Elevated eosinophils
- B. Koebner phenomenon***
- C. Positive hepatitis B antigen
- D. Elevated LDH
- E. Elevated ASO titer

Adult Still's Disease (Adult-Onset JRA)

Yamaguchi Criteria: (5 features with 2 major criteria)

Major:

1. Fever $>39^{\circ}\text{C}$ for >1 week
2. Arthritis/arthralgia >2 wks
3. Typical rash (during febrile episodes)
4. Leukocytosis $>10\text{K}$ with $>80\%$ PMNs

Minor:

1. Sore throat
2. Lymphadenopathy
3. Lg Liver or spleen
4. Abnl LFTs
5. Negative ANA & RF

Adult Still's Disease

- Buzzwords and associations:
Evanescant, salmon-colored rash



=



Adult Still's Disease

- Buzzwords and associations:

Koebner phenomenon

Rash at pressure sites
(bra/underwear)

Can be trivial such as hot shower



Question #3

- A 24-year-old man was referred by the ED for evaluation of ulcers of the mouth and penis. He was born in Japan and is in the U.S. to attend graduate school.
- He has a history of recurrent painful oral ulcers for 3-4 years. Four days ago, he developed a painful ulcer on the penile shaft. He takes no medicines and denies sexual contact for the past 5 years.

Question #3

- Left eye is inflamed and there is a hypopyon
- Numerous painful ulcers on the oral mucosa
- There is a 0.5cm ulcer on the penis



Question #3

Which would support the diagnosis?

- A. HSV PCR
- B. Biopsy of oral ulcer
- C. Papule at site of venipuncture
- D. Painful inguinal lymph node
- E. Histoplasma antigen

Question #3

Which would support the diagnosis?

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- D. Painful inguinal lymph node
- E. Histoplasma antigen

Behçet's Disease

- Pleomorphic vasculitis diagnosed **clinically**
- Recurrent **oral ulcers** (>3 per year) PLUS 2 of the following
- Recurrent **genital ulcers**
- **Eye** (uveitis, retinitis, hypopyon)
- Skin lesions, esp **pathergy**
- Less common manifestations
 - GI disease (abdominal pain, bloody diarrhea)
 - Aseptic meningitis
 - Arterial and venous thrombosis

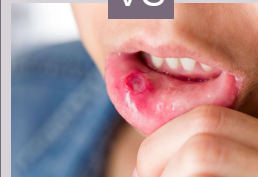


Behçet's Disease

- **Ulcers** is the buzzword, but the trick is differentiation from infectious causes (HSV, coxsackie, etc.)
- Additional Clues
 - **Recurrence**
 - **Ocular findings**
 - **Pathergy** (needle or IV site)



VS



Question #4

- A 38-year-old woman with AML is admitted with fever. She underwent induction chemotherapy 2 weeks prior, complicated by neutropenic fever that resolved with marrow recovery
- She presents with a 1-day history of fever without localizing symptoms
- Exam: T 101.4°F; P 98; otherwise, unremarkable
- CBC showed a white blood cell count of 12,250 with 20% bands

Question #4

Hospital Day 2:

- Fever persists despite broad spectrum antibiotics
- Interval development of raised, red-purple, tender papules and nodules on her face, neck and the dorsum of her hands



Question #4

Hospital Day 3:

- Fever persists; some of the papules develop a plaque-like appearance

Hospital Day 4:

- Skin biopsy with dense perivascular infiltrates of neutrophils without evidence of vasculitis; stains for organisms negative



Question #4

Which is most likely to be diagnostic?

- A. Flow cytometry
- B. Naproxen challenge
- C. Bone marrow biopsy
- D. Skin biopsy
- E. PET scan

Question #4

Which is most likely to be diagnostic?

- A. Flow cytometry
- B. Naproxen challenge
- C. Bone marrow biopsy
- D. Skin biopsy***
- E. PET scan

Sweet Syndrome

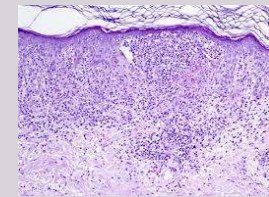
- AKA acute febrile neutrophilic dermatosis
- Three variants:
 - Idiopathic or "classical" >50% (IBD, post viral illness, preg, etc.)
 - Malignancy associated ~20% (may precede dx, AML most frequent)
 - Drug induced-G-CSF most common, antibiotics
- **Fever and Rash** universally present
- Rarely oral ulcers or extra-cutaneous disease characterized by neutrophilic infiltrate on path
- Lab tests with leukocytosis with left shift, inc ESR & CRP
- Path diagnostic – **Neutrophilic infiltrate without vasculitis**

Skin Lesions in Sweet Syndrome



- Lesions appear **abruptly** and usually **tender**
- May be single or multiple, often involving **dorsum of hand**
- Red, violaceous, or yellow center
- Nodular or **plaque-like**
- Central umbilication with **target appearance**

Sweet Syndrome



- Buzzwords and associations:
 - Fever and a rash
 - Neutrophilia (peripheral and on **path**)
- Be suspicious in patients with malignancy (esp **AML**), **IBD**, recent **URI**, vaccination, pregnancy, or colony stimulating factor use in preceding 2 weeks

Question #5

- A 33-year-old recent immigrant from Central America is seen for a leg ulcer
- The ulcer has progressively enlarged over 3 months after he bumped his leg on a table
- There has been no response to oral antibiotics.
- For the past year he has been troubled by an “upset stomach”. On further probing, he describe intermittent abdominal cramps, frequent diarrhea; and, on 2 occasions, blood in the stool

Question #5

Exam:

- T 100.2°F
- Abdo pain to palpation
- Skin lesion

Labs:

- WBC 11,150 (2% eos)
- ESR=79, CRP=110
- BMP normal
- Chest x-ray normal



Question #5

Which one of the following is the most likely diagnosis?

- A. Ulcerative colitis
- B. Cutaneous leishmaniasis
- C. Amebic colitis
- D. Cutaneous blastomycosis
- E. Squamous cell cancer

Question #5

Which one of the following is the most likely diagnosis?

- A. **Ulcerative colitis***
- B. Cutaneous leishmaniasis
- C. Amebic colitis
- D. Cutaneous blastomycosis
- E. Squamous cell cancer

Pyoderma gangrenosum

- **Another** neutrophilic dermatosis
 - Indolent, fever rare (vs Sweet)
- Papule starts at site of often trivial trauma, progressing to a **painful** ulcer with violaceous border and necrotic base
- > 50% of cases occur with systemic illness (but may precede dx, or occur independent of flares)
 - **IBD** (Ulcerative colitis > Crohn's)
 - Inflammatory arthritis
 - Solid organ or heme malignancy

Pyoderma gangrenosum

- Buzzwords & Hooks
 - Minor trauma (**Pathergy**) frequent
 - Painful, progressive **undermined ulcer** with violaceous **edges** and **necrotic base**
 - Associated with **IBD**, arthritis, neoplasm



Question #6

- A 79-year-old woman is seen for 3 weeks of headache, fever and fatigue
- One week earlier she developed jaw discomfort when chewing food and had a brief episode of double vision
- One month ago, she attended a luau and ate roast suckling pork prepared over an open fire



Question #6

- Exam:
 - T 102.2°F, P 104, BP 124/84
 - Slight tenderness over left scalp
 - Rest of exam normal
- Labs:
 - Hb 9.8; WBC 9800, normal diff
 - UA normal
 - Basic metabolic panel normal
 - Sedimentation rate 147

Question #6

Which of the following is most likely to be diagnostic?

- A. Temporal artery ultrasound
- B. Taenia solium serology
- C. TTE
- D. CT angiography
- E. Temporal artery biopsy

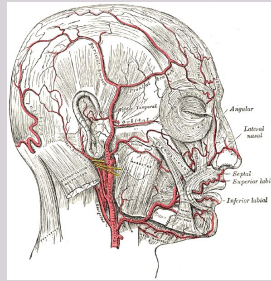
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Which of the following is most likely to be diagnostic?

- A. Temporal artery ultrasound
- B. Taenia solium serology
- C. TTE
- D. CT angiography
- E. Temporal artery biopsy*

Giant Cell Arteritis

- Extracranial branches of the carotid
- Criteria (ACR 1990): 3/5 sensi 93%
 - Age >50 years
 - New onset HA
 - Temporal artery tenderness/dec pulse
 - ESR >50 mm/h
 - Abnl temporal biopsy (vasculitis)
 - > 2 cm with multiple sections reviewed
 - Steroids <2 weeks do not reduce yield and may be sight-saving
- TA u/s, PET, or CT angiography alternative options if biopsy not diagnostic



SEE ENLARGED SLIDE FOLLOWING ENTIRE PRESENTATION

Giant Cell Arteritis

Buzzwords & Associations:

FUO in a patient >50 years PLUS

- Scalp or TA tenderness
- Visual symptoms (diplopia or transient visual loss)
- Jaw or tongue fatigue or pain while chewing
- ESR >100

Empiric steroids indicated when suspicion is high



Overlap of GCA and PMR

- ~50% patients with GCA have concomitant PMR
- Consider GCA in febrile patient with Buzzwords for PMR...
 - Morning stiffness in proximal muscles of shoulder and hip girdle
 - Gel phenomenon (stiffness with inactivity)



Takayasu Arteritis

- Large vessel vasculitis
 - Aorta, carotids and pulmonary arteries
- Buzzwords and associations:
 - Young woman (>80%), Asian ancestry
 - Subacute onset of fever, weight loss, arthralgias and myalgias
 - Carotidynia (pain with palpation), decreased pulses
 - Extremity claudication; visual changes; TIAs
- Dx: Angiography



Question #7

- 37-year-old female presents with fever and joint pain. She is a long-distance runner and in excellent health.
- Three weeks prior she noted R knee pain after a long run. She was treated with a steroid injection with transient improvement but subsequently developed bilateral ankle pain and redness. She notes subjective chills and sweats.
- She recalls several tick bites in the last 2 months.

Question #7

- Exam:
 - T 100.5°F; Pulse 72; BP 110/70
 - Bilateral synovial thickening of ankles with warmth and tenderness to passive movement
 - Skin exam with painful pre-tibial nodules
- Labs:
 - WBC 8.8 (76% segs)
 - CRP=167
 - Uric acid=4.4
 - RF <15, Anti-CCP Ab negative



Question #7

Which of the following is most likely to be diagnostic?

- A. Chest x-ray
- B. Serology for *Borrelia burgdorferi*
- C. Urine Histoplasma antigen
- D. Arthrocentesis
- E. Skin biopsy

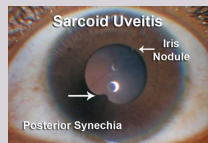
Question #7

Which of the following is most likely to be diagnostic?

- A. Chest x-ray***
- B. Serology for *Borrelia burgdorferi*
- C. Urine Histoplasma antigen
- D. Arthrocentesis
- E. Skin biopsy

Sarcoidosis

- Extra-pulmonary disease in ~1/3 of cases
- Lofgren Syndrome
 - Only form of sarcoid that is a clinical diagnosis
 - Triad of hilar LAN, acute arthritis, EN
 - Women, ankles (>90%), fevers common
- BUZZWORDS
 - Hilar LAN, EN, uveitis, parotid enlargement
 - Non-caseating granulomas
 - Aseptic meningitis with basilar enhancement



Erythema nodosum

- NO cause >50% of cases
- Drugs: sulfonamides, penicillins
- Oral contraceptives
- Sarcoid (Lofgren's syndrome)
- Ulcerative colitis (or Crohn's or Bechets)
- Microbes:
 - Streptococci (16% EN cases), Bartonella, TB
 - Endemic fungi
 - EBV, Hep B/C, covid-19



Erythema nodosum

- **NO** cause >50% of cases
- **D**rugs: sulfonamides, penicillins
- **O**ral contraceptives
- **S**arcoid (Lofgren's syndrome)
- **U**lcerative colitis (or Crohn's or Bechets)
- **M**icrobes:
 - Streptococci (16% EN cases), Bartonella, TB
 - Endemic fungi
 - EBV, Hep B/C, covid-19



Question #8

- A 19-year-old Iraqi immigrant is admitted with 2-days of fever and abdominal pain.
- He has had similar episodes on at least 3 previous occasions over the past 7 years. At the first episode he underwent appendectomy; the appendix path was normal. Subsequent episodes resolved spontaneously after 2-3 days.
- Exam:
 - T 102.2°F; pulse 114; no rash
 - Abdominal guarding, rebound tenderness, hypoactive bowel sounds
- Labs:
 - WBC 16,650; UA normal
 - BMP & LFTs normal
 - No occult blood in stool
 - CT of abdomen and pelvis normal

Question #8

Which of the following is most likely to be diagnostic?

- A. Family history
- B. Colonoscopy
- C. Genetic testing
- D. Angiography
- E. ANCA blood test

Question #8

Which of the following is most likely to be diagnostic?

- A. Family history
- B. Colonoscopy
- C. Genetic testing*
- D. Angiography
- E. ANCA blood test

Familial Mediterranean Fever

- Auto-inflammatory disease causing a periodic fever syndrome
 - Others: PFAPA, TRAPS, hyperimmunoglobulin D
- **Recurrent** attacks (1-3 days) of **fever & serositis** (peritonitis, pleuritis, pericarditis, arthritis)
- Mediterranean ancestry
- Dx:
 - Biallelic mutation of MEFV gene
 - Suggestive: response to colchicine
 - FH insensitive (AR disease)



FMF Buzz Words

- **Mediterranean ancestry**
 - Southern Europe (Greece, Italy, Spain, Balkans)
 - Turkey, Middle East
 - North Africa
- **Periodic, self-limited fevers**
- **Serositis**



Question #9

- A 26-year-old medical student presents with fever and cervical adenopathy.
- She was completely well until 9 days ago when she had the acute onset of fever and vague neck discomfort. She had no sore throat and no dental or scalp problems.



Question #9

- Exam:
 - T 101.4°F; unilateral anterior and posterior cervical enlarged lymph nodes, firm, and mildly tender. Otherwise, unremarkable.
- Labs:
 - Hb 13.9; WBC 4,900 (9% atypical lymphocytes)
 - Basic metabolic panel normal
 - Chest x-ray normal
 - ESR=72
 - Monospot: Negative

Question #9

- Serologic studies:
 - EBV IgM negative
 - HIV negative
 - CMV, Toxo, Bartonella negative
 - RF, ANA, ds-DNA negative
- Lymph node biopsy is performed; Stains for AFB and fungi negative

Question #9

Which of the following is a lymph node biopsy is most likely to show?

- A. Stellate necrosis
- B. Multinucleated giant cells
- C. Non-necrotizing granulomas
- D. Necrotizing histiocytosis
- E. Reed-Sternberg cells

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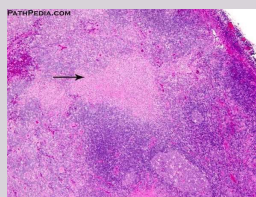
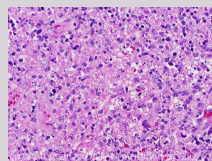
- A. Stellate necrosis
- B. Multinucleated giant cells
- C. Non-necrotizing granulomas
- D. Necrotizing histiocytosis***
- E. Reed-Sternberg cells

Kikuchi Disease

- AKA acute necrotizing histiocytic lymphadenitis
- Self-limited condition of unknown cause
- Typically occurs in **young women**
- May follow infection (EBV, COVID-19)
- Fever & **cervical LAN** (esp posterior, usually unilateral)
- Rarely: morbilliform rash, diffuse LAN, aseptic meningitis, uveitis
- Leukopenia & **atypical lymphocytes** in 25% of cases

Kikuchi Disease

- Diagnosis by pathology:
 - **Necrotizing histiocytic infiltrate** (not neutrophils) and fragments of nuclear debris
- Buzzwords and associations:
 - **Cervical adenopathy in young woman**
 - **Atypical lymphocytes** (mono-like syndrome)
 - Path: necrotizing adenitis with histiocytosis

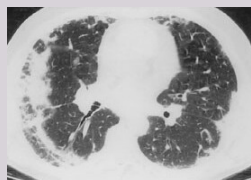


Question #10

- A 41-year-old woman is seen for fever, worsening respiratory symptoms, and a rash.
- She has long-standing asthma with frequent exacerbations.
- She uses an inhaler several times a day and was recently placed on a leukotriene receptor antagonist. She is being tapered off steroids which she has taken for several months.

Question #10

- Exam: Temp 101.5°F; RR 24
- Diffuse wheezing; palpable purpura with nodules on elbows and legs
- Labs: WBC 15,230 (22% eosinophils)
- CT scan: bilateral peripheral infiltrates
- Skin nodule biopsy: granulomas



Question #10

Which one of the following is the most likely diagnosis?

- A. Strongyloidiasis
- B. Allergic reaction to Montelukast
- C. Sarcoidosis
- D. Allergic bronchopulmonary aspergillosis
- E. Eosinophilic granulomatosis with polyangiitis

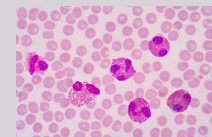
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- C. Sarcoidosis
- D. Allergic bronchopulmonary aspergillosis
- E. Eosinophilic granulomatosis with polyangiitis*

EGPA (Churg-Strauss)

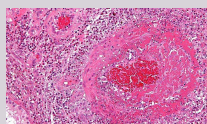
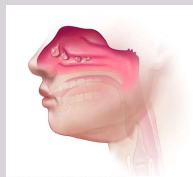
- Eosinophilic granulomatosis with polyangiitis
- Small vessel vasculitis associated with allergic rhinitis, asthma, peripheral and lung eosinophilia
- Most often involves lung and skin, but can involve heart, GI tract, and nervous system
- Labs may be helpful here
 - Eos usually >10%
 - IgE often elevated
 - P-ANCA positive in only 30-40% of EGPA patients
- Tapering of steroids often “unmasks” EGPA



EGPA

2022 ACR/EULAR Classification: >6 points c/w diagnosis

Criterion	Points
Eosinophilia ($\geq 1 \times 10^9/L$)	5
Obstructive lung disease	3
Nasal polyps	3
Extravascular eosinophilic inflammation	2
Mononeuritis multiplex/motor neuropathy	1
Negative points:	
• Pos c-ANCA or anti-proteinase 3	Minus 3
• Hematuria	Minus 1



EGPA

• Buzzwords and associations:

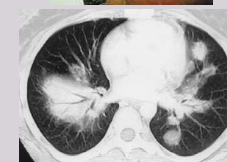
- Longstanding **asthma/COLD**
- New infiltrates and **eosinophilia** (>10%) as **steroids tapered**
- **Nasal polyps/allergic rhinitis**
- **Rash** (tender nodules on extensor surfaces with purpura, ecchymosis, or necrosis))
- Fever UNCOMMON (until late)

Question #11

- A 38-year-old man is seen for a 6-week history of cough, intermittent fever and night sweats
- He has had nasal stuffiness for 4-5 months with occasional epistaxis
- He lives in Philadelphia, and 6 months ago traveled to Cincinnati on business
- He has no pets and takes only an OTC decongestant. He admits to use of intranasal cocaine in last 2 months

Question #11

- Exam:
 - T 100.2°F; RR 18;
 - Nasal deformity with perforation of septum
 - Lungs clear; rest of exam normal
- Labs:
 - WBC 6,900 with normal differential
 - UA 30-50 RBC; BMP normal
 - Chest CT: bilateral nodules with cavitation



Question #11

Which of the following will most likely support the diagnosis?

- A. c-ANCA
- B. Anti-glomerular basement membrane Ab
- C. Urine toxicology screen
- D. Angiotensin converting enzyme (ACE)
- E. Pulmonary angiogram

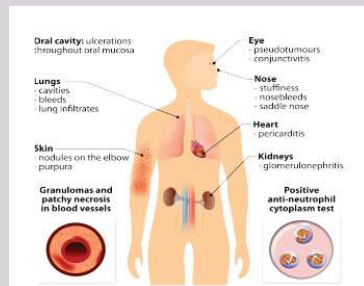
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Granulomatosis with Polyangiitis (GPA)

- Systemic vasculitis of medium and small arteries
- Primarily involves upper and lower respiratory tracts and **kidneys**
- Variably involves joints, cartilage, eyes, skin, and nervous system



SEE ENLARGED SLIDE FOLLOWING ENTIRE PRESENTATION

2022 ACR/EULAR Criteria for GPA

Criterion	Points
Pos c-ANCA or anti-proteinase 3	5
Nasal findings • symptoms (drainage, congestion) • Imaging (consolidations, inflammation)	3 1
Cartilaginous abnl (nasal or auricular)	2
Pulmonary nodules or cavities	2
Granulomas or giant cells on bx	2
Pauci-immune glomerulonephritis	1
Negative points: • Eosinophilia • P-ANCA or anti-myeloperoxidase positivity	Minus 4 Minus 1

≥5 diagnostic

SEE ENLARGED SLIDE FOLLOWING ENTIRE PRESENTATION

GPA vs EGPA: Pearls

Feature	GPA	EGPA
Pulmonary	Nodules, cavitation, infiltrate	h/o Asthma, infiltrates
Sinuses	Chronic sinusitis/destruction	Chronic sinusitis, polyps
Kidneys	Pauci-immune GN	Rare
CNS	Rare	Neuropathy
Eos	No	Yes (peripheral and tissue)
ANCA	C-ANCA/PR3 positive (85%)	P-ANCA (<40%)

Question #12

- A 42-year-old man is seen for his third episode of cellulitis of the external ear.
- Two previous episodes involving the same ear, 2 and 5 months ago, responded very slowly to antibiotics.
- He has a several year history of chronic nasal stuffiness and had an episode of knee arthritis in the past year but is otherwise well.

Question #12

• Exam:

- Afebrile
- Left auricle is inflamed and tender, ear lobe is spared
- He has a saddle-nose deformity; the nasal mucosa is normal
- Labs: CBC normal



Question #12

Which of the following is most likely to be diagnostic?

- A. ANA
- B. Comprehensive H&P
- C. ACE level
- D. CRP
- E. Rheumatoid factor

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Relapsing Polychondritis (RPC)

- Immune-mediated condition
- Inflammation of cartilaginous structures, particularly ears, but also nose, eyes, joints, and airways
- Clinical diagnosis



RPC Diagnostic Criteria

Criteria

- Bilateral auricular chondritis
- Seronegative polyarthritides
- Nasal chondritis
- Ocular inflammation
- Upper respiratory chondritis
- Cochlear and/or vestibular dysfunction

Scores

- 3 clinical criteria
- OR
- 1 clinical criteria plus histopath
- OR
- Chondritis at 2 or more locations responding to steroids and/or dapsone

Saddle-nose Deformity

- Collapse of nasal bridge
 - GPA
 - Relapsing polychondritis
 - Lepromatous leprosy
 - Congenital syphilis
 - Cocaine use
 - Trauma



Relapsing Polychondritis

- Buzzwords and associations:
 - Recurrent “cellulitis” (**cartilage inflammation**)
 - Saddle-nose
 - Cauliflower ear
 - Sparing of ear lobe
 - Parasternal joint involvement



Question #13

- A 29-year-old woman presents following a T-C seizure
- Her family notes that she has been c/o headache and increasingly paranoid in the last 2 weeks
- On exam, she is afebrile but confused. Note is made of involuntary twitching of both upper extremities
- Non-contrast head CT is unrevealing
- LP shows a CSF WBC of 22 (50% lymphocytes)

Question #13

Which of the following is most likely to impact management?

- A. Contrasted MRI of the brain
- B. CSF ACE level
- C. Anti-thyroid antibodies
- D. Meningo-encephalitis PCR panel
- E. CT of the abdo-pelvis

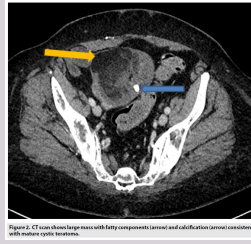
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Anti-NMDAR Encephalitis

- Autoimmune encephalitis
- Epidemiology:
 - Median age 21 years (children/young adults predominate)
 - **Women>>Men** (80% of premenopausal adults are female)
- Triggers
 - **Ovarian teratomas** (present in ~50% of women age 12-44 years)
 - HSV-1 encephalitis (para-infectious)



Anti-NMDAR Encephalitis

- Clinical presentation
 - **Psychiatric symptoms** in ~90% of teens/adults
 - **Mutism or aphasia**, pressured speech
 - **Movement disorders**
 - Autonomic instability
 - Seizures
- Treatment
 - Immunomodulators
 - PLEX
 - **Teratoma removal!**



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