Noon Report

“Two is Better Than One”
Boys Like Girls ft Taylor Swift

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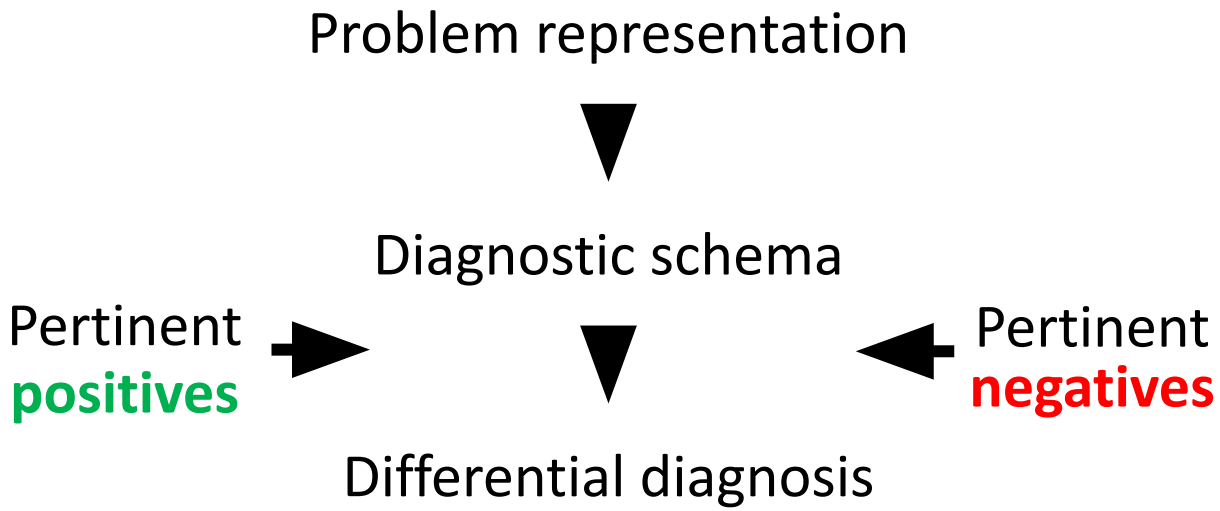
41 y.o. male presents to the ED with joint pain.



Diagnostic schema for joint pain

1. **Autoimmune**
   a. **Seropositive arthritides**: Rheumatoid, SLE, Sjogren's, mixed connective tissue disease
   b. **Seronegative arthritides ("PEAR"):** Psoriatic, enteric, ankylosing spondylitis, reactive
2. **Crystals:** Gout, CPPD
3. **Infection-related:** Bacterial, mycobacterial, viral, etc.
4. **Degenerative:** Osteoarthritis
5. **Hemarthrosis**: Spontaneous vs traumatic
6. **Periarticular**: Tendonitis, bursitis, fibromyalgia, etc.
7. **Potpourri:** Sarcoid, Still's, vasculitis, paraneoplastic, etc.

Features that help narrow your differential:

- Patient demographics
- Number and distribution of affected joints
- Acuity of pain
- Inflammatory vs. non-inflammatory joint pain

e.g. 41 year old male with **acute onset monoarticular inflammatory** joint pain. DDx:

1. Infection-related
2. Crystals
3. Traumatic
4. Seronegative arthritis

Acute pericarditis

Diagnosis:

≥ 2 of the following

1. Hx: Characteristic chest pain
2. P/E: Friction rub
3. ECG: STe and PRd (early)
4. TTE: Pericardial effusion

Etiology: Idiopathic/post-viral (80%), infectious, auto-immune, other…

Management:

- NSAIDs, colchicine (evidence primarily for idiopathic/post-viral)
- Avoid prednisone (increases recurrent risk), but may consider if contraindications/non-responsive to NSAID/colchicine, immune-mediated etiology
- Rule out tamponade if effusion is present (manage accordingly)

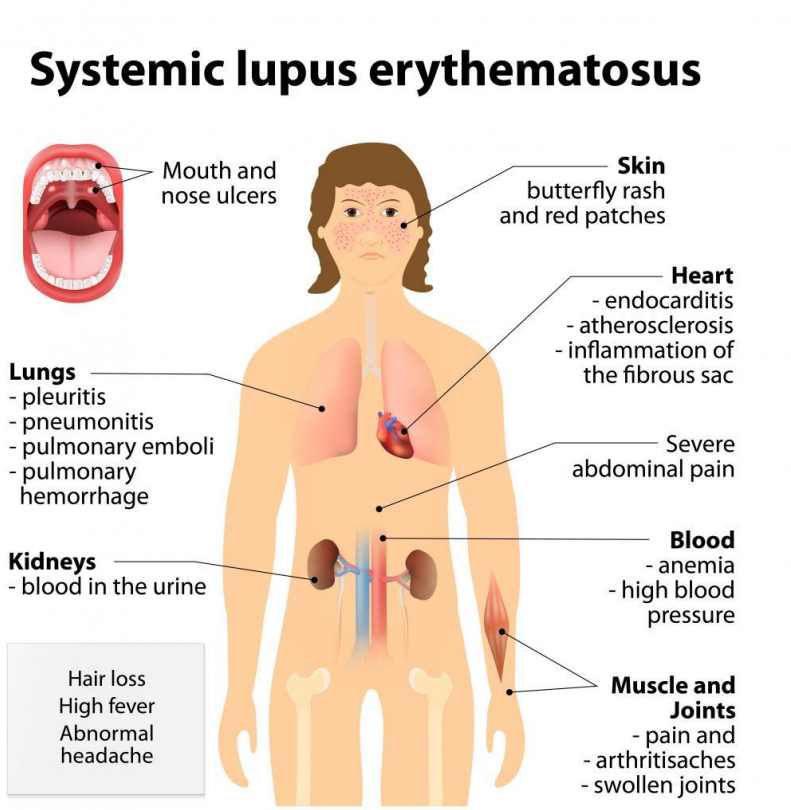
Systemic lupus erythematosus (SLE)

- Chronic autoimmune disease of unknown etiology, that can affect virtually any organ of the body, leading to extremely variable signs and symptoms

- It is classified as a “seropositive connective tissue disorder” and thus generally **requires the presence of antinuclear antibodies** (ANA positive)

- Serologic testing
  - ANA is Sn (>95%) but not Sp
  - Anti-dsDNA is Sp (>95%) but not Sn
  - May see other ENAs (e.g. anti-Sm, anti-histone, etc.)

- The diagnosis of SLE is based on clinical and laboratory findings after excluding alternative diagnoses. Classification criteria based on 2019 ACR/EULAR guidelines.



New EULAR/ACR criteria for the classification of SLE

Clinical domains	Points
Constitutional domain	
Fever	2
Cutaneous domain	
Non-scarring alopecia	2
Oral ulcers	2
Subacute cutaneous or discoid lupus	4
Acute cutaneous lupus	6
Arthritis domain	
Synovitis or tenderness in at least 2 joints	6
Neurologic domain	
Delirium	2
Psychosis	3
Seizure	5
Serositis domain	
Pleural or pericardial effusion	5
Acute pericarditis	6
Hematologic domain	
Leukopenia	3
Thrombocytopenia	4
Autoimmune hemolysis	4
Renal domain	
Proteinuria > 0.5 g/24 hr	4
Class II or V lupus nephritis	8
Class III or IV lupus nephritis	10

Immunologic domains	Points
Antiphospholipid antibody domain	
Anticardiolipin IgG > 40 GPL *or* anti-β2GP1 IgG > 40 units *or* lupus anticoagulant	2
Complement proteins domain	
Low C3 *or* low C4	3
Low C3 *and* low C4	4
Highly specific antibodies domain	
Anti-dsDNA antibody	6
Anti-Sm antibody	6

REFERENCE: Aringer et al. Abstract #2928. 2018 ACR/ARHP Annual Meeting

- ✓ **Classification criteria are not diagnosis criteria**
- ✓ All patients classified as having SLE must have ANA ≥ 1:80 (entry criterion)
- ✓ Patients must have ≥ 10 points to be classified as SLE
- ✓ Items can only be counted for classification if there is no more likely cause
- ✓ Only the highest criterion in a given domain counts
- ✓ SLE classification requires points from at least one clinical domain

@Lupusreference

- Entry Criterion: **ANA titre ≥ 1:80**
- Weight score requires **≥ 1 clinical criteria** AND **≥ 10 points**

SLE management & monitoring

- **Hydroxychloroquine** is the backbone of therapy for all patients with SLE
- Additional treatment is influenced by whether there is renal involvement
  - *Lupus nephritis*: Determined by lupus nephritis class, usually a combination of immunosuppressive agents.
  - *Non-renal SLE*: Determined by severity of symptoms. May add a limited course of glucocorticoids. If worsening, add other immunosuppressive agents (methotrexate, azathioprine, mycophenolate, cyclophosphamide)
- For all patients: Vaccinations, diet/exercise/smoking, BP/lipid/glucose control
- Monitoring
  - Clinical assessment
  - Anti-dsDNA & serum complements (C3, C4) can be used to monitor disease activity during a flare. ANA is not helpful (do not repeat over time)

Diagnostic schema for oral ulcers

(for the Internist, high yield!)

Note: Aphthous ulcer (i.e. Canker sore) is a dermatological description of the type of oral lesion. It can occur idiopathically (most common) or in association with other systemic diseases.

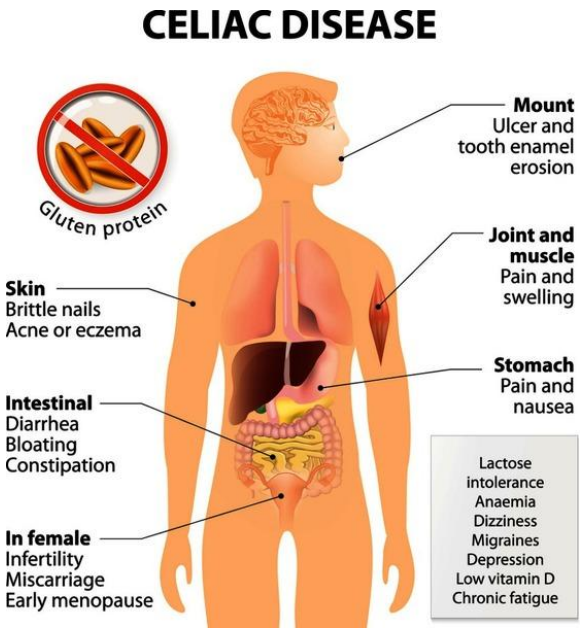
1. **Idiopathic/mechanical**
2. **Infectious** – HSV, VZV, HIV, Syphilis, Coxsackievirus
3. **Immune-mediated**
   - Rheumatological – SLE, Behcet's, SVVs
   - GI-associated – IBD, celiac
   - Dermatological – Mucous membrane pemphigoid, bullous pemphigoid, pemphigus vulgaris, etc… (hi Derm!)
4. **Drug-associated**
   - Reaction – EM, SJS/TEN
   - Side effect – chemotherapeutics, methotrexate

What features help narrow your differential?

- Acute vs chronic
- Painful vs painless
- Associated signs/symptoms
- Risk factors / exposures

Celiac disease

- Immune-mediated enteropathy due to gluten hypersensitivity
- Classically presents with GI symptoms (diarrhea, bloating), but can see **a variety of systemic features**, including rash (dermatitis herpetiform), liver enzyme elevation, oral ulcers, etc.
- Can lead to micronutrient deficiency (vitamin B12, D, Fe, Ca)
- Diagnosis
  - Serology with anti-TTG and IgA levels (other tests if IgA low or specific population at risk)
  - If positive serology or high risk, proceed with upper endoscopy & duodenal biopsies for diagnosis (Marsh Classification)
  - The above testing must be done while the patient is on **a gluten-containing diet** (ideally 3 slices of wheat bread daily for 1 – 3 months)
- Treatment: Gluten-free diet
- Monitor disease activity with serology. If ongoing symptoms despite negative serology, consider repeat biopsy



Atypical celiac disease

- Celiac disease **without clinically apparent gastrointestinal symptoms**, presenting with extraluminal manifestations
- The true prevalence is unknown but is thought to be substantially underrecognized
- The diagnosis is made **the same way as typical celiac disease** (anti-TTG serology, duodenal biopsy)
- There is no current evidence to recommend screening in asymptomatic individuals, but celiac disease should be considered when presenting with extraluminal or systemic complaints