Presentation of Chest Pain with Uncommon Diagnosis

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Introduction:

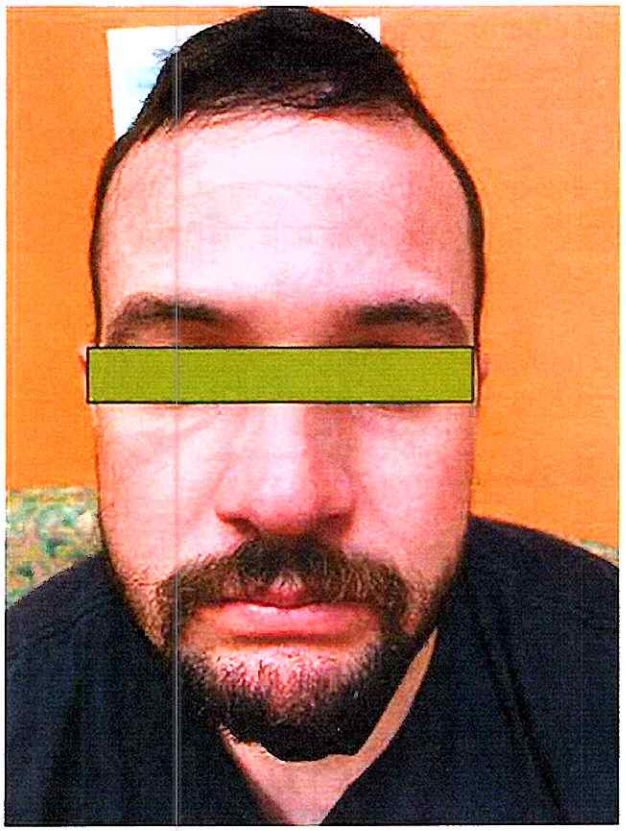
- Adult Onset Still Disease (AOSD) is a **rare multisystem inflammatory** disease of **unknown etiology.**
- Clinical presentation is highly variable. Characteristic **clinical triad** include fever, arthralgia and/or arthritis, and evanescent salmon-colored rash.
- We are presenting an interesting case of AOSD with an unusual presentation.

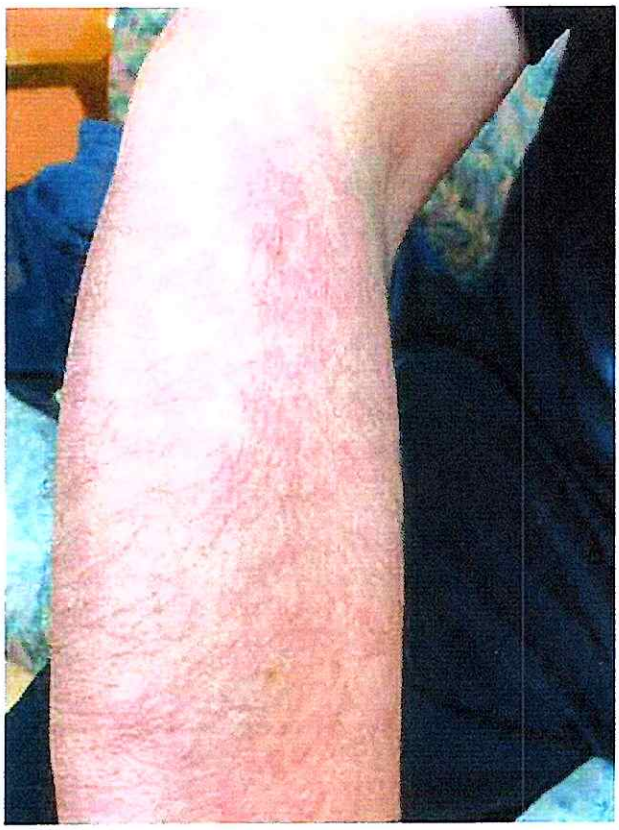
Case presentation:

- A 37-year-old gentleman, no significant PMHx
- ***At time of presentation***
  - Symptoms: left-sided pleuritic chest pain, mild dry cough, and feeling feverish. The physical exam was unremarkable.
  - Labs: Basic labs including troponin were unremarkable, respiratory viral panel negative.
  - ECG: subtle diffuse ST-segment abnormalities with normal sinus rhythm.
  - X-ray: unremarkable, CTPA: mildly enlarged left hilar LN.
  - Exercises stress echo: negative for ischemia or pericardial effusion.
  - **Patient managed symptomatically with NSAIDs.**
- ***Five days later***
  - Presented again with chest pain, fever (reaching up to 102.5 F), arthralgia, and new non-scaly maculopapular skin rash over face and elbows.
  - Labs: neutrophilic leukocytosis, normocytic anemia, high CRP, and mildly elevated troponin of 77 ng/L which trended down.
  - ECG: normal sinus rhythm otherwise unremarkable.

- **Suspected viral myopericarditis, treated with colchicine and high dose ibuprofen.**
- Cardiac MRI was negative for myopericarditis. But showed enlarged infra-hilar lymph node and mild splenomegaly with bilateral minimal pleural effusions.
- Labs: negative workup for HIV, QuantiFERON TB, HBV, HCV, syphilis, Group A strep, CMV, EBV, and parvovirus B19.
- Autoimmune workup including ANA, RF, anti-CCP and ANCA was unremarkable.
- Ferritin: >12500ng/mL (upper limit is 400).
- **Patient was diagnosed with Adult-Onset Still disease and was started on prednisolone 0.5mg/kg daily.**

- ***Two months later***
  - The patient continued to have fever.
  - Pan CT showed lymphadenopathy with sizable left axillary lymph node.
  - Biopsy was negative for TB, fungal infection, and lymphoma.
  - He was started on Anakinra.
- ***One-year follow up.***
  - Marked improvement of symptoms and remained in remission for at least a year.





Clinical Significance:

- The two widely accepted criteria for AOSD have their limitation.
- The **Yamaguchi criteria** has the highest sensitivity (96.2%), but it requires exclusion of other differential diagnoses.
- The **Fautrel criteria** has the highest specificity (98.5%), but glycosylated ferritin is not widely available in most hospital systems.
- **A glycosylated ferritin** level < 20% is a better diagnostic marker for AOSD than elevated serum ferritin alone. Combining both abnormalities can be especially helpful for differential diagnosis.

Discussion:

- **The diagnostic challenge of AOSD arises from the rarity of the disease, its heterogeneity of clinical presentation and the lack of specific diagnostic tests, often resulting in delayed diagnosis.**
- The median interval between onset of symptoms and a definite diagnosis of AOSD ranged between 1 and 4.1 months across studies.

- More studies demonstrated a major role of inflammatory cytokines, such as IL-1, IL-6, IL-18, and IL-37, and other biomarkers in the diagnosis and management of AOSD.
- Further research is essential to identify at-risk groups, update diagnostic criteria, and explore the use of other biomarkers, to expedite the diagnosis and improve outcomes for individuals with AOSD.

1992 Yamaguchi criteria*		2002 Fautrel criteria	
Two major criteria and at least five total criteria		Four or more major criteria or three major criteria + two minor criteria	
Major criteria	**Minor criteria**	**Major criteria**	**Minor criteria**
Fever ≥39°C, ≥1 week	Sore throat	Spiking fever ≥39°C	Maculopapular rash
Arthralgias or arthritis ≥2 weeks	Lymphadenopathy	Arthralgia	Leukocytes ≥10,000/mm³
Typical rash	Hepatomegaly or splenomegaly	Transient erythema	
Leukocytosis ≥10,000/mm³ with ≥80% granulocytes	Abnormal liver function studies	Pharyngitis	
	Negative ANA and RF	PMN ≥80%	
		Glycosylated ferritin ≤20%	

Notes: *Absence of infection, malignancy, or other rheumatologic disorders known to mimic AOSD.
Abbreviations: AOSD, adult-onset Still's disease; ANA, antinuclear antibody; RF, rheumatoid factor; PMN, polymorphonuclear leukocyte.