Challenges in Diagnosis: Adult Onset Still's Disease- a rare systemic inflammatory arthritis

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Initial Presentation

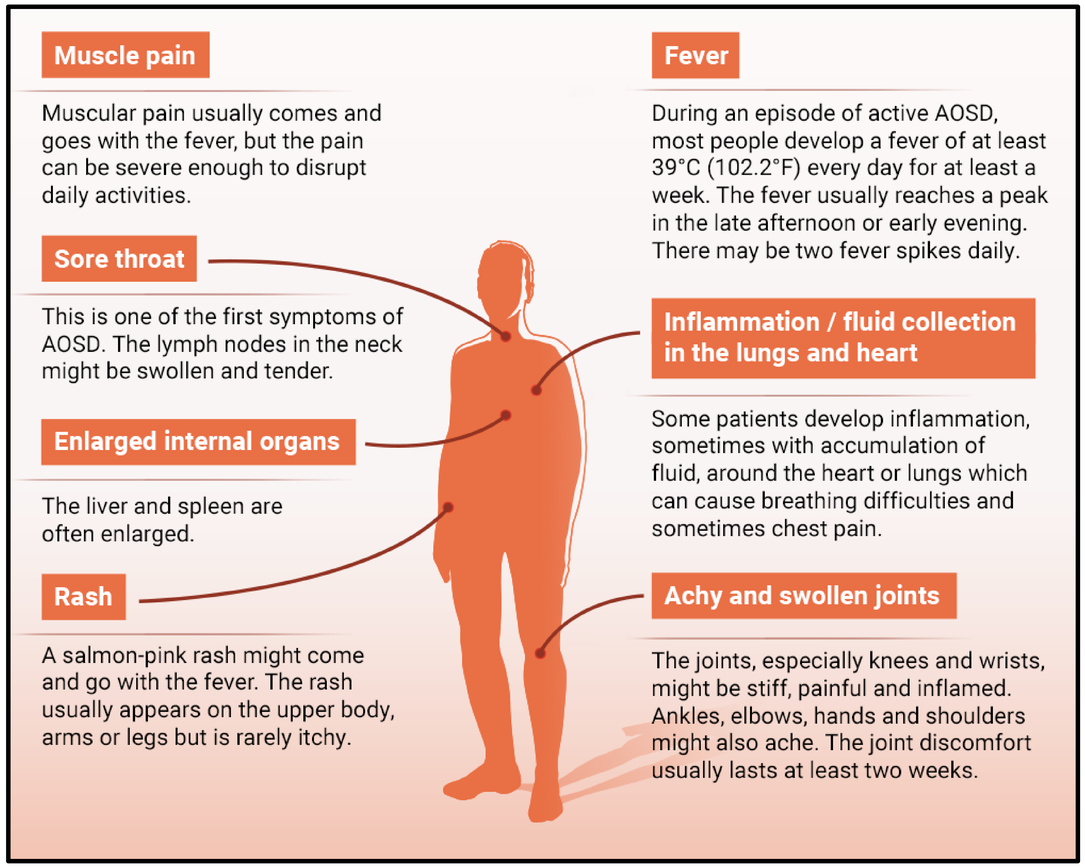
Demographic: 54 year old, Caucasian male.
Chief Complaint: Fatigue, nonproductive cough and fluctuating fevers.
Vitals: Temperature of 102.6 ºF, heart rate of 95 beats/min and blood pressure of 122/81 mmHg, with a respiratory rate of 16 breaths/min and 99% O2 saturation on room air.
Labs: WBC 15,260/ul with 85% Left Shift, C-reactive protein 214 mg/L, Erythrocyte sedimentation rate (ESR) of 84 mm/hr.

Hospital Course

- Treated with Broad Spectrum Antibiotics
- Sepsis workup negative- No Growth Blood Cultures
- Pulse steroids initiated with improvement in fever, cough and stiffness.
- AST/ALT upon discharge being up to 187 U/L and 220 U/L, respectively.
- Discharged with 7 days of antibiotics.

Outpatient Course

One Week Follow Up: recurrence of cough and dyspnea, despite having a normal repeat chest x-ray. He was started on prednisone 20mg for five days.
Two Week Follow Up: Cough improved during the first part of the day, but he developed worsening symptoms in the afternoon and evening. He also started developing a hoarse throat along with shoulder and neck joint stiffness. He admitted receiving a diagnosis of Juvenile Still's Disease during his teenage years, but has been symptom free since the age of 25. Started on high dose steroid regimen of 60mg prednisone a day, with a slow taper until Rheumatology could see him.
Three Week Follow Up: Marked improvement in symptoms, resolution of fevers, and relief of arthralgias. He performed self-guided physical therapy.
Continued Management: Rheumatology reviewed- started on Methotrexate. The patient continues to taper his steroid, slowly and under Rheumatology's close monitoring, and has found himself recovering and regaining functionality.



https://www.periodicfevers.com/thescience/AOSD/

Discussion

Constellation of symptoms recognized as its own specific disease entity among adults in 1971 by E.G. Bywaters for categorization as Adult Onset Still's Disease.[1]
Cases of this disease are rare with 0.16:100,000 effected among the population.[2]
Adult Onset Still's Disease affects men and women likely equally, predominantly among people ages 16-35 (about 75% of all cases), although cases are reported even among people in their 70s.[3]
Classically taught as a triad of symptoms: fevers, joint pain, and a salmon colored rash sparing the face.
Other nonspecific symptoms and signs include fatigue, neutrophilia, swollen lymph nodes, splenomegaly, pericarditis, and pleural effusions.[4]
Diagnosis is one of exclusion. Multiple diagnostic criteria have been developed, but the Yamaguchi Criteria is the most sensitive.[5]
Treatment targets inflammatory conditions - with mainstay starting with NSAIDs for minor presentations vs. steroids for more severe.
Targeting chronic conditions includes the use of methotrexate, anakinra, and other immunomodulatory agents.[6]

Yamaguchi Criteria

Major criteria	Minor Criteria
Fever >39 º Celsius for one week	Sore Throat
Arthralgia for >2 weeks	Lymphadenopathy
Nonpruritic Salmon Rash while Febrile	Hepatosplenomegaly
Leukocytosis >10,000/ul w/PMN predominance	Abnormal Liver Enzymes
	Negative ANA and RF

Conclusion

This case demonstrates the diagnostic challenges and importance of thoroughly evaluating patients who present with acute exacerbations of Adult Onset Still's Disease. Maintaining diagnostic suspicion and reviewing clinical progression is essential to ensuring diagnostic certainty, decreasing patient discomfort, and preserving patient functionality and well-being.

Additional Information

Contact Information

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