International Journal for Multidisciplinary Research (IJFMR)
E-ISSN: 2582-2160 ● Website: www.ijfmr.com ● Email: editor@ijfmr.com

A Case Study Report: Adult-Onset Still's Disease

JP Grace Angeline

RN, RM. MSc N, Associate Professor, College of Nursing CMC Vellore Chittoor campus, Andhra Pradesh India

ABSTRACT:
Background: Adult-onset Still's disease (ASOD) is a rare systemic inflammatory condition, commonly known as systemic juvenile idiopathic arthritis (SJIA). It usually affects young adults in the age group of 16 – 35 years with an unknown etiology, characterized by a triad of symptoms like high-grade fever, arthralgia, and maculopapular salmon-colored skin rash over the trunk or extremities. The diagnosis of ASOD is based on a thorough patient history and clinical examination and is supported by laboratory findings and the exclusion of other diseases. There are no specific guidelines to treat this ASOD until now, however, a targeted treatment approach aims for comprehensive disease control of articular and systemic manifestations and prevention of complications and chronic, irreversible disability.

Keywords: Systemic Inflammatory, Salmon Coloured Rash, Arthralgia, Targeted Therapy

Introduction

Adult-onset Still's Disease (AOSD) is a rare systemic inflammatory condition that usually affects young adults. These disorders are named after a British physician Sir George Fredric Still who first described this disorder in the medical literature in the year 1896 as a childhood arthritis associated with fever. An adult with "Stills disease" was reported in 1971, but due to a lack of clarity in diagnosing and overlapping of symptoms it was reported in the late 1800s. The hallmark features of this ASOD are characterized by a triad of symptoms like high spiking fevers, a characteristic salmon-colored rash, and joint pain, muscle pain. However, to date, there is no definite laboratory or imaging test available for diagnosing AOSD. Since no clear clinical pathway for treating this disorder, the focused history, clinical examination, and related lab investigations help to pivot the healthcare team to provide a comprehensive approach.

Epidemiology

This is a rare multisystem disease. Very few published articles are available to know the prevalence and incidence of the disease. It is reported that worldwide annual incidence of AOSD is estimated to be between 0.16 and 0.62 per 100,000 individuals and it predominantly affects women more than men. Available research shows that 45 – 80% had the disease onset occurrence between the age group of 16 – 35 years[1].

Cause and risk factors

ASOD has an unknown etiology. This is not a hereditary disease but the hypothesis states that it might be caused by genetic factors, exaggerated response to viral or bacterial infection, or exposure to environmental factors. Recent studies state that some people show the symptoms of Stills disease after the COVID-19 vaccination[2].

Viral infections: rubella; measles; mumps; Epstein-Barr virus; hepatitis A, B, or C virus; HIV; cytomegalovirus; parvovirus B19; adenovirus; echovirus; human herpes virus; influenza and parainfluenza viruses; Coxsackie virus

Bacterial infections: Yersinia enterocolitica, Campylobacter jejuni, Chlamydia trachomatis or pneumoniae, Mycoplasma pneumoniae, Borrelia burgdorferi [3] .

Pathophysiology

Innate and Adaptive immunity dysregulation play a vital role in the pathogenesis of Stills disease. The cytotoxic function of natural killer cells is decreased in patients with active AOSD[4]. Interleukin (IL) 18 and 1β, are the two key proinflammatory cytokines processed through the inflammasome machinery factors in the pathogenesis of AOSD.

They cause IL-6 - in skin rash and serum correlating with disease activity

Th1 cytokine secretion- contributes to the complex immunopathology associated with the disease process.

NK cell dysregulation leads to macrophage activation.

Activation of neutrophils and macrophages which recruit neutrophils to the inflammation site leads to the persistence of chronic articular AOSD

Disease patterns

Three major disease patterns have been observed in AOSD patients[5]:

1. **Monocyclic**: The patient may show a single systemic symptom that completely resolves within months.
2. **Polycyclic or Intermittent**: one or more disease flares and characterized by complete remissions that can last up to a couple of years
3. **Chronic articular pattern**: usually associated with long-lasting polyarthritis

Clinical manifestations

Stills disease patients usually present with 3 clinical and one biological manifestations. These are similar to other diseases hence the diagnosis gets delayed in this AOSD[5].

Clinical symptoms: Fever, maculopapular salmon-colored rash, and arthritis or arthralgia.

1. **Fever**: Characterized by sudden rise of temperature ≥39 °C which may be present constantly during the active disease period. Fever may occasionally spike twice daily and temperature rises daily in the evening for more than a week. The temperature can fluctuate by 4 °C within 4 hours [4].
2. **Arthralgia or Arthritis:** Second most common symptoms. All joints including sacroiliac and distal interphalangeal joints. Fusion of the wrist joint and bilateral carpal ankylosis without structural damage of metacarpophalangeal or proximal interphalangeal joints are very suggestive of AOSD 6.
3. **Skin rash:** During the fever, skin rashes are observed on the trunk and proximal extremities, characterized by transient, nonpruritic, salmon-colored, maculopapular lesions. occasionally rashes are seen on the palms and soles**.**

Other common symptoms: sore throat, pharyngitis, lymphadenopathy, and hepatosplenomegaly are the symptoms that can be seen in Stills disease.

Biological symptoms: Increased leukocyte and Neutrophil Counts are predominantly observed in patients with AOSD.

Diagnosis

Like other inflammatory processes, in AOSD during the acute phase, the neutrophil counts and leukocytes are increased. The serum C-reactive protein (CRP) erythrocyte sedimentation rate (ESR), and liver enzymes are increased.

Recent research suggests that marked elevation of serum ferritin (hyperferritinemia) levels is suggestive of AOSD. Also, 20-50% of decreased levels of Glycosylated Ferritin (GF) suggests adult onset of Stills disease. The GF level normally represents more than half of the total ferritin level [6].

Classification Criteria

There are two sets of criteria available now to evaluate stills disease. The Yamaguchi and Fautrel classification criteria are the most widely used criteria for AOSD

A. Yamaguchi et al. classification criteria for AOSD [7] :

For Diagnosis: "5 or more criteria, of which at least 2 should be major"

Major criteria:

1. Fever ≥39 °C lasting at least 1 week
2. Arthralgia or arthritis for ≥2 weeks
3. Typical nonpruritic salmon-pink skin rash
4. Leukocytosis ≥10,000/mm3 with ≥80% polymorphonuclear cells

Minor criteria:

1. Sore throat
2. Lymph node enlargement
3. Hepatomegaly or splenomegaly
4. Abnormal liver function tests
5. Negative ANA and RF tests

Exclusion criteria:

1. Infections (especially, sepsis and infectious mononucleosis)
2. Malignancy (mainly malignant lymphoma)
3. Other rheumatic disorders (mainly polyarteritis nodosa and rheumatoid vasculitis with extraarticular features)

The Fautrel criteria set does not require exclusion criteria and it has the advantage of including ferritin and GF levels as diagnostic biomarkers

B. Fautrel et al. classification criteria for AOSD [8].

For diagnosis: "4 or more major criteria OR 3 major criteria + 2 minor criteria"

Major criteria:

1. Spiking fever ≥39 °C
2. Arthralgia
3. Transient erythema
4. Pharyngitis
5. Polymorphonuclear cells ≥80%
6. Glycosylated ferritin < 20%

Minor criteria:

1. Maculopapular rash
2. Leukocytes ≥10,000/mm3

Management

Symptomatic therapy:

Traditionally the inflammatory and systemic symptoms of AOSD are treated by steroids, NSAIDs, and Disease-Modifying Antirheumatic Drugs (DMARDs). The recent advancement helps reduce the inflammatory cell reactions by the targeted therapy.

Targeted therapy: effective in refractory AOSD [9,10].

a. Anti-interleukin 1 (IL-1Ra and anakinra)
b. Anti-interleukin 6 (tocilizumab)
c. Anti- IL1b (canakinumab)
d. TNF-a inhibitors (infliximab, etanercept, and adalimumab)

Case report:

Background: Mr. X, an 18-year-old BCA student came to the emergency department with a history of high-grade fever associated with chills for the past 10 days and severe body pain and weakness for 7 days. He had no history of vomiting, seizure breathlessness, or any GI Symptoms. He had a sore throat a month ago and was well during the hospital visit without any signs of any respiratory illness.

Past history: He has no specific illness or hospitalization from his childhood. During the COVID–19, he was not infected however he had a dose of injection as a prophylaxis dose in the year 2021. He has no family history of any communicable, non-communicable, or genetic illness.

Physical examination reveals GCS 15/15 thin body built, No signs of pallor icterus or clubbing or noticeable rashes. Temperature: 105.9* Fahrenheit, Pulse 140/mt, Respiratory rate 34 breaths/ mt Bp; 90/60 mmHg. All blood and Urine investigations also, a bone marrow biopsy was done to rule out the disease. He was initially treated for the symptoms of fever and was supported with Noradrenalin for the hypotension. His blood investigation results shown as HB – 13.1 gm%, WBC – 20400mm3, Platelet – 330000mm3; CRP – 106 mg/l; Blood culture – Negative for growth; Liver enzymes, and electrolytes were within normal limits. He was evaluated for Dengue, scrub, Malaria, TB, and Infective endocarditis as a differential diagnosis. Later the blood investigation of Ferritin showed a high value of 15397ng/ml, which is supposed to be 22- 322ng/ml for males. He was referred to rheumatology, and diagnosed to have Adult-onset Stills disease. He was started on Inj Methotrexate corticosteroids and NSAIDs. Targeted therapy of TOCILIZUMAB was given. Although he had the signs of cardiac complication of myocarditis, at discharge he had good improvement in his mobility, and his temperature reduced and was within normal limits.

Nursing care management

1. Assessment Data:

Subjective Data:

a. Reports of severe joint pain
b. Describes pain as sharp and throbbing
c. Pain scale rating: 8/10

Objective Data:

d. Swelling and redness observed in joints
e. Limited range of motion
f. Elevated temperature (fever)
g. Laboratory **results showing elevated inflammatory markers (CRP, ESR)**

Nursing Diagnosis: Acute Pain related to inflammatory processes as evidenced by patient reports of severe joint pain, pain scale rating, and physical signs of inflammation.

Expected Outcomes:

The patient will report a reduction in pain to a manageable level (3/10) within 48 hours and exhibit improved joint mobility and decreased inflammation.

Nursing Interventions and Rationale:

Pain Assessment and Monitoring

- Assess the pain location, intensity, aggravation factors, duration of pain, and type of pain
- Observe for noncommunication clues of pain. Eg. facial expression
- Use the standardized pain scale to measure the subjectivity of pain every 4 hourly
- Document the findings every 4 hours to evaluate the effectiveness of pain management and provide a modified nursing care plan

Positioning and Mobility

- Position the client to ensure comfort and support affected joints with pillows or foam supports to alleviate pressure on painful joints and reduce pain.
- Encourage to perform gentle range-of-motion exercises as tolerated to prevent stiffness and improve joint function.

Pharmacological Management

- Administer prescribed nonsteroidal anti-inflammatory drugs (NSAIDs) or corticosteroids as ordered to reduce inflammation and pain in AOSD
- Reassess the pain after 20 minutes to evaluate the effectiveness of the intervention
- Reassure the client
  Non-Pharmacological Pain Relief Measures
- Apply cold packs to affected joints for 15-20 minutes, several times a day to reduce swelling and numb the area, providing pain relief.
- Encourage the patient to perform relaxation techniques. Eg., deep breathing exercises or guided imagery to reduce muscle tension and anxiety
- Engage the patient in guided imaginary technique in their favorite or preferred topics

Patient Education

- Educate the patient about the importance of adhering to the medication regimen and the potential side effects of prescribed medications to enhance compliance and help in the early identification of adverse effects.
- Teach the patient about recognizing early signs of a flare-up and the importance of seeking timely medical intervention to prevent severe pain episodes.

Emotional Support

- Provide emotional support and encourage the patient to express feelings and concerns about their condition to reduce anxiety and improve overall well-being.

Evaluation:

The patient reports a reduction in pain to 3/10 or less within 48 hours, demonstrates effectiveness in using non-pharmacological pain relief techniques, and shows improved joint mobility and decreased signs of inflammation.

2. Assessment Data:

Subjective Data:

a. Reports of severe joint pain
b. Describes pain as sharp and throbbing
c. Pain scale rating: 8/10

Objective Data:

d. Swelling and redness observed in joints
e. Limited range of motion
f. Elevated temperature (fever)
g. Laboratory results showing elevated inflammatory markers (CRP, ESR)

Nursing Diagnosis: Hyperthermia related to the inflammatory process of Adult-Onset Still's Disease as evidenced by elevated body temperature, flushed skin, and patient reports of chills.

Expected Outcomes:

The patient will maintain a body temperature within normal limits (36.5°C - 37.5°C) within 48 hours.
The patient will verbalize understanding of fever management strategies.
The patient will demonstrate effective use of cooling measures.

Nursing Interventions and Rationale:

Temperature Monitoring

- Monitor and document the patient's temperature every 2-4 hours to assess the effectiveness of interventions.
  Pharmacological Management
- Administer antipyretic medications such as acetaminophen or NSAIDs as prescribed to help lower the body temperature.

Hydration

- Encourage the patient to take more fluid (e.g., water, electrolyte solutions) to prevent dehydration.
- Administer intravenous fluids as prescribed if oral intake is insufficient to maintain fluid balance.

Environmental Adjustments

- Make sure the room temperature to a comfortable level (around 20-22°C or 68-72°F).
- Provide lightweight clothing and linens to reduce insulation aid in heat dissipation and comfort.
  Cooling Measures
- Apply cool compresses to the patient's temperature of 100 degrees to lower body temperature through conduction.
- Keep a fan to increase air circulation in the patient's room in evaporative cooling, reducing body temperature.

Rest and Activity

- Encourage rest and limit physical activity to conserve energy and prevent excessive heat production.
- Educate the patient about the importance of fluid intake and recognizing signs of dehydration.
- Teach the patient and family about the proper use of antipyretics and when to seek medical attention if the fever persists or worsens.

Evaluation:

Within 48 hours the patient's body temperature is maintained within normal limits (36.5°C - 37.5°C), verbalizes understanding of fever management strategies, demonstrates effective use of cooling measures, and reports feeling more comfortable.

3. Assessment Data:

Subjective Data:

a. Reports feeling self-conscious about skin rashes
b. Expresses concerns about appearance and social interactions

Objective Data:

c. Presence of salmon-colored maculopapular rash
d. Rashes primarily on the trunk, arms, and legs
e. Avoidance of social activities

Nursing Diagnosis: Body Image Disturbance related to visible skin rashes and changes in appearance as evidenced by patient reports of self-consciousness, concerns about appearance, and avoidance of social interactions.

Establish a Therapeutic Relationship

- Establish a trusting relationship with the patient
- Encourage open communication about their feelings and concerns to promote emotional healing.

Assessment of Body Image Perception

- Assess the patient's perception of their body image and the impact of skin rashes on their self-esteem and social interactions.
- Educate the patient about Adult-Onset Still's Disease, including the nature of the skin rashes and potential treatments to reduce their appearance.
- Apply the prescribed skincare routines and products to alleviate the appearance of rashes.
- Encourage the patient to focus on their positive attributes and strengths to shift the attention away from the skin rashes and improve self-esteem.
- Encourage the choice of clothing options that make the patient feel comfortable and confident while minimizing the visibility of rashes.

Support Groups and Counseling

- Allow the patient to ventilate his /her feelings
- Refer similar patients if admitted to the ward
- Refer the patient to support groups or counseling services to enhance emotional well-being.

Coping Strategies

- Teach the patient coping strategies such as relaxation techniques, stress management, and positive self-talk.
- Encourage the patient to participate in social activities and interactions gradually to build confidence and reduce feelings of isolation.
- Assess the patient's psychological status regularly for signs of depression or anxiety.

Evaluation:

The patient verbalizes feelings and concerns about body image within 24 hours, demonstrates positive coping mechanisms to deal with body image disturbances, and engages in social activities and interactions within one week.

Conclusion:

Adult-onset Stills Disease is a rare disease; however, nursing care plays a major role in supportive, preventive, and promotive care. A clear understanding of Still's disease will help the nurses render

effective nursing care during acute care. Meeting the physical, emotional, social, and spiritual support to the patient and the family during this crucial period will promote the quality of life for the patient and reduce the caregiver's burden.

Declaration of patient consent:

After the patient's consent, the details were collected. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References:

1. Efthimiou P, Kontzias A, Hur P, Rodha K, Ramakrishna GS, Nakasato P. Adult-onset Still's disease in focus: Clinical manifestations, diagnosis, treatment, and unmet needs in the era of targeted therapies. Semin Arthritis Rheum. 2021 Aug;51(4):858-874.
2. Magliulo D, Narayan S, Ue F, Boulougoura A, 2. Badlissi F. Adult-onset Still's disease after mRNA COVID-19 vaccine. Lancet Rheumatol. 2021 Oct;3(10):e680-e682. doi: 10.1016/S2665-9913(21)00219-8. Epub 2021 Jul 22. PMID: 34316726; PMCID: PMC8298008.
3. Fautrel B. Adult-onset Still disease. Best Pract Res Clin Rheumatol. 2008;22(5):773–792.
4. Bhargava J, Panginikkod S. Still Disease. [Updated 2024 Feb 26]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK538345
5. Gerfaud-Valentin M, Maucort-Boulch D, Hot A, Iwaz J, Ninet J, Durieu I, Broussolle C, Sève P. Adult-onset still disease: manifestations, treatment, outcome, and prognostic factors in 57 patients. Medicine (Baltimore). 2014 Mar;93(2):91-99
6. Mitrovic S, Feist E, Fautrel B. Adult-Onset Still's Disease. Periodic and Non-Periodic Fevers. 2019 Oct 30:93–132. doi: 10.1007/978-3-030-19055-2_6. PMCID: PMC7123329.
7. Yamaguchi M, Ohta A, Tsunematsu T, Kasukawa R, Mizushima Y, Kashiwagi H, et al. Preliminary criteria for classification of adult Still's disease. J Rheumatol. 1992;19(3):424–430.
8. Fautrel B, Zing E, Golmard J-L, Le Moel G, Bissery A, Rioux C, et al. Proposal for a new set of classification criteria for adult-onset still disease. Medicine (Baltimore) 2002;81(3):194–200.
9. Ortiz-Sanjuán F, Blanco R, Riancho-Zarrabeitia L, Castañeda S, Olivé A, Riveros A, et al. Efficacy of anakinra in refractory adult-onset Still's disease: multicenter study of 41 patients and literature review. Medicine (Baltimore) 2015;94(39):e1554.
10. Barsotti S, Neri R, Iacopetti V, d'Ascanio A, Talarico R, Tripoli A, et al. Successful treatment of refractory adult-onset still disease with canakinumab: a case report. J Clin Rheumatol. 2014;20(2):121.