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ADULT ONSET STILLS DISEASE MIMICKING A RUPTURED HEMORRHAGIC OVARIAN CYST: A CASE REPORT

Medicine

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ABSTRACT

Adult Onset Stills Disease (AOSD) is a rare systemic autoinflammatory disorder characterized by quotidian fever, evanescent salmon-colored rash, arthralgia, and multisystem involvement. Diagnosis remains challenging due to its heterogeneous presentation and lack of specific markers. We present a case of a 45-year-old female who initially presented with acute severe abdominal pain mimicking a ruptured hemorrhagic ovarian cyst. Comprehensive investigations, including markedly elevated serum ferritin (>1000 ng/mL), neutrophilic leukocytosis, and negative infectious and autoimmune workup, supported the diagnosis of AOSD per Yamaguchi criteria. The patient responded favorably to systemic corticosteroid therapy with rapid clinical improvement. This case highlights the importance of considering AOSD in the differential diagnosis of unexplained systemic inflammation, even when presentation mimics acute surgical conditions.

KEYWORDS

Adult-onset Stills Disease, Pyrexia Of Unknown Origin, Serum Ferritin, Corticosteroid Therapy

INTRODUCTION

Adult-Onset Stills Disease (AOSD) is a rare systemic inflammatory disorder characterized by quotidian fever, evanescent rash, arthralgia, and multisystem involvement. Due to its heterogeneous presentation and lack of specific diagnostic markers, AOSD remains a diagnosis of exclusion, often posing a significant diagnostic challenge to clinicians. The estimated incidence is approximately 0.16 cases per 100,000 persons per year, underscoring its rarity (Gerfaud-Valentin et al., 2020). This case report describes an unusual presentation of AOSD mimicking a ruptured hemorrhagic ovarian cyst, emphasizing the importance of a systematic exclusion approach in the diagnostic process.

Patient Profile

A 45-year-old female with no known comorbidities presented to the emergency department with acute systemic symptoms. She had a history of vaginal hysterectomy 17 months prior. There was no history of autoimmune disease, malignancy, chronic infections, or similar prior episodes.

History of Present Illness

The patient presented with sudden onset of severe abdominal pain, high-grade fever, bilateral lower limb swelling, and multiple skin lesions over the trunk and limbs. Symptoms were acute in onset, progressive in nature, and associated with significant fatigue and myalgia.

Physical Examination

On examination, the patient appeared unwell with diffuse abdominal tenderness without guarding or rigidity. Dermatological examination revealed multiple blanchable, erythematous maculopapular skin lesions over both arms, forearms, chest, abdomen, thighs, and legs. Musculoskeletal examination demonstrated generalized tenderness with pitting edema over both lower limbs. Cardiovascular, respiratory, and neurological examinations were within normal limits.

Investigations

Extensive investigations were performed to rule out infectious, malignant, and autoimmune etiologies. Laboratory findings are detailed in Table 1. Key findings included elevated inflammatory markers (ESR and CRP), leukocytosis with neutrophil predominance, markedly elevated serum ferritin (>1000 ng/mL), and negative autoimmune and infectious markers. Imaging studies, including abdominal ultrasonography and computed tomography (CT), revealed no acute intra-abdominal pathology.

Table 1: Laboratory Investigations and Reference Ranges

Parameter	Patient Value	Reference Range
Hemoglobin (Hb)	11.2 g/dL	12–16 g/dL
Total Leukocyte Count (TLC)	15,800 /mm³	4,000–11,000 /mm³
Neutrophils	85%	40–70%
Lymphocytes	10%	20–40%
Platelet Count	420,000 /mm³	150,000–400,000 /mm³
ESR	65 mm/hr	<20 mm/hr
C-Reactive Protein (CRP)	78 mg/L	<5 mg/L
Serum Ferritin	>1000 ng/mL	13–150 ng/mL (female)
Aspartate Transaminase (AST)	58 U/L	10–40 U/L
Alanine Transaminase (ALT)	62 U/L	7–56 U/L
Antinuclear Antibody (ANA)	Negative	Negative
Rheumatoid Factor (RF)	Negative	Negative
Blood Culture	No growth	No growth

Note. *ESR = Erythrocyte Sedimentation Rate; CRP = C-Reactive Protein; ANA = Antinuclear Antibody; RF = Rheumatoid Factor.*

Management

The patient was initially managed symptomatically in the emergency department with intravenous fluids, analgesics, antipyretics, and empirical broad-spectrum antibiotics while infectious etiologies were actively investigated. Imaging studies including abdominal ultrasonography and CT were performed to exclude surgical causes, including ruptured ovarian cyst and intra-abdominal abscess. However, no significant intra-abdominal pathology was detected.

Following comprehensive evaluation and exclusion of infectious, malignant, and autoimmune conditions, the clinical constellation of persistent fever, evanescent rash, arthralgia, leukocytosis with neutrophilia, and markedly elevated ferritin levels satisfied the Yamaguchi and Fautrel criteria for AOSD (Fautrel et al., 2024). Systemic corticosteroid therapy was initiated with oral prednisolone at an anti-inflammatory dose. The patient demonstrated rapid clinical improvement with resolution of fever, reduction in abdominal pain, disappearance of skin lesions, and improvement in overall functional status. Inflammatory markers declined progressively during subsequent monitoring. Gradual tapering of corticosteroid therapy was planned with close outpatient follow-up.

DISCUSSION

Adult-Onset Stills Disease is a rare systemic autoinflammatory disorder characterized by high spiking fevers, arthralgia or arthritis, transient salmon-colored rash, and multisystem inflammatory involvement. The estimated incidence of approximately 0.16 cases per

100,000 persons per year renders it an uncommon but clinically significant differential diagnosis in patients presenting with systemic inflammatory symptoms and pyrexia of unknown origin (Gerfaud-Valentin et al., 2020).

The pathogenesis of AOSD involves dysregulation of the innate immune system with excessive activation of pro-inflammatory cytokines including interleukin-1 (IL-1), interleukin-6 (IL-6), interleukin-18 (IL-18), and tumor necrosis factor-alpha (TNF-α), contributing to the systemic manifestations observed in the disease (Bindoli et al., 2024). These cytokine-driven mechanisms underlie the diverse and occasionally atypical clinical presentations that characterize AOSD.

AOSD typically presents with a clinical triad of quotidian fever, evanescent salmon-colored rash, and inflammatory arthritis. However, it may manifest with a wide spectrum of features including lymphadenopathy, hepatosplenomegaly, sore throat, myalgia, and serositis. Gastrointestinal symptoms and abdominal pain, although less common, have been reported and can lead to diagnostic confusion with acute surgical conditions (Tariq et al., 2024). In the present case, severe abdominal pain mimicked a ruptured hemorrhagic ovarian cyst, illustrating the diagnostic complexity associated with atypical AOSD presentations.

The diagnosis of AOSD remains largely clinical and based on exclusion of other causes of systemic inflammation. The Yamaguchi criteria, which emphasize clinical features including high spiking fever, arthralgia, leukocytosis with neutrophilia, and characteristic rash along with exclusion of infectious, malignant, and autoimmune conditions, are among the most widely used classification criteria. Elevated serum ferritin, particularly when markedly elevated (>1000 ng/mL) and associated with low glycosylated ferritin fractions, constitutes a supportive laboratory marker (Sahoo, 2025). In the present case, the clinical and laboratory constellation strongly supported the diagnosis following systematic exclusion of alternative etiologies.

Corticosteroids remain the cornerstone of initial treatment, with approximately 60–70% of patients demonstrating clinical response. In patients with refractory disease or those requiring steroid-sparing strategies, disease-modifying antirheumatic drugs (DMARDs) such as methotrexate may be introduced. Biologic agents targeting specific cytokines, including IL-1 inhibitors (anakinra), IL-6 inhibitors (tocilizumab), and TNF inhibitors, have demonstrated promising results in refractory or severe AOSD (Bindoli et al., 2024). Early diagnosis and timely initiation of therapy are associated with improved outcomes and reduced risk of life-threatening complications, including macrophage activation syndrome.

CONCLUSION

This case emphasizes the diagnostic complexity of Adult-Onset Stills Disease and the importance of maintaining a systematic exclusion approach when evaluating patients with unexplained systemic inflammation. Atypical presentations, such as severe abdominal pain mimicking acute surgical conditions, may significantly complicate the diagnostic process. A high index of clinical suspicion, supported by characteristic laboratory findings including markedly elevated serum ferritin and neutrophilic leukocytosis after exclusion of infectious and autoimmune etiologies, is essential for timely diagnosis. Early recognition and prompt initiation of corticosteroid therapy can lead to favorable outcomes and prevent serious disease-related complications.

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