International Journal of Contemporary Pediatrics
Kaushal S et al. Int J Contemp Pediatr. 2025 Nov;12(11):1889-1891
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291
DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20253499

Case Report

Still disease with persistent atypical dermatomyositis-like skin eruption with macrophage activation syndrome: a rare presentation

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Received: 15 September 2025
Accepted: 14 October 2025

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ABSTRACT

Still's disease is a type of inflammatory arthritis that causes fevers, rashes, and inflammation of joints and, sometimes, internal organs. Still's is also called systemic-onset juvenile idiopathic arthritis (SJIA), as it was first diagnosed in children. SJIA is a type of juvenile idiopathic arthritis (JIA), a group of arthritis conditions affecting children. JIA is one of the most common pediatric chronic diseases, with a prevalence rate varying from 3.8 to 400 cases/100,000 children, and a yearly incidence between 1.6 and 23 new cases for 100,000 children. SoJIA when considered as a JIA subtype includes about 10–20% of all JIA patients, but the percentage is higher in some countries and when only severe cases are considered. Indeed, in parts of Asia, it may account for up to 30–40% of all JIA cases. Here we report a case of Still's disease in pediatric patient with a persistent atypical dermatomyositis like skin eruption complicated by MAS, highlighting diagnostic challenges and management strategies.

Keywords: Still's disease, Inflammatory arthritis, SJIA

INTRODUCTION

Systemic-onset juvenile idiopathic arthritis (SoJIA), also known as Still's disease, is a rare and distinct subtype of JIA characterized by systemic features in addition to arthritis. It typically presents with quotidian spiking fevers, evanescent salmon-colored rash, and arthritis, along with variable systemic manifestations such as hepatosplenomegaly, lymphadenopathy, and serositis.

JIA remains one of the most common chronic rheumatologic conditions of childhood, with prevalence estimates ranging from 3.8 to 400 per 100,000 and annual incidence between 1.6 and 23 per 100,000 children.[1] SoJIA accounts for approximately 10–20% of all JIA cases worldwide, with higher rates of up to 30–40% reported in Asian populations.[2] The classic triad of quotidian fever, arthritis or arthralgia, and the evanescent rash is seen in nearly three-quarters of patients.[3] In recent years, beyond the well-known transient rash, persistent pruritic eruptions (PPEs) have been increasingly recognized as atypical cutaneous manifestations of SoJIA and adult-onset Still's disease (AOSD). These eruptions, distinct in morphology and duration, are associated with refractory disease courses, severe complications, and even increased mortality.[4,5] Their recognition is crucial in timely diagnosis and management.

CASE REPORT

Authors report the case of a previously healthy 9-year-old girl who presented to the pediatric casualty at IGMC Shimla with 12 days of quotidian fevers (up to 102°F), accompanied by chills, rigors, proximal muscle weakness, and bilateral lower limb pain. On examination, she had arthritis involving the knees and ankles, pedal edema, and striking dermatologic findings: a heliotrope rash around the eyelids and persistent pruritic cutaneous lesions, which were atypical for classical SoJIA. Laboratory evaluation revealed Hb 8.9 g/dl, platelets 90×10^9/l, WBC 13,000/µl, ESR 155 mm/hr, CRP 188 mg/l, ferritin >2000 ng/ml, triglycerides 387 mg/dl, and

D-dimer >3000. Liver enzymes were elevated (AST 220 U/l, ALT 82 U/l). ANA, RF, and cultures were negative. MRI of the thigh and myositis panel did not suggest myopathy. Ultrasound abdomen revealed hepatosplenomegaly with intra-abdominal lymphadenopathy.

A diagnosis of SoJIA complicated by macrophage activation syndrome (MAS) was established. She was managed with IVIG (2 g/kg) and intravenous methylprednisolone (5 mg/kg daily for 5 days), followed by hydroxychloroquine 200 mg/day. Remarkably, she became afebrile within 72 hours, with resolution of the heliotrope rash and muscle weakness. Laboratory markers normalized, including platelet count, ferritin, liver enzymes, and triglycerides, allowing transfer out of the ICU. At six months' follow-up, she remained in remission on maintenance immunotherapy, with no recurrence of MAS or atypical cutaneous lesions.

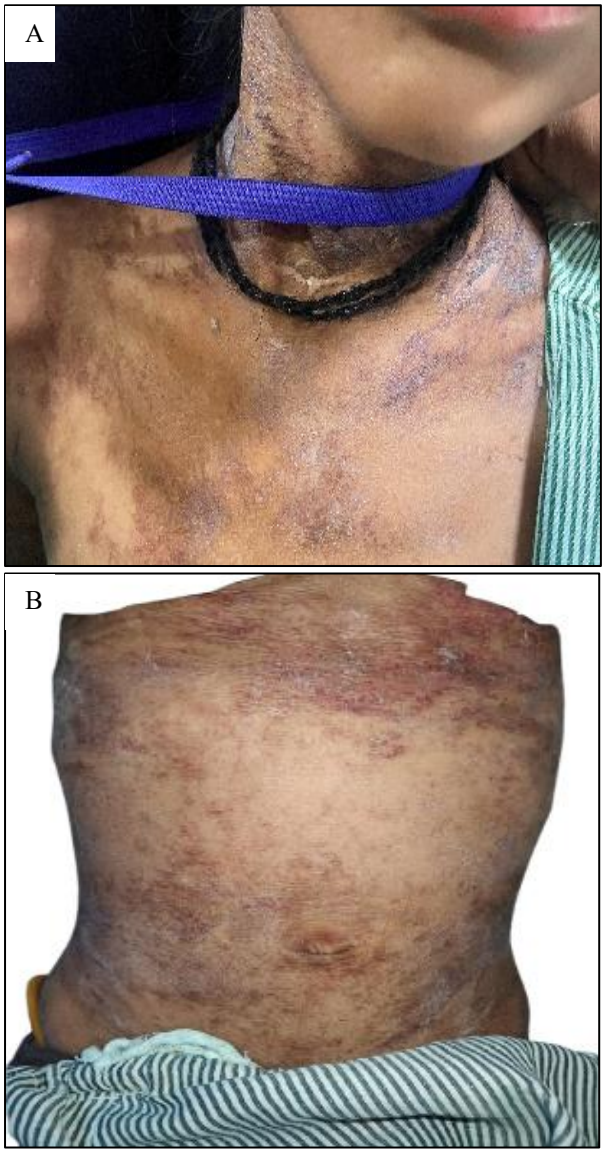

Figure 1: (A) Hyperpigmented patches over neck and chest. (B) Diffuse erythematous rash involving trunk.

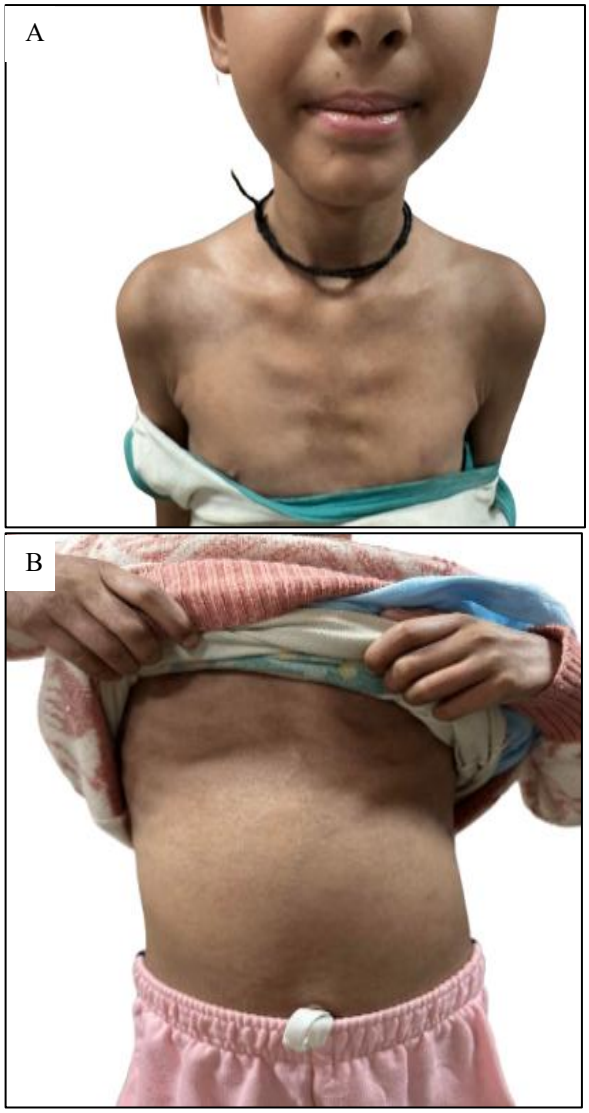

Figure 2 (A, B): Lesions resolved after treatment.

DISCUSSION

SoJIA, also known as Still's disease, is a rare and distinct subtype of JIA characterized by systemic features in addition to arthritis. It typically presents with quotidian spiking fevers, evanescent salmon-colored rash, and arthritis, along with variable systemic manifestations such as hepatosplenomegaly, lymphadenopathy, and serositis. JIA remains one of the most common chronic rheumatologic conditions of childhood, with prevalence estimates ranging from 3.8 to 400 per 100,000 and annual incidence between 1.6 and 23 per 100,000 children.

Still's disease, encompassing SoJIA in children and AOSD in adults, remains a diagnosis of exclusion. AOSD is classified by Yamaguchi criteria, while SoJIA follows the ILAR classification.[6] MAS complicates approximately 10–15% of SoJIA cases and is a potentially life-threatening hyperinflammatory state. It is characterized by fever, cytopenias, hepatosplenomegaly, coagulopathy, and hyperferritinemia.[7] Recognition of

MAS is vital, as delayed treatment significantly increases mortality risk.

CONCLUSION

This case underscores the diagnostic complexity of SoJIA, particularly when atypical dermatomyositis-like rashes are present. Awareness of these unusual cutaneous findings is critical, as they may represent harbingers of severe systemic disease, including MAS. Prompt recognition and initiation of targeted immunomodulatory therapy can be life-saving. report highlights the importance of dermatologic vigilance in juvenile inflammatory disorders, where cutaneous clues may provide the earliest signs of systemic complications.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Thatayatikom A, Modica R, De Leucio A. Juvenile idiopathic arthritis. In: StatPearls. Treasure Island (FL): StatPearls Publishing. 2023.
2. Cimaz R. Systemic-onset juvenile idiopathic arthritis. Autoimmun Rev. 2016;15(9):931-4.
3. Maeda-Aoyama N, Hamada-Ode K, Taniguchi Y, Nishikawa H, Arii K, Nakajima K, et al. Dyskeratotic cells in persistent pruritic skin lesions as a prognostic factor in adult-onset Still disease. Med (Baltimore). 2020;99(6):19051.
4. Lee JY, Hsu CK, Liu MF, Chao SC. Evanescent and persistent pruritic eruptions of adult-onset Still disease: a clinical and pathologic study of 36 patients. Semin Arthritis Rheum. 2012;42(3):317-26.
5. Fernández-Camporro Á, Rodriguez-Diaz E, Beteta-Gorriti V, Gonzalvo-Rodríguez P, Alvarez-Cuesta C. Still disease with persistent atypical dermatomyositis-like skin eruption: two cases associated with macrophage activation syndrome. Clin Exp Dermatol. 2022;47(11):1991-4.
6. Tomaras S, Goetzke CC, Kallinich T, Feist E. Adult-onset Still's disease: clinical aspects and therapeutic approach. J Clin Med. 2021;10(4):733.
7. Grom AA, Horne A, De Benedetti F. Macrophage activation syndrome in the era of biologic therapy. Nat Rev Rheumatol. 2016;12(5):259-68.

Cite this article as: Kaushal S, Chauhan J. Still disease with persistent atypical dermatomyositis-like skin eruption with macrophage activation syndrome: a rare presentation. Int J Contemp Pediatr 2025;12:1889-91.