SYSTEMIC-ONSET JUVENILE IDIOPATHIC ARTHRITIS: A CHALLENGE FOR THE DERMATOLOGIST

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Abbreviations **sJIA** = systemic-onset juvenile idiopathic arthritis;
MAS = macrophage activation syndrome

Case report. A 2-year-old boy presented with recurrent febrile episodes for approximately six months, which worsened one week prior to evaluation, reaching a maximum temperature of 39.5°C, accompanied by arthralgia, abdominal pain, and an evanescent rash. The rash appeared during fever spikes and resolved with defervescence. On physical examination, salmon-colored macules and papules approximately 5 mm in diameter, some coalescing into plaques, were noted on the upper and lower limbs and on both the anterior and posterior trunk (Figs 1, 2). Laboratory tests showed marked elevation of acute-phase reactants: leukocytosis (23,460/µL, nr 4,00-10,200) with neutrophilic predominance (66%, nr 40-60), elevated C-reactive protein (89.1 mg/L, nv <10), erythrocyte sedimentation rate (82 mm/h, nv <20), and ferritin (1,040 ng/mL, nr 17.9-464). Ultrasound revealed arthritis in the left wrist, left hip, and both knees. The patient exhibited severe hyperferritinemia with decreased hemoglobin levels; hence, a diagnosis of systemic-onset juvenile idiopathic arthritis associated with partial macrophage activation syndrome (MAS) was made. Immunomodulatory treatment was initiated with systemic corticosteroids and methotrexate; however, due to MAS involvement, therapy was escalated to include cyclosporine and tocilizumab, resulting in decreased fever frequency, improvement in inflammatory markers, and no new skin lesions.

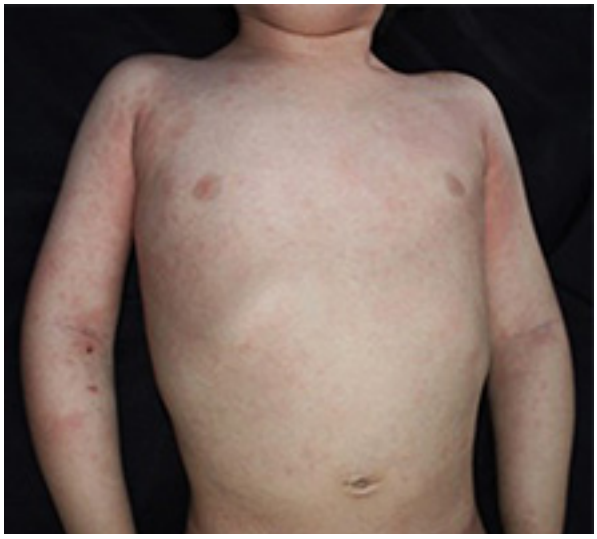

Fig. 1

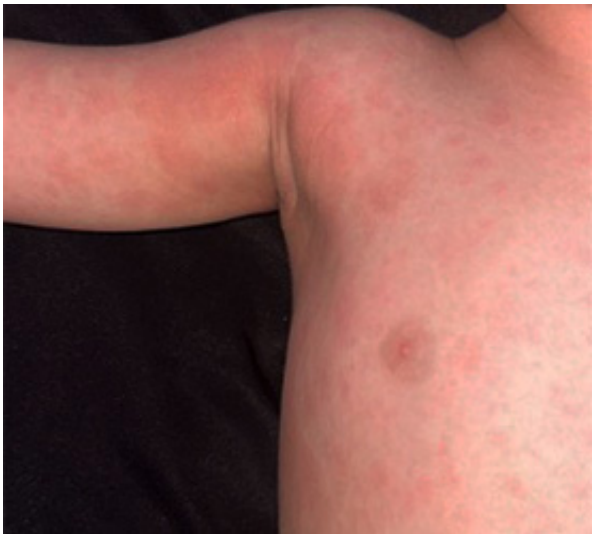

Fig. 2

Fig. 1, 2: Evanescent rash in systemic-onset juvenile idiopathic arthritis.

Discussion. From a pathophysiological standpoint, systemic onset juvenile idiopathic arthritis (sJIA) differs from other forms of JIA due to the predominance of autoinflammatory mechanisms driven by innate immune system dysfunction, with interleukin-1 and interleukin-6 playing central roles (2, 5); sJIA is the least common JIA subtype, accounting for approximately 10-20% of cases in Western cohorts. The estimated annual incidence ranges from 0.4 to 0.9 per 100,000 children under 16 years, with point prevalence as high as 7 per 100,000, as reported in population-based studies from Western Australia (6). There is no clear sex predominance, although some studies suggest a slightly higher incidence in females (6). The average age of onset is typically between 5 and 7 years, though onset can occur at any age in childhood (6).

sJIA is characterized by generalized inflammation affecting joints and other organs. Approximately 10% of patients develop macrophage activation syndrome (MAS), a severe complication characterized by hemophagocytic activity (7), as suspected in our patient. Clinically, sJIA manifests with daily high fevers and arthritis. A non-pruritic, salmon-colored, evanescent rash associated with fever is common, typically involving the trunk and extremities (8). Histopathology is nonspecific and shows variable infiltration by lymphocytes and neutrophils (9). Thus, skin biopsy is not mandatory, but may aid in diagnosis. Our patient presented with the classic rash of sJIA, appearing with fever spikes and disappearing afterward, which was a key diagnostic clue.

Diagnosis is based on the International League of Associations for Rheumatology (ILAR) criteria, which include persistent fever for at least 3 days associated with either two major criteria (evanescent rash and arthritis), or one major and two minor criteria (such as generalized lymphadenopathy, hepatosplenomegaly, persistent arthralgia for at least two weeks, neutrophilic leukocytosis, or serositis). Management includes nonsteroidal anti-inflammatory drugs, corticosteroids, methotrexate, and, in some cases, biologic therapy (1-4).

Conclusion. Systemic-onset juvenile idiopathic arthritis is a condition that requires early diagnosis due to its systemic involvement and potential for serious complications. The ILAR criteria are critical for identification. It is essential for dermatologists to be familiar with the clinical signs of this disease, as symptoms may mimic more common pediatric conditions.

Conflicts of interest

The authors declare that they have no conflicts of interest.

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