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A RARE PRESENTATION OF SYSTEMIC-ONSET POLYARTHRITIC JUVENILE IDIOPATHIC ARTHRITIS WITH SEVERE ITCHING WITHOUT RASH

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Abstract
Systemic-onset juvenile idiopathic arthritis (SJIA) is a distinct subtype of juvenile idiopathic arthritis characterized by quotidian fever, arthritis, lymphadenopathy, hepatosplenomegaly, and an evanescent rash. Dermatologic manifestations are typical in systemic disease, commonly presenting as a salmon-pink rash. However, isolated severe pruritus without rash is extremely uncommon and may create diagnostic uncertainty. We report a 5-year-old male with a 2-month history of intermittent high-grade fever, progressively worsening polyarthritis, and severe pruritus without visible skin rash. Examination revealed generalized lymphadenopathy, multiple polyarticular joint swellings, and scratch marks without erythema. Laboratory evaluation showed anemia, neutrophilic leukocytosis, thrombocytosis, markedly elevated C-reactive protein, and positive antinuclear antibody with nuclear and speckled pattern. Radiography revealed periarticular osteopenia without erosions. The child was treated with NSAIDs, antibiotics, and subsequently high-dose pulse methylprednisolone following ANA positivity, resulting in dramatic clinical improvement. He was discharged on weekly methotrexate with folic acid supplementation. This case highlights an atypical presentation of SJIA where severe itching without rash may obscure the diagnosis. Early recognition and initiation of immunosuppressive therapy can prevent long-term sequelae.

Keywords: Juvenile Idiopathic Arthritis; Systemic-Onset JIA; Autoimmunity; Antinuclear Antibody; Methylprednisolone pulse therapy.

Introduction

Juvenile idiopathic arthritis (JIA) encompasses a heterogeneous group of chronic inflammatory arthritides of unknown aetiology occurring before 16 years of age [1]. Among these, systemic-onset JIA (SJIA) is uniquely characterised by quotidian fever, arthritis, and systemic features such as rash, hepatosplenomegaly, and serositis [2,3]. Unlike other JIA phenotypes, SJIA is now conceptualised more as an autoinflammatory disorder driven by innate immune system dysregulation, with major involvement of macrophages and neutrophils and overproduction of pro-inflammatory cytokines, particularly interleukin-1 (IL-1) and interleukin-6 (IL-6) [4–6].

Epidemiologically, SJIA accounts for approximately 10–20% of all JIA cases and is associated with higher rates of systemic complications [5]. Clinically, its hallmark features include high-spiking quotidian fever, arthritis/arthralgia, and an evanescent salmon-pink maculopapular rash temporally associated with fever spikes [7]. Laboratory abnormalities typically include neutrophilic leucocytosis, thrombocytosis, elevated C-reactive protein (CRP) or erythrocyte sedimentation rate (ESR), anemia of inflammation and elevated serum ferritin [8].

Cutaneous manifestations — primarily the salmon-pink rash — are seen in most cases, yet presentations without rash are rare and may obscure early recognition [8]. Moreover, severe pruritus (itching) without any visible skin eruption is extremely uncommon in SJIA, and may be mistaken for allergic, dermatologic or infectious aetiologies [9,10]. Because delayed diagnosis and treatment are associated with increased risk of chronic arthritis, joint damage and life-threatening complications such as macrophage activation syndrome (MAS) [11], awareness of atypical manifestations is essential. Herein we describe a case of SJIA in a 5-year-old boy presenting with severe generalized pruritus without rash, in addition to prolonged fever and polyarthritis, to illustrate the need for heightened clinical suspicion and prompt immunomodulatory therapy.

Case Presentation

A 5-year-old boy presented with a 2-month history of intermittent fever and worsening joint pain and swelling for the last 15-16 days, resulting in inability to walk. Fever was insidious in onset, initially low-grade (2–3 spikes/day), gradually escalating to high-grade (3–4 spikes/day) with chills and intermittent antipyretic relief. Joint involvement included bilateral elbows, wrists, proximal interphalangeal joints, knees and ankles, with morning stiffness and progressive limitation of motion. The child also complained of severe generalized itching for 15–16 days, without any visible rash. There was no recent history of loose stools, cough/cold, difficulty in breathing, trauma, or other systemic illness. A similar episode a year ago was associated with a cheek rash per parental report. No family history of rheumatologic disease was noted.

On general examination, the child was moderately built, poorly nourished, and pale. He had generalized lymphadenopathy. Musculoskeletal examination revealed symmetric swelling of bilateral elbows, wrists, proximal interphalangeal joints, knees and ankles; the swellings were warm and tender but lacked erythema or trauma signs. Multiple excoriations (scratch marks) were evident around involved joints. Passive motion exacerbated pain. Respiratory, cardiovascular and neurological examinations were unremarkable; there was no hepatosplenomegaly. (Figure 1-5)

Figure 1-5: Clinical photographs showing symmetrical swelling of bilateral knees, wrists and ankles with surrounding scratch marks (due to intense pruritus).

The lab findings revealed Hemoglobin 6.4g/dl, Total Leucocyte count 13,400/ml with predominant neutrophilia (69.2%), platelet count of 8.45 lakhs/ml. Peripheral blood smear revealed microcytic hypochromic blood picture with reactive neutrophilia with reactive thrombocytosis. The C-reactive protein was 204.9 mg/dl, KFT, LFT, serum electrolytes were within normal limits. RA factor was done which was found to be negative. Anti-Nuclear antibody test was done which was positive and revealed a nuclear and speckled (1:100) (AC-2,4,5) pattern. Radiological investigation like X-ray Bilateral knee antero-posterior view was done, which did not show any osteophytic growth, and showed slight osteopenia around the joint.(Figure 6)

Management and Outcome

On admission, the patient was started on intravenous ceftriaxone, paracetamol and syrup ibuprofen. By day 3 of hospitalisation, the frequency and intensity of fever spikes decreased, joint swelling reduced slightly and joint mobility improved. Following positive ANA results, intravenous methylprednisolone pulse therapy was administered for 5 days. The patient showed marked clinical improvement: resolution of fever, significant reduction in pruritus, improved joint swelling and mobility. He was discharged on weekly oral methotrexate and daily folic acid. The patient remains under outpatient follow-up.

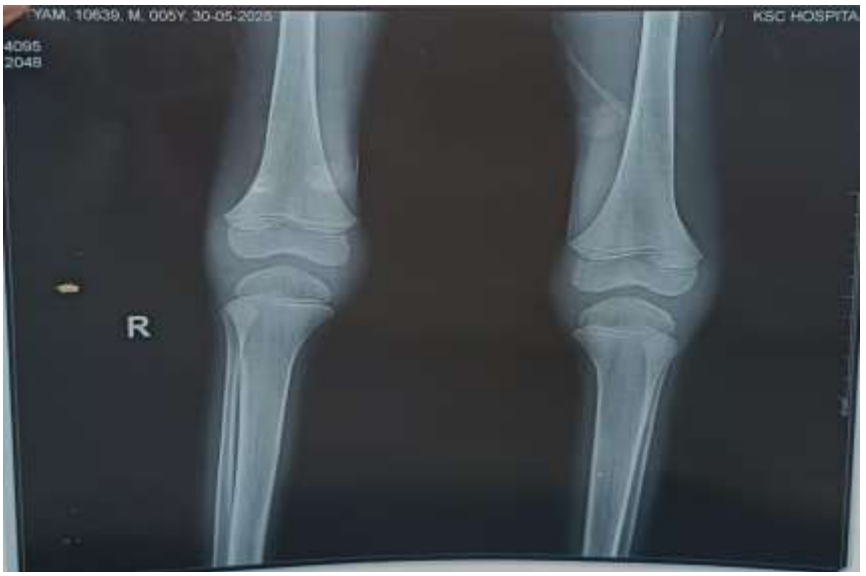

Figure 6: Antero-posterior radiograph of bilateral knees showing mild periarticular osteopenia without joint space narrowing or erosions.

Discussion

Systemic-onset JIA (SJIA) is characterised by a combination of systemic hyperinflammation and arthritis, distinct from other JIA subtypes [1,2]. It represents an autoinflammatory entity rather than classical autoimmune disease, with IL-1 and IL-6 playing key pathogenetic roles [4–6]. Elevated serum IL-6 levels are associated with the extent of systemic symptoms such as thrombocytosis and osteopenia, reflective of aggressive inflammatory milieu [8].

Typical presentation of SJIA includes quotidian fever, arthritis, rash, lymphadenopathy, hepatosplenomegaly, and serositis [7]. The salmon-pink evanescent rash is one of the more recognisable features, reported in most cases [8]. Notably, our patient lacked any visible rash yet had severe generalized pruritus — an atypical dermatologic manifestation scarcely documented in the literature. While pruritus is common in dermatoses or allergic conditions, its occurrence without rash in SJIA may lead to diagnostic delay and misdirection towards allergy, dermatology or infection.

Atypical presentations, such as persistent plaques or papules instead of the classic rash, have been described [9], but cases of isolated pruritus without skin eruption remain exceptional. Such a presentation suggests possible cytokine-mediated neuro-immune activation or subclinical dermal inflammation rather than overt cutaneous involvement. Awareness of such variant features is necessary to broaden diagnostic suspicion.

Laboratory findings in SJIA often include anemia, neutrophilic leukocytosis, thrombocytosis, elevated CRP/ESR, and occasionally high ferritin levels [8,11]. Radiologic features in early disease may show periarticular osteopenia without joint destruction, consistent with inflammatory bone changes rather than chronic damage [12]. In our case, the presence of microcytic anemia may reflect chronic inflammation combined with iron deficiency, while high CRP and thrombocytosis underscored systemic inflammation. ANA positivity, though uncommon in SJIA, has been noted and does not exclude the diagnosis [11].

Therapeutically, prompt immunomodulation is imperative, as untreated or delayed disease may progress to chronic arthritis, growth impairment, and macrophage activation syndrome (MAS) — a life-threatening hyper-inflammatory complication [11]. Corticosteroid pulse therapy remains a mainstay for severe systemic disease; methotrexate is commonly used for maintenance therapy [13].

More recently, biologic agents targeting IL-1 (anakinra, canakinumab) and IL-6 (tocilizumab) have transformed the treatment landscape, improving outcomes and reducing steroid burden [4,6,14]. Early initiation of biologics (so-called 'window of opportunity') may favour remission and alter disease trajectory [15].

Our case illustrates several key points: (i) SJIA may present without classic rash, instead manifesting as severe pruritus; (ii) early recognition and management with pulse steroids and methotrexate can result in excellent clinical improvement; (iii) variant presentations necessitate broad clinical suspicion to avoid delays. Monitoring biomarkers (e.g., S100A8/A9, IL-18, ferritin) may aid in flare prediction and risk stratification, though these were not measured in our patient [12].

Conclusions

Systemic-onset JIA may rarely present with severe pruritus without any rash, complicating early recognition. Clinicians should maintain a high index of suspicion in children presenting with prolonged fever, polyarthritis and unexplained pruritus. Early immunosuppressive therapy is critical to prevent long-term morbidity and life-threatening complications.

Declarations

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Ethics & Consent: Written informed consent was obtained from the parent of the patient for publication of this case report and accompanying images.

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