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Case Report

A Case Report: Systemic-Onset Juvenile Idiopathic Arthritis Complicated by Peripheral Gangrene in A 9-Year-Old Girl

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Abstract

Juvenile idiopathic arthritis (JIA) is a chronic inflammatory disorder that can lead to a range of complications in children. Vasculitis is a rare but severe complication that may result in peripheral gangrene, which requires prompt diagnosis and aggressive treatment. Systemic-onset Juvenile Idiopathic Arthritis (SoJIA) is classified within the spectrum of JIA. We present the case of a 9-year-old girl diagnosed with SoJIA who experienced high fever, severe joint involvement, and transient skin rashes for six weeks without receiving adequate treatment. After intensive therapy, the incipient gangrene of the right fifth toe fully resolved, and all other joints, which had been severely swollen and painful, wholly recovered. However, the established gangrene of the fingers showed no improvement.

This case underscores the critical importance of early diagnosis and timely intervention in preventing irreversible complications in patients with SoJIA.

Keywords: Systemic-onset juvenile idiopathic arthritis; Vasculitis; Peripheral gangrene

Introduction

SoJIA, also known as Still's disease, is a systemic inflammatory disorder classified within the spectrum of Juvenile Idiopathic Arthritis (JIA) [1]. According to the International League of Associations for Rheumatology (ILAR) classification, SoJIA is characterized by arthritis in one or more joints, either preceded or accompanied by a fever lasting at least two weeks. This fever is typically daily ("quotidian") for at least three days and is associated with one or more of the following features: an evanescent (nonfixed) erythematous rash, generalized lymphadenopathy, hepatomegaly and/or splenomegaly, and serositis [2].

The clinical presentation of SoJIA is often marked by a severely ill child exhibiting fever, anemia, and significant joint pain [3].

Case Presentation

A previously healthy 9-year-old girl presented with a 6-week history of high-grade fever (temperature: 39.6°C), severe joint swelling, intense pain, and poor general condition. Upon examination, significant swelling was observed in the hips, knees, hands, fingers, ankles, and toes. Extensive necrosis was present at the tips of her fingers, which caused severe pain (Figure 1). Cyanosis was noted at the proximal phalanx of the fifth toe on the right foot, which progressively worsened. Small areas of skin necrosis were also observed on the legs. Due to severe pain and swelling, the patient was unable to walk. The general condition was poor, with no lung abnormalities, heart, abdomen, or pharynx abnormalities. There was no family history of chronic disease, and the patient's parents were not related.

Laboratory tests revealed marked systemic inflammation, including severe anemia (Hb 6.5 g/dL), neutrophilia, thrombocytosis, elevated CRP, and an elevated erythrocyte sedimentation rate (ESR). Liver enzymes and renal function were within normal limits, and autoantibodies were absent—laboratory and radiologic investigations excluded infectious and oncological causes. The patient was diagnosed with SoJIA due to prolonged high fevers (peaking once or twice daily for 6 weeks), polyarthritis, transient skin rashes (especially during febrile episodes), and exclusion of other potential causes.

The treatment plan included intravenous methylprednisolone (1 mg/kg/day), methotrexate (10 mg weekly), folic acid (5 mg/day), paracetamol, tramadol (IV), erythrocyte suspension transfusion (EST), and subcutaneous enoxaparin sodium.

During clinical follow-up, the patient's general condition improved. Although high-grade fever persisted (once or twice a day), joint swelling in the hips, knees, hands, and feet gradually decreased, and scars remained at the sites of previous swelling (Figure 2). Transient skin rashes were still observed during febrile episodes. The cyanosis in the fifth toe of the right foot wholly resolved, but necrosis at the fingertips of both hands worsened over time.

Due to the lack of improvement in the necrosis of the fingers, a vascular surgeon recommended amputation. As the gangrene in the fingers was irreversible, amputation of the affected fingers was performed by the orthopedic team (Figure 3). The patient also

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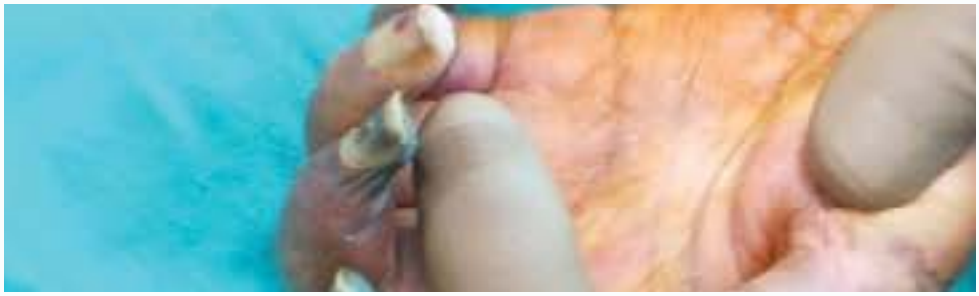

Figure 1: (A) The necrosis, the tips of the second and fifth fingers on the right hand (B) The severe necrosis of the first, second, third, and fifth fingers on the left hand and the moderate necrosis on the fourth left finger and significant edema in the left hand. (C) Small skin necrosis on the legs.

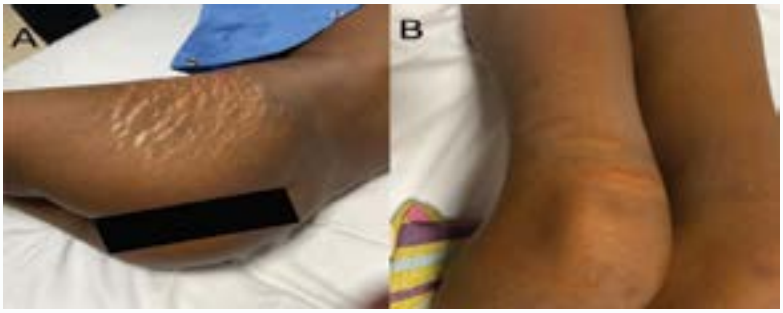

Figure 2: (A) Very large scars on both hips after recovery. (B) Scars on both upper knees after recovery.

Figure 3: Photograph taken 15 months and 16 days after discharge from hospital.

received psychiatric support.

She was discharged on a regimen of prednisone (1 mg/kg/day) and methotrexate (10 mg weekly).

Post-discharge follow-up included visits at 1 week, 1 - 2 months, and subsequently every 3 months. At the 2-month follow-up, the patient's hemoglobin improved to 12.1 g/dL; at 10 months, it reached 14 g/dL. Liver, kidney function, and blood glucose remained normal.

Leukocytosis and thrombocytosis gradually normalized, with complete resolution at 6 months. CRP and ESR levels gradually returned to normal, with complete resolution observed by 12 months.

The patient continued prednisone (1 and 0.5 mg/kg/day) and methotrexate (10 mg weekly) for a total of 12 months until both ESR and CRP values normalized.

Discussion

Juvenile Idiopathic Arthritis (JIA) can lead to severe systemic complications, one of the most serious being vasculitis. In this case, vasculitis resulted in peripheral gangrene, a rare but potentially life-threatening complication of Systemic-onset JIA (SoJIA). Previous literature has reported cases of SoJIA leading to peripheral gangrene, with some resulting in foot amputations [4]. The pathogenesis of rheumatoid vasculitis is believed to involve immune complex deposition within blood vessel walls, triggering inflammation, endothelial damage, and subsequent tissue necrosis [5]. In the context of JIA, vasculitis is a rare but potentially life-threatening complication, which can lead to significant morbidity if not promptly addressed.

Treatment for vasculitis in JIA typically involves immunosuppressive therapy, including corticosteroids and disease-modifying anti-rheumatic drugs (DMARDs) such as methotrexate. In some cases, biologic DMARDs in combination with conventional DMARDs may be considered [6]. In this case, despite experiencing prolonged high fever and severe joint involvement for six weeks, the patient had not received adequate treatment prior to hospitalization. Upon admission, corticosteroids and methotrexate were initiated, significantly improving the non-necrotic joint symptoms within three weeks. The cyanosis of the fifth toe of the right foot was entirely resolved by the third week (Figure 3). However, the necrosis in the

fingertips did not regress.

Literature on reversing finger necrosis once it has fully developed is limited. Based on current evidence, once gangrene has become established, it is unlikely to resolve with medical therapy alone. This case underscores the importance of early diagnosis and aggressive treatment to prevent irreversible tissue damage.

At admission, the patient weighed 33 kg (75-90th percentile) and had a height of 142 cm (90-97th percentile). By the 16-month follow-up, her weight increased to 43 kg (75-90th percentile), and her height grew to 149 cm (90-97th percentile). Despite the severe course of her illness, she was protected from the long-term side effects of the disease and low-dose methylprednisolone.

Conclusion

SoJIA, complicated by peripheral gangrene due to vasculitis, is a rare and severe manifestation that requires early recognition and aggressive management. The complete resolution of cyanosis in the right fifth toe within three weeks (Figure 3) highlights the positive impact of timely intervention, while the irreversible necrosis of the fingers emphasizes the need for early and aggressive treatment to prevent tissue loss. This case underscores the importance of prompt diagnosis and intervention in managing SoJIA complicated by vasculitis, with the rare occurrence of peripheral gangrene in children making this case a valuable addition to the literature.

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