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CUTANEOUS MANIFESTATIONS IN JUVENILE IDIOPATHIC ARTHRITIS PATIENTS, A CHALLENGE IN DIAGNOSIS: A CASE REPORT

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ABSTRACT
Juvenile idiopathic arthritis (JIA) is a heterogeneous group of chronic arthritides in children under 16 years old, characterized by persistent arthritis of unknown etiology with variable systemic and extra-articular manifestations. Systemic juvenile idiopathic arthritis (sJIA) represents a distinct subtype with clinical features including intermittent high fever, arthritis, and cutaneous eruptions, which often pose diagnostic challenges due to overlapping manifestations with other autoimmune or vasculitic diseases. The purpose of reporting this case is to highlight the diagnostic challenges of sJIA with atypical cutaneous manifestations, which may mimic other autoimmune disorders, and to emphasize the importance of careful clinical evaluation for early recognition and management. We report the case of an 8-year-old girl who presented with generalized blackish maculopapular rash, butterfly-shaped erythematous plaques on the face, high intermittent fever, and symmetrical arthritis of both knees. Laboratory examinations revealed elevated inflammatory markers and liver enzymes, with negative antinuclear antibody and anti double stranded DNA results. The data were collected through patient follow-up during hospitalization and regular outpatient visits at the Dermatology, Venereology, and Aesthetics Polyclinic of H. Adam Malik Hospital Medan over a 3-month period. Histopathological findings showed vasculitis. Differential diagnoses considered were systemic lupus erythematosus (SLE) and Henoch-Schönlein purpura (HSP), which were excluded based on clinical, laboratory, and histopathological evaluations. The final diagnosis was systemic juvenile idiopathic arthritis. The patient was treated with methotrexate, folic acid, topical corticosteroids, and emollients, with gradual clinical improvement observed during follow-up. This case highlights the importance of recognizing cutaneous manifestations in JIA, which may mimic other systemic diseases, and emphasizes the role of a multidisciplinary approach for early diagnosis and management to prevent long-term complications.

Keywords: butterfly rash; cutaneous manifestation; juvenile idiopathic arthritis; systemic JIA; vasculitis

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INTRODUCTION

Juvenile idiopathic arthritis (JIA) is a heterogeneous group of chronic arthritides affecting children under 16 years of age, persisting for at least six weeks with no identifiable etiology. Since 1995, the term juvenile rheumatoid arthritis has been replaced by JIA, which is currently classified into seven subtypes: systemic arthritis, oligoarthritis, polyarthritis with negative rheumatoid factor (RF), polyarthritis with positive RF, psoriatic arthritis, enthesitis-related arthritis, and undifferentiated arthritis. Each subtype demonstrates distinct clinical features, genetic predisposition, laboratory findings, disease course, and prognosis, although all share chronic arthritis as the main manifestation. Extra-articular and systemic features are also common in JIA, including cutaneous, ocular, and visceral involvement. (Piette WW., 2019; Thatayatikom A., et al., 2023).

The incidence of JIA ranges between 1.6 and 23 cases per 100,000 children per year, while prevalence varies from 3.8 to 400 per 100,000 children, with the highest incidence occurring in those aged 1–3 years. (Giancane G., et al., 2019; Cimaz R., 2019). Oligoarthritis is the most common subtype globally, while systemic JIA (sJIA) and enthesitis-related arthritis are reported more frequently in Southeast Asia (Weiss JE., et al., 2017; Consolaro A., et al., 2019). Most JIA

subtypes are more prevalent in females, except enthesitis-related arthritis, which predominantly affects males, whereas systemic JIA occurs equally in both sexes. (Cattalini M., et al., 2019).

The etiology of JIA remains unclear, but the interplay of genetic and environmental factors plays a critical role. Genetic contributions are supported by familial aggregation and twin studies, while environmental risks such as antibiotic exposure and cesarean delivery have been implicated. Conversely, protective factors include exclusive breastfeeding and having multiple siblings. arthritis, which predominantly affects males, whereas systemic JIA occurs equally in both sexes. (Cattalini M., et al., 2019; Horton DB., et al., 2019).

The clinical course of JIA is unpredictable, ranging from self-limiting to progressive forms with severe joint destruction and disability (Ravelli A., et al., 2017). Besides joint inflammation, JIA may affect growth, bone maturation, and systemic organs. Cutaneous manifestations are among the most important extra-articular findings. The classic skin manifestation in systemic JIA is a non-pruritic salmon-pink macular rash, typically associated with fever and located on the trunk and extremities (Piette WW., 2019). However, atypical lesions, including hyperpigmented plaques or butterfly-shaped rashes, may mimic other autoimmune or vasculitic diseases such as systemic lupus erythematosus (SLE) and Henoch–Schönlein purpura (HSP), creating a diagnostic challenge.

Here, we report a case of systemic juvenile idiopathic arthritis with cutaneous manifestations mimicking butterfly rash and vasculitis, highlighting the importance of thorough clinical, laboratory, and histopathological evaluation to distinguish JIA from other autoimmune disorders. This case report aims to describe the clinical features, diagnostic process, and management of systemic juvenile idiopathic arthritis (sJIA) in a pediatric patient. The report also seeks to underline the importance of distinguishing sJIA from other autoimmune or vasculitic diseases that may present with overlapping cutaneous and systemic manifestations. Against this background, the present case report aims to highlight the clinical presentation and diagnostic challenges of systemic juvenile idiopathic arthritis (sJIA) in children, and to emphasize the importance of early recognition and comprehensive evaluation in differentiating sJIA from other autoimmune or vasculitic diseases with overlapping cutaneous and systemic features.

METHOD

This study is presented as a single case report. Data were obtained through direct clinical observation, patient anamnesis, physical and dermatological examination, laboratory investigations, and histopathological evaluation. Only one patient was routinely followed up, as this represents a single case report. The patient was purposively selected as she presented with cutaneous manifestations and systemic symptoms suggestive of juvenile idiopathic arthritis. Data collection included clinical photographs, hematological and immunological test results, urinalysis, and histopathological findings. The information was descriptively analyzed and compared with existing literature on systemic juvenile idiopathic arthritis and other autoimmune diseases with overlapping cutaneous features.

CASE ILLUSTRATION

An 8-year-old girl was referred from the Pediatric Department to the Dermatology, Venereology, and Aesthetics Polyclinic of H. Adam Malik Hospital Medan on May 22, 2024, with complaints of blackish spots throughout the body accompanied by joint pain for the past month. The rash initially appeared four months earlier as reddish macules on the abdomen, later spreading to the face and extremities, and evolving into generalized lesions with xerosis but without pruritus or pain. Three months earlier, she developed swelling and pain in both knees, which progressed to involve the ankles, resulting in difficulty walking. Two weeks before admission, she also experienced intermittent high fever up to 40.1°C, responsive only to antipyretics. There was no history of trauma, familial autoimmune disease, or abnormal birth and development.

On physical examination, the patient was compos mentis, with body weight 22 kg, height 121 cm, blood pressure 100/60 mmHg, pulse rate 88/min, respiratory rate 18/min, and body temperature 40.1°C. Dermatological examination revealed erythematous-hyperpigmented plaques on the face forming a butterfly-shaped rash, generalized hyperpigmented maculopapular lesions on extremities and trunk, edema around the knees, and generalized xerosis cutis (Figure 1).

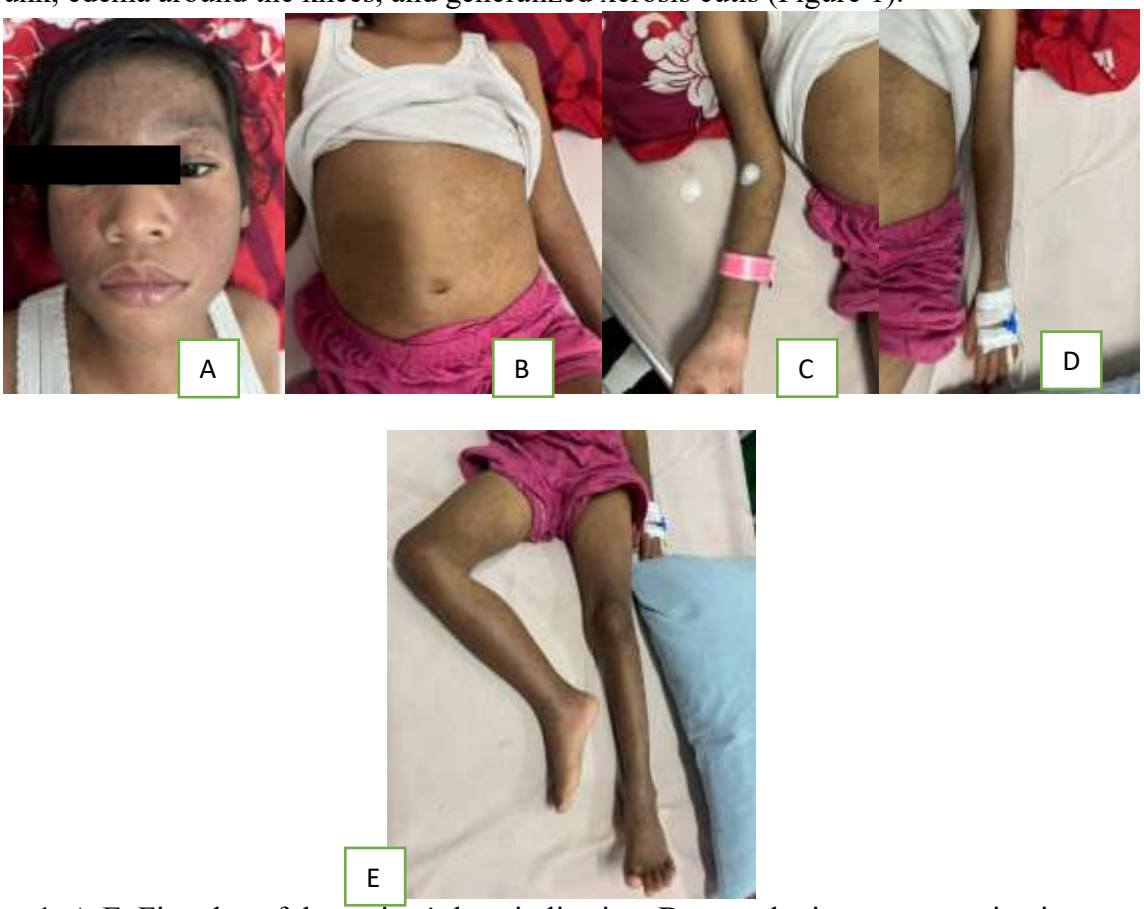

Figure 1. A-E. First day of the patient's hospitalization. Dermatologic status examination revealed partially erythematous hyperpigmented plaques in the facial region forming a butterfly rash (A), macules and generalized hyperpigmented plaques with maculopapular on the superior and inferior extremities (B,C,D,E), edema in the patella region dextra (E) and generalized cuticular xerosis.

Laboratory results showed leukocytosis (13,420/µL), neutrophilia (79.9%), thrombocytosis (428,000/µL), elevated AST (126 U/L), borderline ANA (18.5 IU/mL), and slightly elevated CRP (0.7 mg/dL). Urinalysis revealed glucosuria (1+) with no hematuria or proteinuria. A skin biopsy taken from the lesion revealed an intact epidermis with squamous epithelium, normal basement membrane, and fibrous connective tissue in the dermis. Perivascular inflammatory infiltrates of lymphocytes and neutrophils were observed, consistent with vasculitis, with no dysplastic or malignant cells (Figure 2).

Based on the anamnesis, clinical findings, and supporting investigations, the patient was diagnosed with systemic juvenile idiopathic arthritis (sJIA). Initial therapy included paracetamol, folic acid, urea cream 10%, and desoxymethasone 0.25% cream. After histopathology results confirmed vasculitis, methotrexate 10 mg weekly was added in combination with folic acid, and topical therapy was continued.

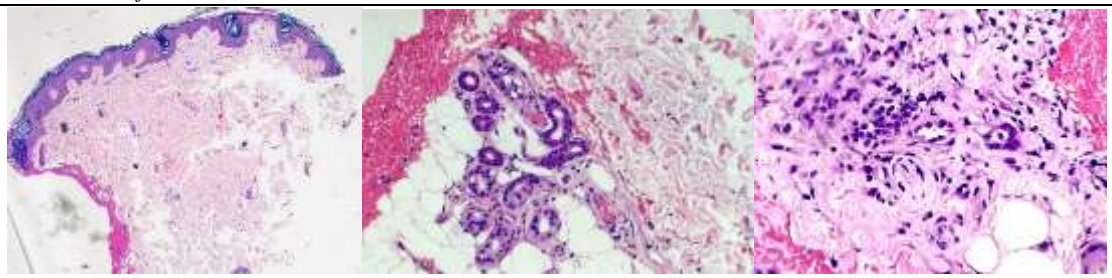

Figure 2. Histopathologic results showed a picture of vasculitis. The epidermal layer is borderline normal, the basement membrane is intact. The dermis layer consisted of fibrous connective tissue. In some foci, there was infiltration of inflammatory cells of lymphocytes and neutrophils surrounding the blood vessels.

At the first follow-up (1 week), fever had resolved, but blackish skin lesions and joint pain persisted. At the second follow-up (1 month), cutaneous lesions had improved, joint pain was reduced, and the patient was able to stand and walk with less difficulty, although residual hyperpigmented lesions and knee edema were still present (Figures 3–4).

Figure 3. A-D. Dermatologic examination revealed partially erythematous hyperpigmented plaques in the fascial region forming a butterfly rash (A), hyperpigmented plaques in the abdominal region (B) hyperpigmented maculopapular, erosion, excoriation in the superior and inferior extremities (C,D), edema in the patella dextra region (D) and cuticular xerosis.

The prognosis in this case was *quo ad vitam bonam, quo ad functionam dubia ad bonam, and quo ad sanationam dubia ad bonam.*

DISCUSSION

Systemic juvenile idiopathic arthritis (sJIA) is a subtype of juvenile idiopathic arthritis characterized by persistent arthritis accompanied by systemic features such as fever and rash. The patient, an 8-year-old girl, presented with intermittent high fever, persistent symmetrical arthritis, and a non-pruritic maculopapular rash resembling a butterfly pattern, which are consistent with the clinical criteria for sJIA established by the International League of Associations for Rheumatology (ILAR).[1] Laboratory results showed leukocytosis, neutrophilia, and elevated liver enzymes, while histopathology demonstrated perivascular inflammatory infiltrates supporting an autoimmune inflammatory process. (Grevich S., et al., 2017).

Differential diagnoses considered in this case included systemic lupus erythematosus (SLE) and Henoch–Schönlein purpura (HSP). SLE was excluded due to negative ANA and anti-dsDNA results, while HSP was excluded because the patient did not present with palpable purpura, renal involvement, or abdominal pain. (Ozen S., et al., 2020) Taken together, the clinical features and investigations strongly supported the diagnosis of sJIA.

The pathogenesis of sJIA is complex, involving dysregulated cytokine activity, particularly interleukin-1 (IL-1) and interleukin-6 (IL-6), which play major roles in systemic inflammation, rash, and arthritis. (Ravelli A., 2015; Schett G., 2016; Martini A., 2012). Methotrexate was selected as the main disease-modifying antirheumatic drug (DMARD) for this patient, combined with folic acid and topical therapy to manage cutaneous symptoms. Conventional DMARDs such as methotrexate and leflunomide remain the standard options, while biologic DMARDs targeting IL-1 and IL-6 are highly effective and well tolerated for sJIA, though availability remains limited in many healthcare settings. (Piette WW., 2019; Onel KB., et al., 2022). The prognosis of sJIA is variable. Early initiation of disease-modifying therapy is associated with better outcomes in terms of joint preservation and systemic control.(Grevich S., et al., 2017)/ In this patient, clinical improvement was noted after methotrexate therapy, although residual hyperpigmentation and joint symptoms persisted.

CONCLUSION

Systemic juvenile idiopathic arthritis (sJIA) is a rare but severe subtype of juvenile idiopathic arthritis that requires careful diagnostic evaluation due to its overlapping features with other autoimmune and inflammatory diseases. In this case, the diagnosis of sJIA was established based on the presence of intermittent high fever, persistent symmetrical arthritis, and a non-pruritic maculopapular rash. The patient was managed with systemic therapy using methotrexate and folic acid, alongside supportive dermatological treatment with emollients and topical corticosteroids to control skin manifestations. Early recognition and prompt initiation of appropriate therapy are essential to reduce disease activity, prevent long-term complications, and improve functional outcomes.

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