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**Case Report**

# Juvenile dermatomyositis in a 6 year old boy: A rare case

**Olivia Awwalin Sunarto, Rahmadewi, Putri Hendria Wardhani, Yuri Widia, Trisiswati Indranarum, Irmadita Citrashanty, Damayanti, Sawitri Sawitri, Dwi Murtiastutik**

Department of Dermatology and Venereology, Faculty of Medicine Universitas Airlangga/ Dr. Soetomo General Academic Hospital, Surabaya, Indonesia.

**Abstract** Juvenile Dermatomyositis (JDM) is a systemic capillary vasculopathy and idiophatic inflammatory myopathies (IIM) in children. It is a rare disease with incidence in the United States 3 per million children per year. A 6-year-old child complained about red spots on his right and left knees. He had red spots which became white spots and thickening skin on his knuckles since 4 months ago before hospitalized. Redness occurred around the eyes and his back since 2 months ago. He had muscle weakness especially on his limb since 2 months ago. JDM has peak occurrence during infancy with ages of 5 and 15. Genetic, environmental and immunological factors are believed to contribute to the development of JDM. The clinical symptoms of this condition are varied, with different levels of skin, muscle, gastrointestinal and cardiac. The goals of treatment are to control inflammatory myositis and prevent complication. It is important for dermatologist to know this disease clearly, so that can make a proper diagnosis and carry out appropriate management.

***Key words***
Juvenile Dermatomyositis; Vasculophaty disease; Rare disease; Systemic autoimmune disease.

## Introduction

Juvenile Dermatomyositis (JDM) is a systemic capillary vasculopathy disease and idiophatic inflammatory myopathies (IIM) in children. It is a rare disease with an incidence in the United States 3 per million children per year. JDM commonly affects children with age between 4 and 10 years old. There is a 2:1 ratio of JDM in girls compared to boys.[1]

Ten percent of patients experience muscular symptoms before to the development of skin abnormalities, and thirty to fifty percent of patients have cutaneous disease three to six months before developing myositis. The precise pathogenesis of JDM is still not entirely understood. Some studies stated that it could be linked to genetics, an immune response to a virus, an adverse drug reaction, exposure to sunlight or internal malignancy.[2]

The first sign of JDM is often a skin rash. Heliotrope rash, Gottron's papules, and periungual erythema with anomalies in the capillary loops are examples of phagonomonic rash. This may be misdiagnosed as allergies. Moreover, JDM is characterized by symmetrical proximal muscular weakness. It is likely a weakening in all the muscular groups, but it is particularly noticeable in the neck flexors, hip flexors, and shoulder girdle. The muscles that are impacted could be sore and swollen.[1]

Early, aggressive treatment of JDM associated with a better prognosis. Corticosteroids in combination with another immunosuppressive medication, commonly methotrexate is the mainstay of treatment.[1] Some patients have prolonged cutaneous disease, but they have resolution of their muscle involvement after

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**Address for correspondence**
Dr. Rahmadewi
Jalan Prof. Dr. Moestopo No. 6-8 Surabaya,
East Java, 60286, Indonesia.
Ph: +62 812-3572-112
Email: rahmadewi@fk.unair.ac.id

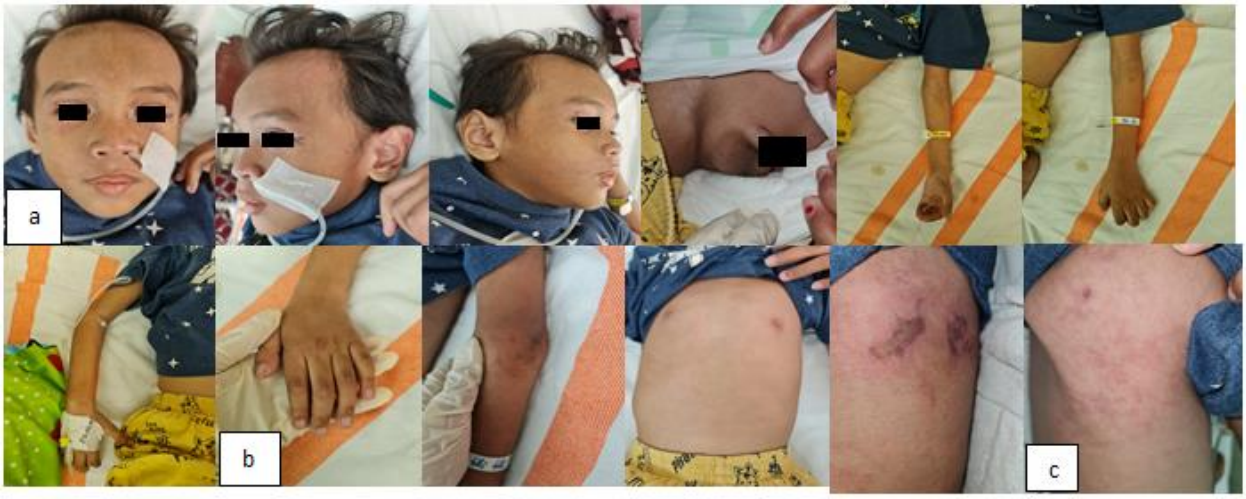


**Figure 1** Clinical findings on November 22nd, 2023 (1st day) of examination. a. Pink erythema to hypopigmentation, edema on upper eyelid, and erosion (Helioptrope sign). b. Light pink, ill-defined papules over the proximal and distal interphalangeal joints (Gottron papules). c. Thoracalis posterior region, we found pink to red patches with minimal white squama (Shawl sign).

therapy. JDM has a diagnostic and therapeutic challenge due to one of rare disease.[4] This case description aims to help recognize and confirm the diagnosis in JDM disease.

## Case report

A 6-year-old children was consulted by pediatric department with suspect Chronic inflammatory demyelinating polyneuritis (CIDP) on November 22nd, 2023. Patient complained about red spots on his right and left knees, moreover he had red spots which became white spots and thickening skin on his knuckles since 4 months ago before hospitalized at Soetomo Hospital. He also complained of the wounds on the back of right and left knees. The wounds started with blisters and then ruptured. He felt pain on his wounds. Redness occurred around the eyes and his back since 2 months ago. He did not have complaint of itchy and burning sensation.

Furthermore, he had muscle weakness especially on his limb. Initially, he had difficulty to raise his arms and legs. After that, he was harder to move, walk and sit. It was getting worse since 1.5 months ago. History of seizures, loss of consciousness and trauma were denied by his mother. The patient can urinate and defecate with full awareness. He had no history of headache, fever, difficult to swallow food, nausea, vomiting, bloody stools and weigh loss. He had no history of hypertension, arrhythmia, allergic rhinitis, asthma, food allergy, drug allergy, and malignancy. His family did not have similar complaint, autoimmune disease, food allergy, drug allergy, allergic rhinitis, and asthma. He did not apply oil, herbal, and any medications. He got therapy from pediatric department with D5 ½ NS infusion 1000ml/24 hours, Ampicilline sx 450mg every 6 hours for 7 days, Paracetamol orally 200mg every 6 hours and Methylprednisolone 12mg every 8 hours (2mg/weight/day).

On physical examination, the patient's general condition was weak, vital signs were within normal limits. On head and neck examination, anemia, icterus, cyanosis, and dyspnea were not found. The result of thorax photo was lung inflammatory suspect specific process, but his heart was normal. Abdominal examination was within normal limits, bowel sounds were audible and normal, liver and spleen were not palpable. There was no oedema in the upper and lower extremities, the extremities felt warm, dry and red. No enlarged lymph nodes.

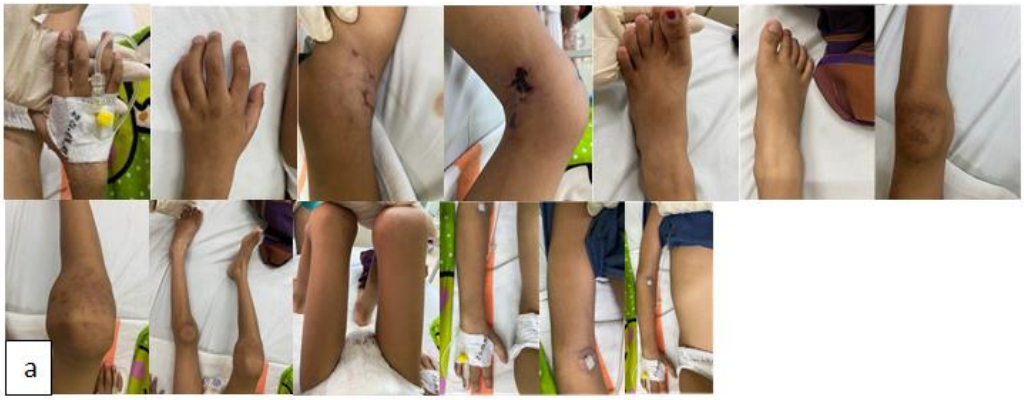


**Figure 2** Clinical findings on November 23rd, 2023 (2nd day) of examination.
a. ill-defined violaceous erythema and hyperpigmentation (Gottron sign).

Dermatological examination of bilateral periorbital region revealed pink erythema to hypopigmentation, edema on upper eyelid, and erosion (Helioptrope sign) **(Figure 1a)**. Fingers of bilateral hands revealed light pink, ill-defined papules over the proximal and distal interphalangeal joints (Gottron papules) **(Figure 1b)**. Posterior thorax region revealed pink to red patches with minimal white squama (Shawl sign) **(Figure 1c)**. On bilateral knees, ill-defined violaceous erythema and hyperpigmentation (Gottron sign) were found **(Figure 2)**.

Laboratory examinations were performed for the patient. Complement C3/C4 levels within normal limits, ANA test and anti ds-DNA results were negative. He had leukocytosis (17.570), high SGOT level (118), high SGPT level (57), hemoglobin 11.6 g/dL, platelet 212.000, creatinine 0.5, BUN 14.6, random blood glucose 115, blood cultur showed there were no bacterial and fungal. Chest x-ray did not show any abnormalities.

The result of histopathology examination showed skin biopsy tissue section covered by atrophic epidermis, the rete ridges are partially flattened. Basal cell hypopigmentation was visible, no basal cell vacuolar degeneration was seen. In the dermis, lymphocyte infiltration was seen at the dermoepidermal junction and superficial dermis. The conclusion revealed that can be found in dermatomyositis or cutaneous lupus erythematosus **(Figure 3)**.

Based on the history, physical examination and supporting examination, the patient was diagnosed with juvenile dermatomyositis. Pediatric department gave D5 ½ NS infusion 1000ml/24 hours, Ampicilline sx 450mg every 6 hours for 7 days, Paracetamol orally 200mg every 6 hours if patient got fever, methylprednisolone orally 12 mg every 8 hours until patient came to outpatient clinic. The patient was consulted to medical rehabilitation doctor for strength training his muscle. Dermatology and venereology department gave

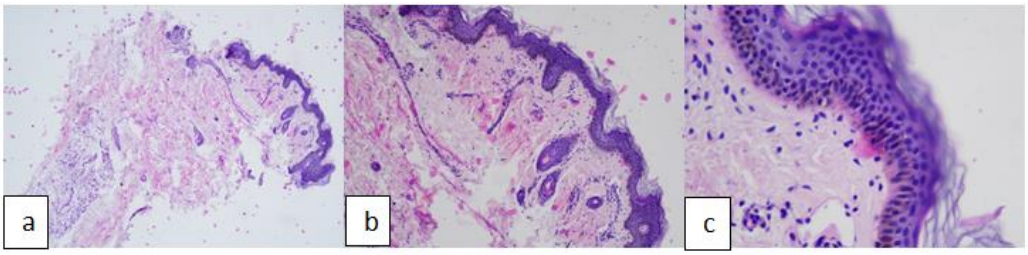


**Figure 3** Histopathology examination results on December 1st, 2023. a. Atrophic epidermis, the rete ridges are partially flattened was found (40x), b. Basal cell hypopigmentation was visible, no basal cell vacuolar degeneration was seen (100x), c. Lymphocyte infiltration was seen at the dermoepidermal junction and superficial dermis (400x).

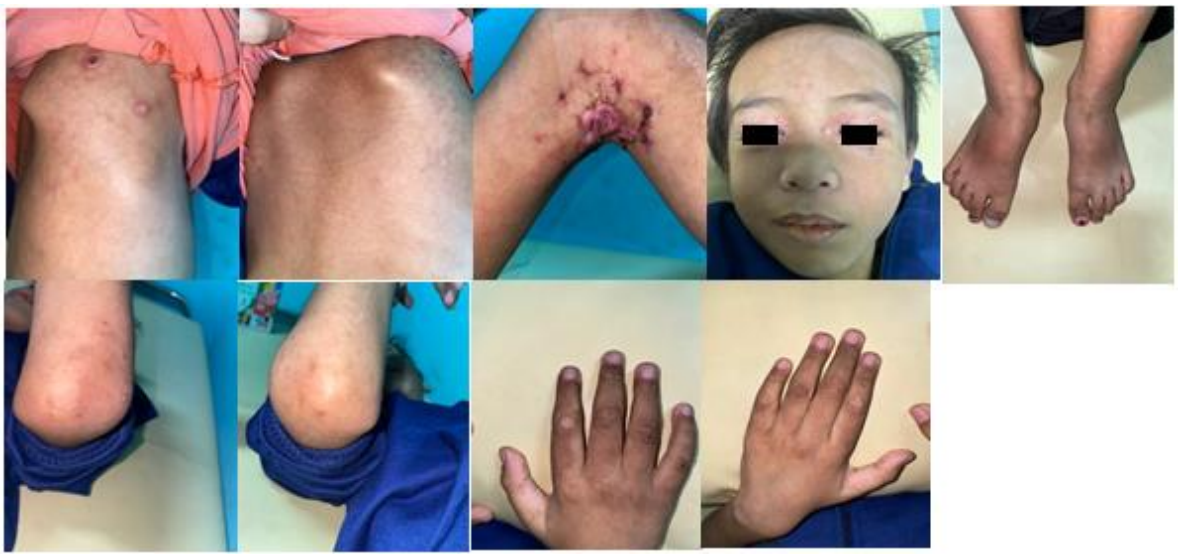

**Figure 4** Clinical findings on December 15th, 2023 (25th day) of examination. The red spots on his back have decreased.

mometasone furoate 0.1% cream every 12 hours on red spots and treated the wound with duoderm gel, framycetine and covered by sterile kassa for his right feet. On the 23rd day of evaluation, the red spots on his back have decreased **(Figure 4)**. We thought several differential diagnoses such as contact dermatitis, angioedema, cutaneous lupus erythematous, polymyositis, systemic sclerosis, and mixed connective tissue disease.

## Discussion

Idiopathic inflammatory myopathies (IIMs) are a diverse set of complicated illnesses marked by persistent and symmetrical muscle weakening and injury, along with the participation of other organ systems. Dermatomyositis (DM) is one of IIM disease. The etiology is characterized by microangiopathy that specifically impacts the skin and muscle tissues.[1] JDM is characterized by two periods of peak occurrence: one during infancy which is called juvenile DM (JDM), typically between the ages of 5 and 15, and another during maturity, typically between the ages of 40 and 60.[2,3]

The development of DM involves multiple factors, is intricate, and not fully comprehended. Genetic, environmental, and immunological factors, together with the newly identified autoantibodies mentioned later, are believed to contribute to the development of both adult DM and JDM. In children, UV radiation has been linked to the development of JDM and the presence of anti-TIF1 antibodies. There was a frequent occurrence of past interactions with diseased animals.[1,3,4]

The typical age at which JDM symptoms first appear and at which the disease is officially diagnosed is between 5.7 and 6.9 years and 7.4 and 7.7 years, respectively. There is a belief that a younger age when symptoms first appear, a higher age when the condition is officially diagnosed, or delays in receiving a diagnosis may be linked to a less favourable outlook for the patient.[5,6]

The most prevalent symptoms of JDM include heliotrope discolouration and the presence of Gottron's papules. Additionally, individuals with JDM often experience muscle weakness in the proximal areas. Cutaneous calcinosis, which forms over areas of pressure, is a typical type of lesion seen in JDM. Additional lesions that are

more frequently observed in JDM involve the formation of calcifications beneath the skin, which might protrude to the surface and lead to the development of ulcers and infections.[7,8]

In our case, a 6-year-old children consulted with 4 months history of red spots on his right and left knees. He also complained of the wound on the back of right and left knees. The wound on the back of his knees started with blisters and then ruptured. He felt pain on his wound. He had a complaint of red spots which became white spots and thickening skin on his knuckles since 4 months ago. Redness occurred around the eyes and his back since 2 months ago. Patient also had muscle weakness especially on his limb which was getting worse since 1.5 months ago.

Involvement of inflammation in muscle of JDM patients may be severe, typically results in weakness, loss of endurance, and alterations in physical function. Older children might be better to quantify and complain of early weakness, however, younger patients may present as to be carried more often or difficulties in getting down or up off the floors, dressing, combing hair, climbing stairs and moreover, choking with drinking liquids or voice changes. Muscle weakness is the primary feature of the JDM, usually progressive, symmetric, non-selective and greatly affects proximal muscles dysphagia, respiratory problems, and aspiration pneumonia can occur due to the involvement of respiratory and oropharyngeal muscles in severe cases.[3,5,9,10]

The patient's general condition was weak with normal vital signs and physical examination results. Dermatological examination left and right eyelids revealed pink erythema to hypopigmentation and edema with erosion, which was consistent with Helioptrope sign. Examination on both hands revealed light pink, ill-defined papules over the proximal and distal interphalangeal joints, which was consistent with Gottron papules. On posterior thorax, pink to red patches with minimal white squama was found, which was cosistent with Shawl sign. Both knees had ill-defined violaceous erythema and hyperpigmentation, which was consistent with Gottron sign.

Elevated blood muscular enzymes levels are indicative of muscle involvement. Serum CK is a highly sensitive muscle enzyme that is released into the bloodstream when muscle damage occurs, particularly during the early stage of the disease. In the active stage of the disease, levels of serum inflammatory biomarkers such as Erythrocyte Sedimentation Rate and C-reactive protein might be elevated. LDH and ALP return to normal levels 2.53 and 3.68 months, respectively, following the onset of symptoms. CK and aldolase, on the other hand, take approximately 4.5 months to normalize. Thus, whereas CK is commonly used in international categorization criteria, its reliability diminishes after approximately 4 months from the onset of symptoms to the clinical evaluation.[10,11]

In acute stage, the erythematous eruption reveals mild hyperkeratosis and epidermal atrophy with effacement of the rate ridge. Basal cell liquefactive degeneration is a common occurrence, and occasionally cytoid structures could be detected. Evidence upper dermal edema is present. Additionally, the presence of melanophages could be observed. There is an occurrence of activated T-lymphocytes and macrophages infiltrating the perivascular area. Mucin deposits, found in the dermis, are indicative of dermatomyositis, although they are not diagnostic.[3,12]

Gottron papules exhibit hyperkeratosis, acanthosis, and minor papillomatosis. The manifestations of interface dermatitis are observed as previously described. Lesional skin could be examined using direct

immunofluorescence to detect the presence of immunoglobulin and complement deposits at the junction between the dermis and epidermis.[3,12]

In our case, laboratory examinations revealed normal level of Complement C3/C4 and negative results ANA test and anti ds-DNA. The patient had leukocytosis, and high AST and ALT The result of histopathology examination showed skin biopsy tissue section covered by atrophic epidermis, the rete ridges are partially flattened. Basal cell hypopigmentation was visible, no basal cell vacuolar degeneration was seen. In the dermis, lymphocyte infiltration was seen at the dermoepidermal junction and superficial dermis. Based on the history, physical examination and supporting examinations, the patient was diagnosed with dermatomyositis.

Management of JDM is complex and warrants a multidisciplinary approach including physiotherapists, dermatologist, specialist nurses and paediatric rheumatologists, with other specialists, as needed, for example, cardiologist/pulmonologist. The mainstay of therapy is high-dose corticosteroid initially in combination with disease modifying drugs like MTX or cyclosporin A (CsA). In 2010, the Childhood Arthritis and Rheumatology Research Alliance (CARRA) reached consensus on treatment plans for moderately severe JDM for the first two months after diagnosis that include a combination of steroid (intravenous methylprednisolone followed by oral prednisolone or high-dose oral prednisolone alone) and MTX ± intravenous immunoglobulin (IVIG). The combination of steroids and MTX had the best outcome for efficacy and safety.[13]

The conventional treatment for systemic and muscular illness in DM involves the use of systemic corticosteroids at a dosage of 1 mg/kg/day, followed by a gradual reduction in dosage. The maximum dosage is continued for a minimum of 2-4 weeks and until there is clear indication of clinical progress. If the condition exhibits a quick response, the dosage of corticosteroids can be gradually reduced to 50% of the initial amount within a span of 6 months. Using this methodology, a significant proportion of patients, specifically 75%, achieve a state of being free from clinical disease during a period of 2 to 3 years. In cases of advanced or recalcitrant disease, treatment options such as pulse prednisone, split-dose prednisone at dosages higher than 1 mg/kg/day, or medications that reduce the need for steroids might be started.[14]

In our patient, we gave both systemic (methylprednisolone orally 12 mg every 8 hours for 14 days) and topical corticosteroid (mometasone furoate 0.1% cream every 12 hours) with wound care and strength training for his muscle by medical rehabilitation doctor. On the 23$^{rd}$ day of evaluation, there was no clinically significant improvement. The red spots on the back have decreased.

JDM is very poor prognosis, with one third of patients recovering without complications, one third developing disability and one third dying. The mortality rate dropped to 1%, after many clinician use corticosteroid treatment.

**Conclusion**

Juvenile dermatomyositis is a rare multisystem inflammatory disease typically affecting children between the ages of 5 and 15. It has cutaneous manifestation, muscle disorders, and systemic involvement. The cutaneous manifestations tend to be chronic, debilitating and recalcitrant to therapy.

The diagnosis is mostly dependent on physical examination. JDM has diagnostic and

therapeutic challenge. It is important for dermatologist to know this disease clearly, so that they can make a proper diagnosis, carry out appropriate management, prevent debilitating effect and sequele in the future. This disease has negatively impact for quality of life.

***Declaration of patient consent*** The authors certify that they have obtained all appropriate patient consent.

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***Conflict of interest*** Authors declared no conflict of interest.

***Author's contribution***

**OAS:** Identification and management of the case, manuscript writing, final approval of the version to be published.

**R:** Diagnose and management, critical review, final approval of the version to be published.

**PHW, YW, IC, D, S, DM, TI:** Diagnose and management of the case, critical review, final approval of the version to be published.

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