



A Case of Juvenile Dermatomyositis Responding to Methotrexate and Steroid

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ABSTRACT

A 4-year-old patient presented with skin rash and muscle weakness. She was diagnosed with juvenile dermatomyositis based on Bohan and Peter criteria and laboratory result. The treatment consisted of steroid combined with methotrexate. The response was good.

Keywords: Juvenile dermatomyositis, methotrexate, steroid

ABSTRAK

Pasien anak perempuan berusia 4 tahun dengan keluhan bercak kemerahan di kulit dengan kelemahan otot. Pasien didiagnosis *juvenile dermatomyositis* berdasarkan kriteria *Bohan and Peter* dan hasil laboratorium. Pasien mendapat terapi steroid dikombinasikan dengan *methotrexate*, menunjukkan hasil baik. **Prastiya Indra Gunawan, Darto Saharso. Juvenile Dermatomyositis yang Berespons terhadap Methotrexate dan Steroid - Laporan Kasus**

Kata kunci: *Juvenile dermatomyositis, methotrexate, steroid*

INTRODUCTION

Juvenile dermatomyositis (JDM) is the common idiopathic inflammatory myopathies that occurs during childhood. The incidence is estimated 2-3 per million children per year.^{1,2} Most cases of JDM occur between 5 and 14 years old. Females are more commonly affected. The etiology of dermatomyositis is unknown, but autoimmunity is suspected to be the underlying cause of inflammation. In the last few decades, mortality has significantly declined, and there has been a progress in functional outcome due to earlier diagnosis and more effective treatment.^{1,2} We report a case focusing on diagnostic and management approach.

CASE REPORT

A 4-year-old female patient presented with erythema of nasal bridge, cheeks, upper and lower eyelids lasted for four months. The complaint did not diminish with topical anti-allergy treatment. Two months later, she developed confined erythematous papular lesions of metacarpophalangeal, proximal part of interphalangeal, and distal part of interphalangeal joints. Simultaneously she suffered from weakness and difficulty to get up from the bed and to run. No fever was

observed. Neurological examination showed weakness of proximal muscle. No sign of arthritis and calcinosis. Skin changes typically involved erythematous rash with purplish discolorisation along nasal bridge, cheeks as well as periorbital regions (heliotrope rash). There were papular, erythematous, and scaly lesions over the knuckles (Gottron's sign) (Figure 1). CPK level was 354 IU/mL. ANA test, anti-ds-DNA, CRP, and LE cell phenomenon were negative. Muscle biopsy revealed perivascular infiltration of inflammatory cells and perivascular atrophy (Figure 2). EMG showed myopathy. Methylprednisolone 18 mg in three divided doses was administered.



Figure 1. Gottron sign before treatment

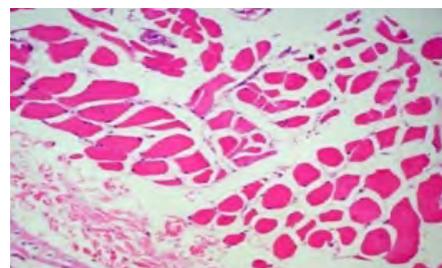


Figure 2. The muscle biopsy shows perivascular infiltration of inflammatory cells and perivascular atrophy.

Within two weeks of treatment, the patient showed remarkable improvement (Figure 3). A few skin lesions manifested as generalised erythema and mild oedema of skin were persisted. Addition of methotrexate as steroid-sparing agent weekly started at 7.5 mg and then increased 2.5 mg weekly to a maximal dose of 25 mg.



Figure 3. Gottron sign after treatment

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On follow-up after four weeks of treatment, the patient showed an excellent response. The muscle strength gradually improved as well as the cutaneous manifestations were moderately disappeared. The methotrexate was well tolerated. No side effect was significantly observed. No mood changes, weight gain, or recurrent infections developed. The treatment resulted in neurological improvement.

DISCUSSION

The rash in JDM may precede or follow the onset of proximal muscle weakness. The characteristic rash is heliotropic or violaceous, occurring most prominently on the eyelids. Erythema can occur over the upper fragments of the body (shawl sign) and extensor surfaces of arms and legs.³ In this patient, the characteristic of rashes compared with clinical presentation by Sunkureddi, *et al*, is shown in table.^{1,4}

Table. The characteristic of rashes of the patient

	<i>The case</i>
Heliotrope sign	+
Gottron's sign	+
Shawl sign	-
V-sign	-
Poikiloderm vasculare atrophicans	-

Bohan and Peter set four criteria for the diagnosis of JDM three of the four criteria must be fulfilled for the definite disease, two of four for probable diseases and one of four for possible diseases.¹ The criteria including symmetric proximal muscle weakness, elevation of serum skeletal muscle enzymes, the electromyographic triad of short, small, polyphasic motor unit potentials; fibrillations, positive sharp waves, and insertional irritability;

and bizarre, high frequency repetitive discharges, muscle biopsy abnormalities of degeneration, regeneration, necrosis, phagocytosis, and an interstitial mononuclear infiltrate. Typical skin rash of DM including a heliotrope rash and Gottron's sign/papules. The patient had characteristic rash manifested as heliotrope and Gottron sign, elevated CPK level, evidence of muscle weakness and myopathy; diagnosed as definite juvenile dermatomyositis (JDM).

As dermatomyositis is not common, there have been few randomized controlled trials, and those that have been completed enrolled small number of patients. Subsequently, optimum therapy has not been defined adequately. Corticosteroids are the mainstay of therapy and accepted as first-line management for JDM.⁵ Treatment typically begins with high-dose corticosteroid. In this patient; methylprednisolone treatment showed a good response. Although controlled studies have not been published, initial treatment with high dose of corticosteroid is recommended. Approximately 90% of patients show partially improvement with corticosteroid therapy, and 50-75% of patients would achieve complete remission.⁴ There is controversy surrounding the best route of administration, dosing regimen, duration and parameters to monitor the treatment.²

During corticosteroid tapering, other immunosuppressive agents, such as azathioprine, methotrexate, cyclophosphamide, and cyclosporine could be used as steroid-sparing agent to relieve symptoms relief.^{4,6} Jamuar reported 3 cases presented classically with skin and muscle

involvement and were treated with high doses of prednisolone. Upon tapering the dose of prednisolone, they had recurrence of symptoms and were started on subcutaneous methotrexate. All patients responded well to the treatment.⁷ The pediatric rheumatology group in Toronto compared outcomes in 31 children with JDM, who were received methotrexate as first line treatment and 22 similar children (control). All patients received 2 mg/kg/day of prednisone administered in three doses for the first six weeks, then in once daily dose. If the disease was under control, the dose was tapered by roughly 10% every second week. Patients received methotrexate at 10–20 mg/m² body surface area/week. The duration, cumulative dose, and toxicity of corticosteroid treatment were significantly reduced in the methotrexate group.⁸ In this patient, methotrexate-steroid sparing agent for symptom relief was instituted after two weeks steroid treatment and it successfully reduced the symptoms. A placebo-controlled trial of azathioprine in myositis did not show any significant evidence at 3 months. Cyclosporine has also been reported with success in combination with immunoglobulin.⁹

The overall prognosis for survival is improved with the use of corticosteroids. The prognosis is good for most patients with dermatomyositis without associated malignancy.⁴

CONCLUSION

Corticosteroids are still the mainstay of medication for juvenile dermatomyositis. Combination of steroid and methotrexate treatment gave a remarkable response in this patient.

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