Juvenile Dermatomyositis in Thai Children

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Abstract

Juvenile dermatomyositis is a rare, chronic multisystemic inflammatory disorder of unknown etiology, characterized by a typical skin rash and proximal muscle weakness. A retrospective study from the medical records of patients diagnosed as juvenile dermatomyositis was performed at Queen Sirikit National Institute of Child Health from 1988 to 1998. There were seven cases of juvenile dermatomyositis diagnosed according to the criteria of Bohan and Peter. Six cases were female and one case was male. The age of diagnosis ranged from 2.5 years to 11 years. (mean age was 7±3.6 years). The presenting symptoms were muscle weakness (6 cases), muscle pain (2 cases) and skin rashes (4 cases). All of the patients developed proximal muscle weakness of the lower extremities varying from grade 3 to grade 4. The cutaneous manifestations were heliotrope signs (6 cases), gottron's papules (2 cases), photosensitivity (2 cases) and calcinosis cutis (4 cases). Electromyography (EMG) was performed in 6 cases and revealed typical change of myopathic type. Elevated muscle enzymes were noted in all cases. Muscle biopsy was performed in 6 cases and was compatible with myositis. Oral prednisolone (1-2 mg/kg/day) was given in 6 cases and the muscle weakness improved. There was no mortality in this study. Four cases developed calcinosis cutis 1 to 3 years after muscle weakness and did not respond to any treatment. In conclusion, juvenile dermatomyositis is a disease which causes chronic disability in children. Early diagnosis and treatment can prevent morbidity and mortality. Calcification at the skin usually occurs after the onset of muscle weakness several months to years after diagnosis.

Key word : Juvenile Dermatomyositis, Thai Children

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Juvenile dermatomyositis (JDM) is a chronic inflammatory disorder that primarily affects the skin, striated muscles and occasionally internal organs. The disease is rare in children with an incidence of 1 to 3.2 cases per million children[1-4]. It is more frequent in Caucasians children compared to Asian children[2-4]. The clinical characteristics are symmetrical proximal muscle weakness and typical skin rash. The cause of juvenile dermatomyositis is unknown but the pathogenic mechanism is thought to be immune mediated muscles and skin damage[5]. Environmental factors and infectious agents such as influenza A or B, hepatitis B and Toxoplasma gondii have been associated with JDM [2,5]. Early diagnosis and treatment of JDM can prevent morbidity and mortality.

Several articles have described the various clinical manifestations, outcomes and prognosis of JDM[6-13]. However, there are no reports of JDM in Thai children.

This report includes the clinical manifestations, laboratory investigations, treatment and outcome of 7 children with JDM seen at Queen Sirikit National Institute of Child Health between 1988 and 1998. We also compared our cases with previous reported series.

MATERIAL AND METHOD

A retrospective study was conducted from the medical records of patients diagnosed with JDM at Queen Sirikit National Institute of Child Health from 1988 to 1998. All of these patients met the diagnosis criteria for JDM proposed by Bohan and Peter (Table 1)[14,15]. Initial clinical signs and symptoms, laboratory findings, treatment, outcome and complications were reviewed.

RESULTS

Clinical manifestations (Table 2)

Seven cases were diagnosed with JDM at Queen Sirikit National Institute of Child Health over a 10-year period. All patients had definite criteria of JDM proposed by Bohan and Peter. Age at onset ranged from 2.5 years to 11 years. The mean age at onset was 7±3.6 years. There were 6 female and 1 male patient.

Muscle weakness was the most common presenting symptom in 6 cases (86 %). Other symptoms were skin rashes 4 cases (57%), muscle pain 2 cases (28 %) and edema 2 cases (28 %). The onset of muscle weakness was insidious ranging from 2 weeks to 1 month before admission and muscle power test was grade III and grade IV. Heliotrope periorbital edema (Fig. 1) was the most common cutaneous manifestation (6 cases). Gottron's papules (Fig. 2) and photosensitivity rashes (Fig. 3) were each seen in 2 cases (28%). Calcinosis cutis (Fig. 4) seen in 4 cases (57%) developed from 1 year to 5 years after onset of muscle weakness.

Laboratory findings (Table 2)

Elevated muscle enzymes (CPK, LDH) were observed in all 7 cases. Abnormal antinuclear factor was found in only one case. Muscle biopsies were performed in 6 out of 7 cases and all cases were compatible with myositis. Electromyography were done in 6 cases and revealed typical myopathic pattern.

Treatment and outcome (Table 3)

All cases were treated with 1-2 mg/kg/day oral prednisolone in the initial phase and then tapered off to a maintenance dose depending of the clini-

Table 1. Diagnostic criteria for dermatomyositis (Bohan and Peter)[14,15].

1) Symmetrical proximal muscle weakness, progressing weeks to months, with or without dysphagia and respiratory muscle weakness.
2) Muscle biopsy evidence of inflammatory myopathy.
3) Elevation of muscle enzymes.
4) Electromyographic features of myopathy.
5) Cutaneous eruption of dermatomyositis such as a heliotrope rash or Gottron's sign.

Definite : Fulfill 3 criteria + rash.
Probable : Fulfill 2 criteria + rash.
Possible : Fulfill 1 criteria + rash.

Criteria # 5 must be one of the stated number of criteria in patients with definite, probable or possible dermatomyositis

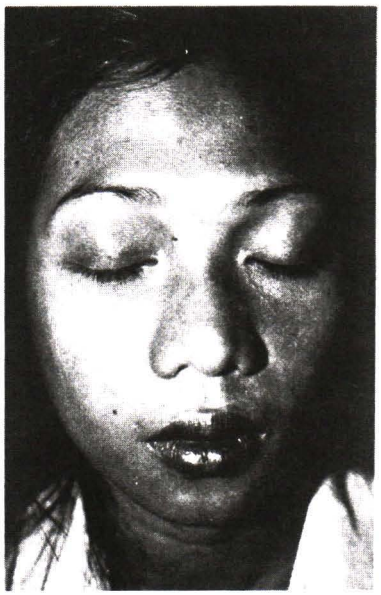

Fig. 1. Patient no 6 at age 15 years shows heliotrope erythema of both upper eyelids.

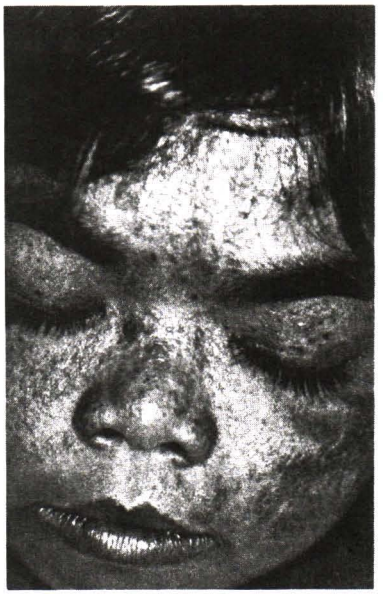

Fig. 3. Photosensitivity rash in juvenile dermatomyositis.

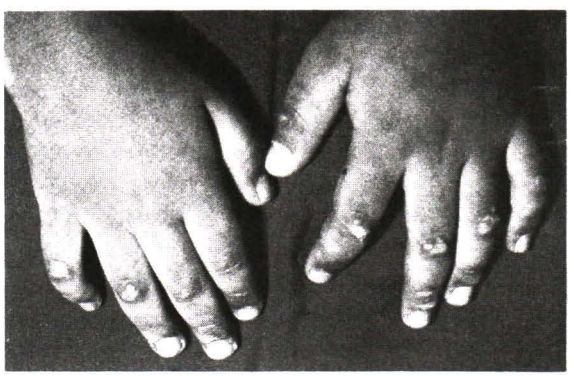

Fig. 2. Gottron's papules over both hands.

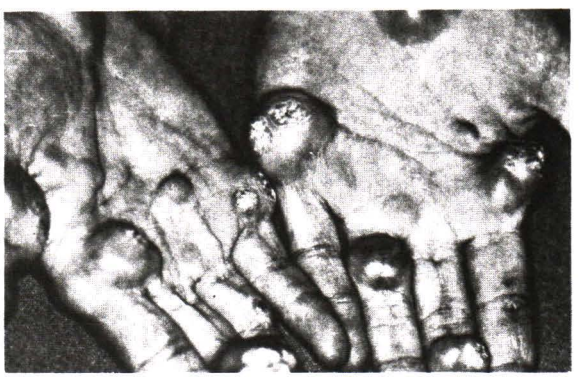

Fig. 4. Calcinosis cutis of both palms.

cal symptoms. Average duration of corticosteroid treatment was at least 1 year. Six patients were followed-up for an average of 2 years. (range from 2 years to 12 years). The longest time of follow-up was 12 years. The outcome was good in 2 cases with complete recovery without muscle weakness. Four cases needed another short course of prednisolone after cessation of initial treatment due to relapse of muscle weakness. One case was lost to follow-up. There were no deaths in this study.

Complications (Table 3)

There was no associated malignancy or pharyngeal involvement in this study. Four cases

Table 2. Clinical and laboratory findings of juvenile dermatomyositis.

Patient	Sex	Age of onset (years)	Presenting symptoms	Muscle weakness	Skin			Muscle enzymes CPK, LDH	ANA	Abnormal EMG	Muscle biopsy
					Gottrons	Heliotrope	Photo				
1	F	2.5	Rash 1 year Weakness 2 weeks	Lower Gr III/V	+	+	+	+	-	+	Myositis
2	F	2.5	Rash 1 year Weakness 3 weeks	Lower Gr III/V	-	-	+	+	-	+	ND
3	M	5	Weakness 1 month Edema 1 month	Lower Gr III/V	-	+	-	+	-	+	Myositis
4	F	9	Weakness 1 month Facial rash 1 week	Upper Gr III/V Lower Gr III/V	-	+	-	+	-	+	Myositis
5	F	9	Weakness 1 month Muscle pain 1 month	Lower Gr. III/V	+	+	-	+	+	+	Myositis
6	F	10	Rash 1 year Weakness 3 years	Upper Gr. IV/V Lower Gr. IV/V	-	+	-	+	-	ND	Myositis
7	F	11	Edema 1 month Muscle pain 1 month	Upper Gr. IV/V Lower Gr. IV/V	-	+	-	+	-	+	Myositis

F = female, M = male, ANA = antinuclear antibody, Photo = Photosensitivity rash, ND = Not detected, + = positive, - = negative

(57%) developed calcinosis cutis within 1 to 5 years of muscle weakness. Treatment of calcinosis cutis was unsatisfactory. Two cases were treated with aluminum hydroxide and the other two with surgical excision.

DISCUSSION

The clinical characteristics including age at onset and symptoms of our patients were similar to those described in previous studies[6-13]. There was a higher female to male ratio (6:1) than other studies. This difference may be the consequence of a relatively small study group.

The degrees of muscle weakness of our patients were mild with an insidious onset, ranging from grade 3 to 4. This could be why the patients came to see the doctor quite late after the onset of illness. The skin rashes were the first presenting symptoms in 4 cases (57%). Hiketa found that there was a tendency for cutaneous manifestations to appear first with muscular manifestations arising later [8]. Our findings also emphasized that the recognition of the skin lesions may lead to early intervention before extensive myositis had evolved.

The mainstay of treatment of juvenile dermatomyositis was systemic corticosteroids. However, there was no consensus about the dose and duration of corticosteroids[17-19]. Most of the authors favored the use of initial high dose corticosteroids therapy until the muscle enzymes had normalized, followed by low doses for an extended period[19]. Intravenous pulse methyprednisolone has been reported with good response[20]. Those patients who did not respond adequately to corticosteroids were treated with immunosuppressive drugs including methotrexate, azathioprine or intravenous immunoglobulin[2,18].

In our study, all cases were treated with oral prednisolone in the initial phase and then tapered off to a maintenance dose depending on the muscle weakness. The responses to treatment varied, with complete recovery in 2 cases, partial recovery in 4 cases and 1 case lost to follow-up. Because of the small number of patients, we could not compare the data to correlate between the clinical course, drug treatment and outcome.

Another important treatment for muscle weakness is extensive physical therapy to prevent joint contracture and muscle atrophy. Topical steroid

Table 3. Treatment and outcome of juvenile dermatomyositis.

Patient	Treatment	Duration	Follow-up	Outcome	Complication
1	Prednisolone	1 year	3 years to now	Partial recovery	Calcinosis cutis after weakness 5 years
2	Prednisolone	1 year	4 years to now	Partial recovery	Calcinosis cutis after weakness 1 year
3	Prednisolone	1 year	3 years	Partial recovery	Calcinosis cutis after weakness 1 year
4	Prednisolone	1 year	5 years	Good recovery	No
5	Prednisolone	Lost follow-up	Lost follow-up	Lost follow-up	Not known
6	Prednisolone	1 year	12 years	Partial recovery	Calcinosis cutis after weakness 3 years
7	Prednisolone	1 year	2 years to now	Good recovery	No

therapy should be used for the pruritus and inflammatory erythematous skin lesions. Routine sunscreen should be used for photosensitive patients[2,22].

In general, the prognosis of juvenile dermatomyositis is good if there is no systemic involvement. The mortality rate is low, about 3 per cent. Patients usually die from pharyngeal, respiratory, gastrointestinal or cardiac involvement[21,22].

Malignancy associated with juvenile dermatomyositis is rare in comparison with adult dermatomyositis which occurred in 30 per cent. No cases of juvenile dermatomyositis complicated by malignancy or pharyngeal involvement were found in this study and there were also no deaths.

Poor prognosis of juvenile dermatomyositis is associated with initial treatment with low dose prednisolone, late onset of treatment, recalcitrant disease and pharyngeal involvement. Two thirds of children in this group had severe complications of calcium deposition[10,23].

Calcinosis cutis is a common complication of juvenile dermatomyositis and occurs in 40-70 per cent of the patients[2,21,23]. The calcium can deposit in muscle, subcutaneous tissue and in fascia. It can cause more long term disability than the acute inflammatory myopathy itself. Pain, limitation of motion, skin ulceration, abscess formation, fever and systemic symptoms can result[23]. The exact mechanism of calcification is unclear. It is generally believed that calcinosis represents a scarring process in damaged muscle during the acute phase of the disease. Calcinosis usually occurs after the acute phase of the disease one to three years after onset and is associated with a longer interval from onset to diagnosis[23,24].

Treatment of calcinosis cutis is unsatisfactory and only anecdotal reports of treatment with diltiazem, probenecid, warfarin, aluminum hydroxide and colchicine have been published[2,24-26]. The mechanism of action of aluminum hydroxide is to form insoluble aluminum phosphate and decrease the intestinal absorption of phosphate which results in a reduction of calcium phosphate in plasma. Surgical debridement of calcification can offer relief in patients who have a decreased range of motion.

In our study, calcification occurred in 4 cases (57%) and all cases were associated with increased time of diagnosis and delayed therapy. We treated two cases of calcinosis cutis with aluminum hydroxide but the response was unsatisfactory. Early diagnosis and treatment of juvenile dermatomyositis with high dose corticosteroids can prevent calcinosis cutis[23].

In conclusion, the present of juvenile dermatomyositis in Thai children revealed data similar to previous studies in Caucasians.

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จูวิไนล์ เดอร์มาโตมัยโอสิสในเด็กไทย

ศรีศุภลักษณ์ สิงคาลวณิช, พ.บ., วนิดา ลิ้มพงศานุรักษ์, พ.บ.*,*
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ผู้รายงานได้ทำการศึกษาผู้ป่วยเด็กที่ได้รับการวินิจฉัยว่าเป็น Juvenile dermatomyositis ที่มารับการรักษา ณ สถาบันสุขภาพเด็กแห่งชาติมหาราชินี ระหว่างปี พ.ศ. 2531–2541 ผลการศึกษา มีผู้ป่วยทั้งหมด 7 ราย เพศหญิง 6 ราย เพศชาย 1 ราย อายุที่เริ่มมีอาการตั้งแต่ 2.5 ปี ถึง 11 ปี (อายุเฉลี่ย 7 $\pm$ 3.6 ปี) อาการสำคัญที่ผู้ป่วยมาโรงพยาบาลได้แก่ กล้ามเนื้ออ่อนแรง 6 ราย (86%) ปวดกล้ามเนื้อ 2 ราย (28%) ผื่นผิวหนัง 4 ราย (57%) ผลการตรวจร่างกายพบกล้ามเนื้ออ่อนแรงตั้งแต่เกรด 3 ถึงเกรด 4 ทุกราย ความผิดปกติทางผิวหนังที่ตรวจพบได้แก่ ผื่น heliotrope 6 ราย (86%), gottron's papules 2 ราย (28%) ผื่นแพ้แสง 2 ราย (28%) และหินปูนเกาะที่ผิวหนัง (calcinosis cutis) 4 ราย (57%) ผลการตรวจทางห้องปฏิบัติการ พบการเพิ่มของเอ็นซัยม์กล้ามเนื้อทุกราย การตรวจคลื่นไฟฟ้ากล้ามเนื้อและผลทางจุลวิทยากล้ามเนื้อเข้าได้กับกล้ามเนื้ออักเสบ 6 ราย ผู้ป่วย 7 รายได้รับการรักษาด้วยยาเพรดนิโซโลนขนาด 1–2 มิลลิกรัม/กิโลกรัม/วัน ในระยะแรกและลดขนาดยา โดยได้รับยาอย่างน้อย 1 ปี ผลการรักษากล้ามเนื้ออ่อนแรงหายเป็นปกติ 2 ราย ดีขึ้นแต่มีอาการเป็นซ้ำ 4 ราย ขาดการติดตาม 1 ราย การศึกษานี้ไม่พบอัตราตาย โรคแทรกซ้อนที่พบคือ หินปูนเกาะที่ผิวหนัง 4 ราย ซึ่งเกิดตามหลังกล้ามเนื้ออ่อนแรงเป็นระยะเวลา 1–3 ปี ซึ่งไม่ตอบสนองต่อการรักษาด้วยยาหรือผ่าตัด สรุป Juvenile dermatomyositis เป็นโรคเรื้อรังและพบไม่บ่อยในเด็ก การรักษาโดยให้ยาเพรดนิโซโลนตั้งแต่แรก จะป้องกันความพิการของแขนขา โรคแทรกซ้อนที่พบบ่อยคือหินปูนเกาะที่ผิวหนัง

คำสำคัญ : เดอร์มาโตมัยโอสิส, เด็กไทย

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