
Original Articles

JUVENILE DERMATOMYOSITIS IN NORTH INDIA**Surjit Singh, Lata Kumar and K. Ravi Shankar**

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Objective: To determine the clinical profile and therapeutic response of patients with juvenile dermatomyositis (JDM). **Design:** Hospital based descriptive follow-up study. **Sample:** 12 patients attending the Pediatric Rheumatology and Immunology Clinic over last five years.

Results: The patients were aged between 3Vi years to 22 years with a male to female ratio of 2:1. All patients had proximal muscle weakness at presentation. Distal muscle weakness and masseter atrophy was seen in 2 patients and neck flexor weakness and pharyngeal weakness was seen in 1 case. Muscle pain, tenderness or swallowing difficulties were not observed. Classical skin manifestations of JDM were present in all except one patient. Vasculitic lesions were not noted. One patient had diffuse lipoatrophy. Two patients developed calcinosis cutis while on treatment. All patients were put on oral corticosteroids (prednisolone 2mg/kg/day) initially which were gradually tapered while monitoring clinical response. Early initiation of steroid therapy resulted in an excellent response. Two patients who did not show significant improvement even with prolonged steroid therapy were given oral weekly methotrexate (10 mg/m²/week).

Conclusions: Most of the children with JDM showed good response to steroid therapy which needs to be continued for a prolonged period. Children who do not respond to this therapy may be given oral weekly methotrexate.

JUVENILE dermatomyositis (JDM) is a multisystem disease characterized by non-suppurative inflammation of striated muscle and skin. Though well recognized, it has been uncommonly reported from India(1). We present our experience of this condition over the last 5 years.

Clinical Profile

Twelve children diagnosed, treated and followed at the Pediatric Rheumatology and Immunology Clinic constitute the subjects of this study. The patients were aged between 3V£ years to 12 years with mean

age of 8.6 years. The male to female ratio was 2:1 (Table I).

These children had been symptomatic for a variable period ranging from 1 month to 3 years. All had proximal muscle weakness at the initial examination. Two of these, in addition had significant distal muscle weakness also. Neck flexor weakness along with pharyngeal muscle weakness was seen in one patient while masseter atrophy was seen in two patients. Skin manifestations at presentation were seen in all except one patient. These dermatological manifestations were variable and

TABLE I—Initial Clinical Profile and Laboratory Investigations

Case No.	Sex	Age at onset (years)	Skin changes	Muscle weakness	CPK (U/L)	EMG	Muscle biopsy
1	F	8	Malar rash, edema-periorbital, sacral and pedal	Proximal and distal	1347	Myopathic	Inflammatory myositis
2	M	9 ½	Heliotrope rash, Gottron's papules	Proximal and distal, neck flexor and pharyngeal weakness	2235	Myopathic	Inflammatory myositis
3	F	8	Malar rash, Gottron's papules, nail bed telangiectasia	Proximal	NA	Myopathic	Inflammatory myositis
4	M	6 ½	Malar rash, heliotrope rash	Proximal	NA	NA	Inflammatory myositis
5	M	1 ½	Nil	Proximal	2962	Myopathic	Inflammatory myositis
6	F	6	Malar rash, Gottron's papules, bluish discoloration of hands and feet	Proximal	1937	Myopathic	Inflammatory myositis
7	M	9	Malar rash, heliotrope rash, photosensitive rash over neck	Proximal	258	Myopathic	Inflammatory myositis
8	M	3 ½	Heliotrope rash, malar rash, nail bed telangiectasia	Proximal	299	NA	Not contributory
9	M	5	Heliotrope rash, Gottron's papules	Proximal	292	Myopathic	Not done
10	M	4	Heliotrope rash	Proximal	1319	Myopathic	Contributory by histochemistry
11	F	8	Heliotrope rash, erythematous rash over pinna and retroauricular area	Proximal	1094	Myopathic	Inflammatory myositis
12	M	12	Heliotrope rash, lipoatrophy, telangiectasia, Gottron's papules	Proximal	68	Myopathic	Inflammatory myositis (predominantly perifascicular atrophy)

consisted of a classical heliotrope rash in 8 cases, malar erythematous rash in 6 cases, Gottron's papules in 6, nail bed telangiectasia in 3, bluish discoloration of hands and feet not amounting to Raynaud's phenomenon in 1, and photosensitive rash over the neck in 1 case. One child had periorbital, sacral and non pitting pedal edema. One patient (Case no. 12) who presented after 2 years of onset of symptoms had diffuse subcutaneous atrophy (lipoatrophy). None of our patients had arthritis, skin ulceration, esophageal or pulmonary symptoms.

The creatine phosphokinase (CPK) levels were elevated in 9 patients; in 2 patients the reports were not available and one patient who presented late had normal CPK levels (Case no. 12). Rheumatoid factor was negative in all patients while one girl had transient anti-nuclear factor (1+ speckled) positivity. Electromyography showed a myopathic pattern in all 10 patients where this investigation was possible. The EMG picture ranged from increased insertional activity, fibrillations and polyphasic motor units to bizarre high frequency discharges. Muscle biopsy was consistent with inflammatory myopathy in 10 of the 11 patients biopsied; in one, it was not contributory. Muscle biopsy findings included myonecrosis, myophagocytosis, perifascicular atrophy and extensive inflammatory infiltrates.

Treatment and Follow-up

The response to treatment and follow-up are summarized in *Table II*. All patients were admitted at the time of first presentation to us and were treated with oral corticosteroids (prednisolone 2 mg/kg/day). Regular clinical assessment of muscle power and serial serum CPK levels whenever possible was done during follow-up. Three patients had excellent response (almost normal muscle power and normal CPK val-

ues within 2 months), 4 had good response (minimal or no muscle weakness and normal CPK values in 6 months), 2 had fair response (normalization of CPK values after 6 months, no relapses on therapy, with or without muscle weakness) and 2 had poor response (relapses while on therapy and/or significant muscle weakness). Case no. 12 presenting after two years of onset of symptoms had minimal muscle weakness with a normal CPK value and probably represented a burnt out disease.

Full dose of corticosteroids was given in all patients except one (case no. 12 who did not receive any treatment as there was only minimal muscle weakness at the time of presentation). Steroids were continued till the CPK values normalized or muscle weakness significantly improved and subsequently the dosage was gradually tapered. Most of the children are still under treatment, save one child with excellent response who had completed two years of steroid therapy and is completely well now. Two children with poor response have had relapses. First, Case no. 3, had increasing weakness two years after prednisolone therapy. At this time the child was also noted to have significant diffuse subcutaneous calcification of the neck and upper limbs. After ruling out steroid myopathy (by looking for any improvement in muscle weakness with decrease in steroid dosage), the child was started on oral weekly methotrexate. Presently, the child has completed one year of therapy and has shown considerable improvement. The second child, Case no. 5, relapsed after 2 years of prednisolone therapy. This child was also started on oral methotrexate after ruling out steroid myopathy. However, he has not returned for follow-up since that time. Another child with fair response, Case no. 9, developed subcutaneous calcifications around pelvic girdle during the course of therapy. At least 4 children, who

TABLE II—Course and Follow-up of Patients

Case No.	Duration (mo) of symptoms prior to presentation	Response* to steroid therapy	Duration of therapy	Relapses	Remarks	Grade of ⁺ disability at present
1.	1	Excellent	2nd yr	—	—	I
2.	8	Fair	2nd yr	—	—	III
3.	12	Poor	4th yr	After 24 mo	On methotrexate, diffuse calcinosis	I
4.	18	Excellent	14 mo	—	Bilateral masseter atrophy	I
5.	24	Poor	3rd yr	After 24 mo	On methotrexate	III
6.	5	Good	8th mo	—	Cushingoid habitus	II
7.	12	Good	12th mo	—	—	I
8.	12	Good	6th mo	—	—	II
9.	12	Fair	4th yr	—	Discrete calcinosis	I
10.	1	Good	24th mo	—	Bilateral masseter atrophy	I
11.	0.50	Excellent	4th mo	—	—	I
12.	24	Not given	NA	—	—	II

* For details please refer to text.

⁺Grade I: Full functional recovery and no abnormality on examination.

Grade II: No abnormality on examination, but easy fatigability.

Grade III: Some atrophy and weakness without serious functional impairment.

Grade IV: Moderately serious functional impairment in walking, climbing stairs and lifting heavy objects.

are presently under treatment have variable degree of residual muscle weakness.

Discussion

JDM is an uncommon disease of childhood⁽¹⁾. It constitutes about 3.3% of pediatric rheumatologic disorders seen in our clinic over the last 5 years. The median age of our patients was 8 years. At all ages the disease is slightly more frequent in females than males with a sex ratio of approximately 1.6:1⁽³⁾. However, most of the children in our study were males (M:F-2:1). We don't have an adequate explanation for this but it may be a reflection of gender bias

due to which more boys are brought to the hospital for treatment as compared to girls. In most of the western countries children have reported to hospital within a few weeks of onset of symptoms⁽⁴⁾. In our series this period has been variable and has ranged from 1 month to 36 months.

The rash of JDM which is nearly pathognomonic of the disease is basically due to cutaneous angitis⁽³⁾. The rash was seen in all except one case (No. 5) in whom it was not clearly noticeable at the time of presentation to us. The rash was mostly classical (malar rash, heliotrope violaceous

discoloration and Gottron's papules). None of our patients had any significant vasculitic lesions.

Proximal muscle weakness was seen in all our children. Cassidy and Petty have reported distal muscle weakness in about 30% cases(5). We noticed significant distal muscle weakness in 2/12 patients only. Masseter atrophy was observed in 2 patients causing difficulty in chewing food. Esophageal involvement has been reported in upto 1/3rd of patients with JDM, but this was not present in any of our patients. Only 1 of our patients had weakness of neck flexors.

Other manifestations not seen in our patients but reported in JDM include arthritis (30%), cutaneous ulceration (25%), Raynaud's phenomenon (15%) and gastrointestinal hemorrhage (5%)(3). One of our patients (Case no. 12) who presented very late in the course of illness had diffuse subcutaneous atrophy (lipoatrophy), a finding which is reported in less than 5% of patients with JDM(5).

Corticosteroids are the drugs of first choice in the management of JDM. Poor response to corticosteroids may be seen under the following circumstances: (i) When there is associated overlap syndrome—none of our patients had a clearly discernable overlap syndrome; (ii) When there is a delay in initiation of therapy—children with fair and poor response in our series have had long duration of symptoms (8 mo-36 mo) prior to initiation of therapy; and (iii) When the starting dose of prednisolone has been less than 1.5 mg/kg/day. We have used a starting dose of 2 mg/kg/day in all our patients.

A 'response' is defined as clinical improvement in a patient's muscular strength and/or normalization of serum enzymes. It is at this point that the corticosteroid dosage is reduced and gradually tapered over

next 2 years or so. Sudden cessation of prednisolone therapy results in relapse. Some authors believe that muscle strength should be the defining criterion because muscle enzymes have been known to reach normal values despite continued clinical weakness(6). In patients with normalized serum enzyme levels, but ongoing muscle weakness, possibility of steroid myopathy should be considered. It is characterized by insidious onset, hip flexor weakness, minimal myopathic changes on EMG and type II fiber atrophy on muscle biopsy. Decreasing the steroid dosage will result in paradoxical increase in strength. In two of our patients we differentiated relapse from steroid myopathy on the basis of above mentioned criteria.

About 1/5th of all patients with dermatomyositis either fail treatment with steroids or are unable to tolerate these(6). In our series, 2 patients (16%) did not show an adequate response to steroid therapy. Immunosuppressants should be used early both for control of disease and for their steroid sparing effect in such patients. Methotrexate is the first line immunosuppressant for both adults and children with either steroid resistance or for fulminant life threatening disease(5). We have used oral methotrexate in 2 patients showing poor response. In one child, the response has been excellent with complete recovery of muscle power. The other child is still to be assessed. The other drugs which have been used with variable results are pulse methylprednisolone, cyclophosphamide, azathioprine, chlorambucil, mercaptopurine, intravenous immunoglobulin, cyclosporine and chloroquine(7). Plasmapheresis has also been tried in the acute stage of illness(7). Needless to say, physiotherapy and other supportive measures must be provided to all patients with JDM.

Calcinosis cutis is one of the specific

complications of JDM which usually develops late in the course of treatment indicating burnt out myositis, but may also occur early during active disease. Two of our patients developed this complication while on therapy. The pathogenesis is unclear but is likely to be dystrophic calcification. It is more likely to occur in patients on low dose prednisolone (<1.5 mg/kg/day) than in patients given a dose above 1.5 mg/kg/day (8). Various drugs (*e.g.*, warfarin, penicillamine) have been tried to treat this condition but none has been shown to be uniformly successful. We did not give any specific therapy for this complication and both of our cases have shown considerable resolution of calcification.

The prognosis of JDM depends on severity of initial involvement (pharyngeal muscle involvement has poor initial response(9) as in Case no. 2), time lapsed after onset of symptoms to initiation of therapy (4 months and beyond has poor prognosis(10) as in Case nos. 2,3,5 and 9) and presence of nail fold capillary loops(11). Taieb *et al.* observed that 30% of the children did well on steroids and were weaned off, while 62% followed a more chronic course that was continuous or interspersed with remissions and relapses(9). In our series, 80% have shown a satisfactory response. The 2 children with poor prognosis have entered 4th year of follow-up and are on methotrexate therapy while another child with fair response is in the 4th year of steroid treatment.

In this series of 12 patients with JDM, the response to prolonged treatment with corticosteroids was quite gratifying, with 10 patients showing almost complete recovery. Weekly oral methotrexate remains a useful second line agent which can be used in children who fail initial therapy with corticosteroids. No significant disability was documented in any of our patients.

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