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Juvenile dermatomyositis: Clinical profile and disease course in 25 patients

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ABSTRACT

A retrospective analysis of 25 Arab patients with juvenile dermatomyositis (JDMS) was conducted between 1988 and 1996. The mean age at disease onset was 8.25 years (range 1.5 - 15 yrs) with a male: female ratio of 1.5: 1. The disease duration before diagnosis was 1 - 108 months. Two patients had a family history of JDMS. The clinical features included fever in 14 patients (56%), weight loss in 20 (80%), muscle weakness in all 25 (100%), and muscle pain in 14 (56%). Skin lesions included Gottron's papules in 15 patients (60%), heliotrope in 13 (52%), erythematous malar rash in 8 (32%), and pigmentary changes in 12 (48%). Seventeen of the 25 patients had arthralgia (68%) and 16 patients had arthritis (64%). Gastrointestinal symptoms were noted in 19 patients (76 %). Myocarditis with cardiac failure was the initial presentation of 1 patient, while 2 had conduction defect. Twelve patients (48%) had respiratory symptoms. The course of the disease was complicated by calcinosis in 10 patients (40%). All of the patients were treated with prednisone; 15 were also treated with methotrexate. The duration of follow up ranged from 6 - 108 months (mean 54.5 months). Twenty-three patients improved, including those who had calcinosis at the time of presentation, with a current muscle power of 4/5 in 10 patients (40%) and 5/5 in 13 patients (52%). No deaths were reported in our series and no patients are currently bedridden.

Introduction

Juvenile dermatomyositis (JDMS) is a multi-system disease characterized by vasculopathy affecting primarily the skin and muscles, and causing symmetrical proximal weakness and typical skin rashes (1). JDMS represents the type IV of idiopathic myositis (2). It seems to differ from the adult form of the disease by the presence of vasculitis, which affects the small blood vessels causing ulceration of the skin and subcutaneous tissues and involving the gastrointestinal tract and myocardium, and the late development of calcinosis (1).

The objective of this study was to review Arab patients with JDMS whose disease onset was before 16 years of age, and to correlate the duration of the disease before diagnosis and treatment with the possible complications of this clinical entity.

Materials and methods

We reviewed the records of 25 patients diagnosed as having JDMS in the Rheumatology Clinic, King Faisal Specialist Hospital and Research Centre and King Khalid University Hospital (Riyadh, Saudi Arabia) between 1988 and 1996. The criteria for the diagnosis of JDMS were applied using the confidence limits defined by Bohan and Peter (3).

A retrospective analysis of the following variables were conducted: the age at onset of the disease, the duration of the disease before diagnosis, the duration of follow up, a family history of dermatomyositis and polymyositis, clinical presentation, laboratory findings including muscle enzymes, electromyographic study (EMG) and muscle biopsy, drug therapy and clinical course. Complications such as calcinosis, and the final outcome were also tabulated. The clinical course of the disease was assessed based on an assessment of muscle strength, skin rash, muscle enzymes and functional status before and after starting the recommended treatment.

Results

All 25 patients had muscle weakness and the typical skin rash. Fifteen of the 25 patients had definite JDMS, while 10 had probable JDMS. Males were more predominant than females in a ratio of 1.5: 1. The mean age at onset was 8.25 years (range 1.5 - 15 years). The duration of the disease before diagnosis ranged from 1 - 108 months. The duration of the follow-up ranged from 6 to 108 months (mean 54.5 months). Two patients had a positive family history of JDMS. All patients had muscle weakness. Weight loss was noted in 76% of the patients, while arthralgia and arthritis were present in 64% and 60% of patients respectively (Table I).

The most common skin rash encountered was Gottron's papules in 15 patients, followed by heliotrope rash in 13 patients; 4 patients had severe cutaneous

Table I. Clinical features at presentation of 25 patients with juvenile dermatomyositis.

Clinical features	n	(%)
Muscle weakness	25	(100)
Skin rash	25	(100)
Weight loss	19	(76)
Arthralgia	16	(64)
Arthritis	15	(60)
Fever	13	(52)
Muscle pain	13	(52)
Respiratory symptoms	11	(44)
Abdominal pain	10	(40)
Calcinosis	10	(40)
Dysphagia	8	(32)
Conduction defect	2	(8)
Myocarditis	1	(4)

vasculitis. Myocarditis was the initial presentation of one patient who also had cardiac failure followed by muscle weakness, elevated muscle enzymes and the characteristic rash of JDMS. Two patients had asymptomatic conduction abnormalities manifested by an abnormal electrocardiogram. Respiratory manifestations in the form of cough, shortness of breath, aspiration pneumonia and restrictive lung disease were observed in 11 out of the 25 patients (44%) and one patient needed ventilator support when he had respiratory failure secondary to aspiration pneumonia. Soft tissue calcification mainly at the elbows, knees and buttocks causing further joint contractures and aggravating the lack of mobility, was seen in 10 of the 25 patients (40%); in the majority of these calcification occurred more than one year after the disease onset.

Muscle biopsies and an EMG examination were performed in 23 and 22 patients, respectively. The most characteristic findings in the muscle biopsies were chronic inflammatory infiltrates composed of interstitial or perivascular mononuclear cells associated with degeneration, necrosis and regeneration of myocytes, which were found in 16/23 patients (69.5%). A polyphasic pattern with wide base high frequency discharges were found on EMG in 11/22 patients. The variations in muscle enzyme levels, as well as laboratory findings including antinuclear antibodies (ANA), are summarized in Table II.

All 25 patients were started on oral prednisone at 1 - 2 mg/kg/d divided into 3 daily doses. Pulse intravenous methylprednisolone (IVMP) at 30 mg/kg/dose daily for 3 consecutive days was given to those patients with severe muscle weakness and to those with long-standing active disease. Oral methotrexate at a maximum dose of 15 mg/m^2 weekly was added if the patient had severe muscle weakness, skin or gastrointestinal vasculitis, respiratory muscle involvement, severe edema secondary to massive inflammation, or lack of response to steroid therapy. All patients tolerated methotrexate well, although one patient stopped after 1 year because of significant nausea and vomiting. Hydroxychloroquine at a dose of 7 mg/kg/day was also added if the patient presented with extensive skin rash and needed high dose steroids.

The treatment protocols were continued for an average of 20 months. Once the muscle power grade and skin rash as well as the muscle enzymes levels had improved, prednisone was tapered to one daily dose in a gradual, step-wise fashion over 2 years. The muscle power grade in the patients improved dramatically after appropriate therapy was begun. Recent follow-up examinations showed a muscle power grade of IV in 10 patients (40%) and V in 13 patients (52%). No patient is currently bedridden, even those who presented with calcinosis universalis (Table III). No patients died in this series and no patients were reported to have developed any malignancy.

Table II. Laboratory data analysis in 25 patients with JDMS at the time of presentation.

Lab test	High (n)	(%)	Normal (n)	(%)	Low	Not done
Creatine kinase	12	(48)	12	(48)	1 (3.8)	—
Aldolase	6	(24)	4	(16)	—	15
Aspartate aminotransferase	10	(40)	13	(52)	—	2
Lactate dehydrogenase	17	(68)	6	(24)	—	2
ESR	22	(88)	3	(12)	—	—
WBC	4	(16)	21	(84)	—	—
Hb	—		18	(72)	7 (28)	—
Urea, Cr	—		25	(100)	—	
Antinuclear antibody titre	15	(60)	7	(28)	—	3
Anti DNA (ds)	2	(8)	20	(80)	—	3
Extractable nuclear antigens (JO)	3	(12)	16	(64)	—	6
C3	1	(4)	13	(52)	—	11
C4	1	(4)	13	(52)	—	11

Discussion

Juvenile dermatomyositis is a multi-system diseases characterized by vasculopathy of the skin and/or muscles causing symmetrical proximal weakness and typical skin rashes (4). In our series 15 patients had definite JDMS while 10 had probable JDMS. It has been reported that juvenile dermatomyositis is at least 10 to 20 times more common than polymyositis in children and tends to have a more acute and severe onset (4). Although females seem to predominate over males in many reports (3, 5, 7), our series revealed male predominance (M:F 1.5:1); there was no obvious reason for this observation. There seemed to be a bimodal age distribution of onset in the individuals with inflammatory myositis, with peaks occurring at 5 to 14 years of age, and at 45 to 64 years (1, 4). In our report the age at onset of the disease was similar to other series.

Two of our patients had a family history of juvenile dermatomyositis, none involving first degree relatives, and there was no consanguinity. Muscle weakness, skin rash and arthritis are the most common clinical features of the disease. The clinical presentations of the patients were similar to those in previous reports (4, 5) (Table IV).

The frequency of calcinosis in our patients was comparable to that in other reports (5, 9). This complication was found in patients who had diffuse vas-

Table III. Muscle power assessment in JDMS patients at presentation and post-treatment.

Muscle power	At presentation n	(%)	Post-treatment n	(%)
Grade I	2	(8)	0	
Grade II	9	(36)	0	
Grade III	8	(32)	2	(8)
Grade IV	6	(24)	10	(40)
Grade V	0		13	(52)

culitis and more than 6 months of active disease before the initiation of appropriate therapy. The delay in initiation of proper treatment in our report was estimated to range from 6 months to 6 years, with an average of 39 months. The reason for this delay was improper diagnosis and delayed referral.

The pathogenesis of the cutaneous involvement and calcium deposition seen in JDMS is thought to usually be due to an insult, possibly viral or bacterial, that initiates an inflammatory process in the muscles and small blood vessels, causing swelling of the endothelial cells lining the dermal vessels which may be severe enough to cause vascular occlusion (10). The papillary basement membrane thickening - which may eventually lead to decreased tissue perfusion and acute ischemia followed by collagen deposition - is proportional to the severity and extent of the initial disease process. This scarring process includes dystrophic calcification and is independent of the serum concentration of calcium and phosphorus.

ANA in low titres was positive in 15 patients (60%), comparable to other reports (11). Three patients (12%) had positive extractable nuclear antigen (ENA) antibodies in the form of JO-1. Two of these patients had significant restrictive lung disease. This finding is not supported by the report of Nishikai *et al.* (17), who described the presence of JO-1 antibodies in adults with dermatomyositis. Two patients had positive anti-ds-DNA. None of those patients had any features of mixed connective tissue disease.

Twenty-three and 22 patients respectively had muscle biopsy and EMG examinations performed. Muscle biopsy in 16 of 23 patients (64%) showed changes typical of JDMS, while 11 of the 22 patients (50%) showed diagnostic features of JDMS on EMG. In the remaining patients the biopsy or EMG results were either suggestive, non-specific, or normal. In one report (12), 10% of children with definite JDMS had normal results on EMG and 5 - 10% of patients with active disease had normal muscle biopsies. These variations may be due to technical differences and sampling problems as the myopathic process is focal, even in a severely affected child. Magnetic resonance imaging (MRI) may be more useful in diagnosing focal inflammatory myopathy (13).

All of our patients were started on corticosteroids and hydroxychloroquine concomitantly. There are few studies reporting the efficacy of IVMP (14). Methotrexate was added to the treatment protocol in severe cases with gastrointestinal vasculopathy, respiratory muscle involvement or severe cutaneous vasculitis. Previous reports have shown the beneficial effects of various immunosuppressive agents including methotrexate in childhood dermatomyositis (15). Data in the literature suggest a 3 - 7.5% mortality rate in JDMS (16). In our series the survival rate was 100% and no patients developed malignancy during the long term follow up.

At the latest follow-up our patients showed a muscle power grade of IV (38.4% of the patients) or V (50%) and no patient is bedridden, including those who had extensive calcification (calcinosis universalis).

In conclusion, calcinosis is a not uncommon complication of JDMS if the patients has suffered extensive vascular damage and a long duration of active disease before initiating proper therapy.

Table IV. Frequency of clinical features of JDMS in our series, and in the two series of Pachman *et al.* and Miller *et al.*, respectively (expressed as percentages).

Clinical features	Our series	Pachman *et al.*	Miller *et al.*
Muscle weakness	100	100	100
Skin rash	100	100	100
Arthritis	60	36	10.3
Fever	52	65	30.8
Muscle pain	52	72	48.7
Abdominal pain	40	37	7.7
Calcinosis	40	22	30.8
Dysphagia	32	45	23.1

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