

A Retrospective Study of 13 Oriental Children with Juvenile Dermatomyositis

Y See,**MBBS, M Med (Paeds), MRCP (Paeds)*, Y C Giam,**MBBS, M Med (Paeds)*, H H Chng,****MBBS, M Med (Int Med)*

Abstract

This is a retrospective study of 13 Oriental children with juvenile dermatomyositis (JDM). We reviewed data found in the hospital records of children diagnosed to have definite (n = 4), probable (n = 7) and possible (n = 2) JDM who presented over a 10-year period at 4 centres in Singapore and compared our results with the experience of others. We found an overall female preponderance (female to male ratio of 3.3:1) but an equal sex ratio in children below 5 years of age. The majority (92%) had insidious onset and good outcome. Diagnosis was often delayed because of the insidious onset, and because weakness occurred late, was mild or absent. Only one child had an acute presentation and refractory course. She died despite aggressive therapy. Clinical features, complications and mainstay medication used were similar to Western studies.

Ann Acad Med Singapore 1997; 26:210-4

Key words: Antinuclear antibodies, Inflammatory myopathy, Muscle enzymes, Photosensitive facial rash

Introduction

Juvenile dermatomyositis (JDM) is an uncommon childhood disease with inflammation of the skin and muscle resulting in characteristic skin lesions and symmetric proximal muscle weakness.¹ It is part of a spectrum of diseases ranging from mainly muscle involvement (polymyositis) to mainly skin involvement (dermatomyositis sine myositis, or amyopathic dermatomyositis).²⁻⁸ Respiratory and palatal muscles may be involved. Calcinosis may occur late in the disease as active inflammation subsides.⁹ Gut and central nervous system involvement are rare.

Most of the data on JDM are based on studies carried out in the West. The little data on Oriental children include two Taiwanese studies.^{10,11} The aims of this study were to describe the demographic, clinical and laboratory features, treatment response and outcome in children in Singapore and to compare these with published data.

Materials and Methods

In November 1994, we carried out a retrospective study of children diagnosed to have JDM in Singapore. Case records with ICD codes 710.3 (dermatomyositis) were traced from 4 centres in Singapore: 3 hospitals with large paediatric units (Tan Tock Seng Hospital, Singapore General Hospital and National University

Hospital) and the National Skin Centre. Paediatricians and rheumatologists in these centres were also contacted for cases they were managing.

We found 14 cases who satisfied the criteria of Bohan and Peter,¹² but excluded one child with connective tissue disease overlap, leaving 13 children. They had first presented during a 10-year period between 1984 and 1993. Using a standardised data entry form, we collected information on race, sex, age at diagnosis, duration of symptoms before diagnosis, presenting features, diagnostic criteria fulfilled, complications, outcomes, laboratory results and treatment given. We then compared the results with published data.

Results

A summary of the 13 patients' key features is shown in Table I. There were 11 Chinese and 2 Malays. Of the 13 patients, 10 were girls and 3 were boys (female to male ratio of 3.3:1). However, there were 2 boys and 2 girls (sex ratio 1:1) below the age of 5 years. With increasing age, there was an increasing predominance of girls: 6 girls and only 1 boy in the 5-10 years age group, and 2 girls and no boys in the group above 10 years old. The age at diagnosis ranged from 2 years 10 months to 16 years 9 months. For girls, the mean age at diagnosis was 8½ years, while for boys it was 5½ years. The overall

* Registrar
Department of Paediatrics
Alexandra Hospital

** Senior Consultant
National Skin Centre

*** Consultant
Department of Rheumatology and Immunology
Tan Tock Seng Hospital

Address for Reprints: Dr Chng Hiok Hee, Department of Rheumatology and Immunology, Tan Tock Seng Hospital, Moulmein Road, Singapore 308433.

mean age at diagnosis was 7 years 9 months.

Four patients fulfilled the criteria for definite JDM, 7 for probable JDM and 2 for possible JDM (Table I). All the patients had rash but only 9 had weakness at presentation. Two other patients developed weakness later. Two children (15%) did not have any subjective nor objective weakness at all, although their muscle enzymes were elevated and one had positive muscle biopsy. Half were initially misdiagnosed as atopic or photosensitive dermatitis or other rash by their family physicians before JDM was recognised. The correct diagnosis was usually made when the child later developed weakness or clumsiness, or when it was realised that the rash was characteristic of JDM and muscle enzyme levels were checked.

The presenting features are shown in Table II. Only child number 3 presented acutely with fever, prostration, weakness and rash. The rest (92%) had insidious onset with symptoms over a period of one month to 1½ years before diagnosis. The 3 most common complaints were rash, weakness and fever. The duration when these were noticed, before diagnosis of JDM, was in the region of months for rash, weeks for weakness and days for fever. Two patients already had calcinosis at diagnosis. Palatal involvement was found in 3 children (23%). They had nasal speech, difficulty in swallowing and regurgitation (which might have contributed to pneumonia suffered by two of them).

The results of laboratory investigations are shown in Table III. Of the muscle enzymes, the majority had at

TABLE I: SUMMARY OF KEY FEATURES OF PATIENTS

No.	Sex/Race	Age/Year of diagnosis	Diagnostic criteria	Other features	Outcome
1**	F/Ch	2 y 10mo (Jul 1991)	rash, weakness, raised enzymes	Gradual onset over 4 months Weakness developed 2 years later	Relapse in 7/91, 1/94
2***	F/Ch	3 y 8 mo (Feb 1993)	rash, weakness	Gradual onset with rash for 1 year Calcinosis at diagnosis	Persistent calcinosis
3**	F/Ch	6 y 2 mo (Oct 1991)	rash, weakness, raised enzymes	Acute onset dysphagia, nasal speech, bed sores, pneumonia	Died of respiratory failure
4*	F/Ch	7 y 8 mo (Aug 1992)	rash, weakness, raised enzymes, positive EMG	Gradual onset over 8 months	Relapse 6/94
5**	F/Ch	8 y 4 mo (Jan 1993)	rash, weakness, raised enzymes	Gradual onset with rash for 1 year Calcinosis at diagnosis	No relapse Mild residual rash
6**	F/Ch	8 y 9 mo (Jun 1993)	rash, weakness, raised enzymes	Gradual onset over 3 months Weakness 1½ years later Palatal involvement	No relapse Mild residual rash
7**	F/Ch	9 y 2 mo (Jul 1987)	rash, weakness, raised enzymes	Gradual onset over 4 months	Mild residual rash Relapse
8*	F/Ch	9 y 11 mo (Feb 1993)	rash, weakness, raised enzymes, positive EMG, positive biopsy	Gradual onset over 4 months	No relapse
9*	F/Ch	11 y 8 mo (Nov 1991)	rash, weakness, raised enzymes, positive EMG	Gradual onset over 2 months Septic arthritis in 1993	Relapse in 3/92, 11/92, 3/93
10"	F/Mal	16 y 9 mo (Apr 1993)	rash, weakness, raised enzymes	Gradual onset over 1 year	Defaulted follow-up
11*	M/Ch	4 y 2 mo (Oct 1984)	rash, weakness, raised enzymes, positive EMG, positive biopsy	Gradual onset over 1 month Palatal weakness Pneumonia	Relapse in 11/84, 2/92
12"	M/Ch	4 y 6 mo (Apr 1990)	rash, raised enzymes, positive muscle biopsy	Gradual onset over 2 months No weakness	Recurrent rashes
13***	M/Mal	7 y 10 mo (Sep 1986)	rash, raised enzymes	Gradual onset over 1½ years No weakness Calcinosis 5 years later	Recurrent rashes

F: female; M: male; Ch: Chinese; Mal: Malay; EMG: electromyography

+: Definite JDM; **: Probable JDM; ***: Possible JDM

TABLE II: PRESENTING FEATURES

Patient No.	Rash	Calcinosis	Weakness	Fever	Others
1	+				
2	+	+			
3	+		+	+	acutely ill, prostration
4	+				
5	+	+	+		
6	+			+	arthritis
7	+		+		
8	+		+	+	
9	+		+	+	arthritis
10	+		+	+	arthralgia
11	+		+		
12	+				
13	+				
Total	13	2	7	5	

TABLE III: LABORATORY TESTS

	Number done	Number abnormal (%)
Creatine kinase	12	10 (83.3)
Aldolase	10	10 (100)
Aspartate aminotransferase	6	5 (83.3)
Lactic dehydrogenase	4	4 (100)
Biopsy	4	3 (75)
Electromyogram	5	5 (80)
Antinuclear antibody	11	3 (23)
Anti-dsDNA antibody	8	0 (0)

least creatine phosphokinase or aldolase done. Of the 4 patients who underwent muscle biopsy, only 3 were positive for myositis. Four of the 5 electromyographic studies ordered were positive. Full blood count was essentially normal in all children. Two girls had transient thrombocytopenia which was probably related to intercurrent infection. Repeat values were normal. Their lupus serology was negative while bone marrow test was not done. Antinuclear antibody was positive in only 3 of the 11 tested (27%) and dsDNA negative in all 8 tested.

All the patients received systemic steroids orally in the form of prednisolone, between 1 and 2 mg/kg/day, with improvement except in case number 3. In addition, topical steroids or hydroxychloroquine were used for rashes. Only one patient (no. 3) with refractory JDM was also given parenteral steroid and cytotoxics (methotrexate, azathioprine). One child with progressive calcinosis was treated with colchicine with transient response. Supportive treatment included sunscreen and antibiotics. At least 3 of the patients also sought traditional Chinese medication. An analysis of one of the samples of Chinese medication received revealed the presence of salicylate and prednisolone. The child

with refractory disease associated with severe muscle weakness died of respiratory failure in another country after defaulting follow-up.

Discussion

We reviewed the demographic and clinical data found in hospital records of 13 Oriental children diagnosed with JDM using the criteria of Bohan and Peter. The chief limitation of this study is the small number of cases. However, as there is currently relatively little information available on Oriental children, it is hoped that this study will contribute to the body of information on this uncommon disease. The small differences between this and other series (for example sex ratio) could be due to small number variations. Another limitation is the retrospective nature of the study. Clinical information sought may not have been recorded and laboratory tests done were not exactly the same for all patients. Also, patients who had died, recovered and discharged or otherwise lost to follow up for more than 5 years preceding this study, would have had their case-sheets culled and not be available for study. Thus, the majority of the children in the study were diagnosed within the later 5 years. Another reason for the greater number of cases in recent years could be a greater awareness of this condition by doctors and dermatologists and thus earlier diagnosis.

We found an equal female to male sex ratio in children below the age of 5 years (2:2) and an increasing proportion of girls in older children especially above the age of 10 years. This pattern of equal sex ratio in very young children but female preponderance in older children was also noted by Hanissian and others² in 1982. Our overall female to male ratio of 3.3:1 appears to be higher than Western studies of 2:1. However, this needs to be confirmed by a larger study. The two Taiwanese studies found a ratio of 1.7:1 and 2.7:1, respectively.^{10,11}

The average age of 7 years at diagnosis is similar to that in the West.¹³ The age range is also similar to reported studies.¹⁴⁻¹⁶ The youngest patient we had was 2 years 10 months at diagnosis, but rash had been present for the preceding 4 months (2 years 6 months at onset). This falls within the range noted by Western authors in which there have been no reports of children below the age of 2 years with JDM.

As observed, onset can be acute or insidious. We had only one child with acute onset. The majority (92%) were of insidious onset, more than those reported in Western literature (75% by Jacobs,⁷ 67% by Cassidy and Petty¹⁷ and 50% by Ansell¹⁸). There appears to be quite a wide variation in the different studies. The range of presenting features in our patients is similar to that reported from elsewhere.

Late diagnosis was often due to misdiagnosis or because parents sought treatment from traditional Chi-

nese healers first. Earlier diagnosis might be possible by stressing to medical undergraduates and general practitioners the characteristic features of the rash of JDM (malar and periorbital nature, photosensitivity and Gottron's papules), and other supportive features to check for, such as weakness and raised muscle enzymes.

Four patients fulfilled the criteria for definite JDM, 7 patients for probable JDM, and 2 patients for possible JDM. When diagnosis was based on rash, weakness and muscle enzymes; muscle biopsy and electromyographic (EMG) studies were not done as these two procedures are invasive. It may explain why only 4 patients had the diagnosis of definite JDM. The authors suggest that EMG and/or muscle biopsy should always be done in patients with uncertain diagnosis.

We note that two children never developed any objective or subjective weakness at all, even after 4 years and 9 years follow-up, respectively. They were both boys (nos. 12 and 13). The former had 3 exacerbations of rash over 3 years while the latter had multiple exacerbations over 5 years until the onset of calcinosis, after which inflammation became quiescent. However, they did not quite fit the category of amyopathic JDM as they had raised muscle enzymes. Amyopathic dermatomyositis has been described in children with the classic skin signs of JDM but no muscle involvement (in terms of weakness, raised enzymes, changes on EMG and muscle biopsy).

The range of complications seen in our patients is similar to that reported in the literature.^{1,17} Variations in frequencies could be due to the small number in this study as well as to the relatively short duration of follow up. Although the longest follow-up was 10 years, the majority of cases were diagnosed within the last 5 years with many diagnosed within the last one year, so the full course of illness may not yet be borne out. We had one death (8%). Recent reports quoted an average death rate of 10%.¹ Most of our other children have done well. None have obvious physical handicap. Again this could be related to the small number and as yet incomplete course of disease.

One child had rash, weakness and calcinosis but normal enzyme level (no. 2). It is probably because she presented towards the end of active inflammation when muscle enzymes can be normal. It is reported that muscle enzymes may be normal early or late in the disease, during remissions and in the amyopathic form. As enzyme levels may be raised to a different extent for different enzymes, it is advisable to check all 4 enzymes viz creatine kinase, aldolase, aspartate aminotransferase and lactic dehydrogenase.

Only 75% of muscle biopsies done were positive while only 80% of EMG done were positive. This could be

because the pathology may be patchy, muscle biopsy and EMG may miss involved areas and be reported as negative. Findings were also non-specific, showing inflammation or degeneration. Thus negative biopsy or EMG does not exclude JDM, nor does a positive result necessarily confirm the diagnosis. However, they add supportive evidence especially when diagnosis is not obvious based on other features.

Oral prednisolone, topical steroid and sunscreen were the main medications used. It is interesting that among the Chinese, traditional medicine were still popular and the analysis of one of these drugs revealed prednisolone and salicylates.

Conclusion

This study of 13 Oriental children with JDM although small and retrospective gave the following information. Overall there was a greater proportion of girls but an equal female to male ratio in younger children. Only one child (8%) presented acutely, did not respond to steroids and cytotoxics and eventually died within 7 months from chest infection and respiratory failure. The children who were initially misdiagnosed with eczema or some other skin condition had an insidious onset of rash preceding weakness for months to years. Clinical features and complications are similar to published data. The overall percentages of children with muscle biopsy and EMG done were relatively small as diagnosis was obviously based on classical clinical features and raised muscle enzymes.

REFERENCES

1. Jacobs J C. *Paediatric Rheumatology for the Practitioner*. New York: Springer-Verlag, 1993:527-55.
2. Hanissian A S, Masi A T, Pitner S E, Cape C C, Medsger T A Jr. Polymyositis and dermatomyositis in children: An epidemiologic and clinical comparative analysis. *J Rheumatol* 1982; 9:390-4.
3. Euwer R L, Sontheimer R D. Amyopathic dermatomyositis: A review. *J Invest Dermatol* 1993; 100(1 suppl):124S-127S.
4. Euwer R L, Sontheimer R D. Amyopathic dermatomyositis (dermatomyositis sine myositis): Presentation of six new cases and review of the literature. *J Am Acad Dermatol* 1991; 24:959-66.
5. Krain L S. Dermatomyositis in six patients without initial muscle involvement. *Arch Dermatol* 1975; 3:241-5.
6. Rockerbie N R, Woo T Y, Callen J P, Giustina T. Cutaneous changes of dermatomyositis precede muscle weakness. *J Am Acad Dermatol* 1989; 20:629-32.
7. Taieb A, Guichd C, Salamon R, Maleville J. Prognosis in juvenile dermatopolymyositis: A co-operative retrospective study of 70 cases. *Pediatr Dermatol* 1985; 2:275-81.
8. Caro I. A dermatologist's view of dermatomyositis/polymyositis. *Clin Dermatol* 1988; 6:9-14.
9. Bowyer S L, Blane C E, Sullivan D B, Cassidy J T. Childhood dermatomyositis: Factors predicting functional outcome and development of dystrophic calcification. *J Pediatr* 1983; 103:882-8.

10. Wang Y J, Lii Y P, Lan J L, Chi C S, Mak S C, Shian W J. Juvenile and adult dermatomyositis among the Chinese: a comparative study. *Chin Med J (Taipei)* 1993; 52:285-92.
 11. Ting C K, Hsieh K H, Lin Y Z, Chou C C, Wang J Y. Clinical observation of juvenile dermatomyositis and polymyositis. *J Rheumatol ROC* 1988; 5:1-12.
 12. Bohan A, Peter J B. Polymyositis and dermatomyositis. *N Engl J Med* 1975; 292:344-7.
 13. Singsen B H. Rheumatic diseases of childhood. *Rheum Dis Clin North Am* 1990; 16:593-9.
 14. Medsger T A Jr, Dawson W N Jr, Masi A T. The epidemiology of polymyositis. *Am J Med* 1970; 48:715-23.
 15. Winkelman R K. Dermatomyositis in childhood. *Clin Rheum Dis* 1982; 8:353-68.
 16. Sullivan D B, Cassidy J T, Petty R E. Dermatomyositis in the pediatric patient. *Arthritis Rheum* 1977; 20(Suppl):327-31.
 17. Cassidy J T, Petty R E, editors. *Textbook of Paediatric Rheumatology*. 3rd ed. WB Saunders Co, 1995:323-64.
 18. Ansell B. Juvenile dermatomyositis. *Rheum Dis Clin North Am* 1991; 17:931-42.
-