Juvenile dermatomyositis

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Summary: Thirteen cases of dermatomyositis seen in children are described. Records of hospitals serving 250,000 children were reviewed for the 10-year period during which these children were seen and no other cases were found. Eleven of the 13 are living, including seven believed to be in complete remission; the remainder have ongoing disease of varying severity. Literature concerning this unusual childhood disease is reviewed and there is general agreement as to the characteristic nature of the clinical picture. Special features, including rash, muscle findings, deformity, calcinosis and gastrointestinal bleeding, are discussed. Emphasis is placed on the value of steroid therapy, though some serious complications are acknowledged. Methotrexate, found promising by others, was without benefit in two cases.

Dermatomyositis is a chronic inflammatory disease of skin and muscle with characteristic signs and symptoms but with a highly variable course. This clinical study is confined to the childhood form, and we present a long-term follow-up of 13 cases, 12 of them seen personally by the authors. Several recent publications[1-5] have summarized the literature, clinical experience and pathology of dermatomyositis. We compare our findings with those of others.

Patient care

Most of these children were followed up in the Children's Arthritis Program of the Canadian Arthritis and Rheumatism Association in Vancouver. A multi-discipline team included a dermatologist, an internist, a pediatrician, a physiotherapist, an occupational therapist and a social worker. Usually the referring physician managed the patient, and the clinic served in a supportive, consultative role.

Case distribution

The 13 children seen over a 10-year period were from the Greater Vancouver hospital district. Nearly 900,000 people live in this district and approximately 250,000 are under 15 years of age.[6] This is certainly not a common childhood disease. Records of all hospitals serving children were reviewed and we could find no other cases of juvenile dermatomyositis seen during this 10-year period.

Clinical features

The main clinical features are listed in Table I. The average follow-up period was 7½ years. Skin changes were prominent. A frequent finding was symmetrical, dull-erythematous to deep-crimson scaling dermatitis of the upper eyelids, often with periorbital edema. Similar dermatitis was common over the malar eminences, elbows, knees, knuckles and interphalangeal joints. Telangiectasia over these joints was usual and scaling was especially marked over the elbows and knees. In fact, the dermatitis over these joints was pathognomonic and of more diagnostic aid than the better known eyelid changes.

In more severe cases and in acute exacerbations the dermatitis was more extensive. Resolution was generally accompanied by mottled hyper- and hypopigmentation. Mucous membrane changes were common in the more acute phases, with moist erosive patches on the vermilion surfaces of the lips and on the labial mucosa. The tongue, buccal mucosa and palate were not affected. Muscle weakness ranged from mild to profound, and muscle mass also varied according to the severity of disease. Proximal muscle groups were always affected, and a "woody" indurated feeling was a valuable diagnostic aid. In more severe cases there was widespread muscle involvement, including pharyngeal and esophageal groups. Calcinosis developed in five children, all of whom had been ill

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for several years. Gastrointestinal bleeding occurred in four children, one of whom had a proved duodenal ulcer. Three of these patients (including the one with the ulcer) were on prolonged adrenocortical steroid therapy.

General management and physical therapy

All children received physiotherapy to maintain as good a range of movement as possible in the face of acute inflammation. At this stage, joints were moved passively and plaster resting-splints minimized contractures. As muscle function improved, active exercises and hydrotherapy were added. Despite daily therapy, contractures could not be prevented in the more severely affected children. Manipulation under anesthesia was carried out on one patient (Case 10), and although an improved position was achieved the casts had to be removed postoperatively because of excessive pain; the contractures returned.

The occupational therapist advised severely handicapped children on the activities of daily living, and also provided diversional therapy. The social worker helped to resolve problems arising from the presence in the family of such a child.

Drug therapy

Prednisone was the most important available drug, and was used in 11 patients. The duration of therapy in each case is indicated in Table I. No specific dosage schedule was followed, but the usual initial dose was 40-50 mg. daily, with a maximum of 60 mg. In most cases significant improvement in muscle power and general well-being were obvious after a period of up to six weeks, and dosage could then be reduced to 10-15 mg. daily without occurrence of exacerbation. Subsequent flare-ups were not infrequent and nearly always subsided with temporary increase of prednisone dosage. We were able to discontinue prednisone in the five steroid-treated cases now in remission. Of the four children with continuing disease, one is improving on a small dose of prednisone, whereas the other three had complications due to steroids requiring withdrawal of this drug.

ACTH was given briefly in two cases and anabolic steroid in three cases without obvious benefit. Methotrexate was used in two cases.

Treatment results

A. Patients recovered

Seven patients are now in complete remission. Two of these received no steroids. One (Case 6) recovered after only four months of mild disease, while the other (Case 7), with marked muscle weakness in the early stages, recovered after three years of more severe disease. Four patients were on prednisone therapy for eight months to two years (Cases 1 to 4). One (Case 3) was severely affected and his course was complicated by intestinal hemorrhage requiring ileal resection. Recovery is complete, and he is now working as a truck driver. One patient (Case 5) was on prednisone therapy for eight years. Recovery was incomplete, in that she was left with residual muscular weakness and severe calcinosis. However, there has been no active disease of skin or muscle for six years. Calcium deposition in her case is illustrated in Fig. 1. Surgical removal of this mass relieved pain and increased range of movement.

B. Patients deceased

Two children died. The first child (Case 8) had severe acute derma-

TABLE I
Clinical features in 13 cases of juvenile dermatomyositis

	Recovered							*Died*		*Continuing disease*			
	Steriod treated					*No drugs*				*Little activity*		*Severe*	
Case no.	1	2	3	4	5	6	7	8	9	10	11	12	13
Sex	*M*	*M*	*M*	*F*	*F*	*F*	*M*	*F*	*F*	*M*	*F*	*F*	*F*
Year of birth	1954	1960	1945	1950	1947	1952	1956	1946	1940	1953	1952	1954	1960
Age of onset	5	2	15	11	6	10	4	11	9	7	12	10	6
Duration of disease (years)	8/12	1	2	2½	10	4/12	3	6/12	16	8	5	5	3
Follow-up years from onset	6	8	8	8	16	7	8	6/12	16	8	5	5	3
Rash	+	+	+	+	+	+	+	+	+	+	+	+	+
Peri-orbital edema	+	+	?	+	+	0	0	+	+	+	0	+	+
Muscle induration	+	+	+	+	+	+	+	+	+	+	+	+	+
Muscle wasting	?	0	+	+	+	0	+	+	+	+	+	+	+
Weakness	+	+	+	+	+	+	+	+	+	+	+	+	+
Deformity	0	0	+	+	+	0	0	0	+	+	0	+	+
G.I. bleeding (gross)	0	0	+	0	0	0	0	0	+	+	0	0	+
Calcinosis	0	0	0	0	+	0	0	0	+	+	0	+	+
Steriod therapy (years)	8/12	9/12	2	1	8	0	0	2/12	1	6	5	4½	3

tomyositis of six months' duration. There was no response to prednisone in high dosage. Death was from respiratory infection. The other patient (Case 9) has been reported previously.[7] She was the first of our patients treated with steroids. First cortisone and then ACTH produced an initial response, but fluid retention and hyperlipemia occurred and these drugs were discontinued. She became progressively crippled with calcinosis. Gastrointestinal bleeding, hyperlipemia and renal disease were additional problems. The dermatomyositis had been considered in remission for a number of years before her death, 16 years after onset. Her inclusion in the deaths from dermatomyositis is questionable, as the exact cause of death was not established.

C. Patients with continuing disease

Four children have continuing disease and their cases are briefly reviewed.

Case 10—A 15-year-old boy had profound muscular weakness and an associated skin eruption at the age of 7. There was initial improvement with prednisone 25 mg. daily, but he then became worse, with increased muscular weakness and muscle contractures. Multiple vertebral compression fractures occurred after two years of treatment. Efforts to reduce prednisone dosage resulted in exacerbation of the dermatomyositis. An anabolic steroid (Danabol) produced precocious masculinization. Calcinosis became prominent. Over the next four years there were multiple respiratory infections along with continuing muscle and skin disease. Low steroid dosage was continued, and two years ago gradually increasing muscular strength was noted; prednisone was discontinued. In the past two years he has shown continued improvement in muscle power and general well-being, with persistent but diminishing rash. Severe muscle contractures remain. Diffuse calcinosis also persists (Fig. 2).

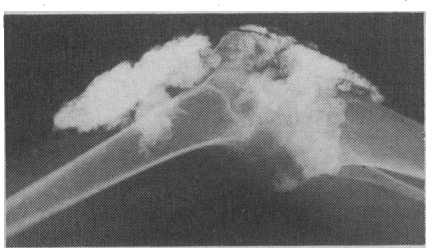

FIG. 1—Localized masses of calcium in Case 5.

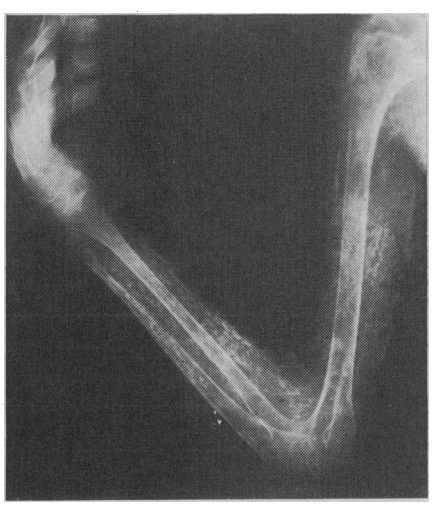

FIG. 2—Diffuse calcinosis and osteoporosis in Case 10, showing maximal extension possible in upper limb.

Case 11—A 17-year-old girl noted a mild skin eruption at the age of 12. This was soon followed by severe muscle induration and weakness. Prednisone therapy was started and there was prompt improvement. Initial dosage was 30 mg. daily and the present dosage is 5 mg. daily. Muscle strength is normal and there is only minimal rash. Efforts to reduce further the prednisone dosage have so far resulted in symptoms rather than signs of muscular weakness.

Case 12—A 13-year-old girl had the characteristic skin eruption and moderate muscle induration and weakness beginning at age 10. For the first two years good function was maintained, but continuing high steroid dosage was necessary. Severe back pain led to the discovery of multiple vertebral compression fractures. Dosage of prednisone was gradually reduced and finally discontinued in September 1968.

Muscular weakness became profound, and because of severe palatal and respiratory weakness, a course of methotrexate 1.5 mg. per kg. intravenously every two weeks was started in November 1968. Dosage was increased to 3 mg. per kg. in February 1969 but was discontinued in June because no improvement had occurred.

Case 13—A 9-year-old girl had the onset of characteristic skin rash, muscle induration and quite marked weakness at the age of 6. She was treated with prednisone and remained fairly well for two years, attending school despite continuing rash and muscular involvement. Over the subsequent nine months, however, her course was complicated by ischiorectal abscess, intestinal bleeding and vertebral compression fractures. Prednisone was gradually reduced to 2.5 mg. daily with concurrent increase in muscular weakness and contractures. Methotrexate 1.5 mg. per kg. intravenously every two weeks was started in February 1969. Dosage was increased to 3 mg. per kg. in April, but the agent was discontinued in June 1969 because there was still no improvement.

Review of literature

Bitnum *et al.*[1] quoted over 200 references in 1964 and culled about 1000 cases of dermatomyositis from the world literature. There were 240 below the age of 16. A classification of polymyositis indicated that the childhood type constituted 10% of the overall incidence.[2] Cook, Rosen and Banker[3] in 1963 gave an excellent summary of clinical findings based on their own series of 50 cases seen in Boston since 1916. They estimated that two new cases of juvenile dermatomyositis might be seen each year in a large referral centre. Banker and Victor[4] made authoritative observations on pathological findings based on autopsy and biopsy material. A further summary by Hanson and Kornreich[5] included details of 17 more cases. The stereotyped nature of clinical findings and the individual character of the pathology in children, fundamentally an angiopathy, are emphasized. Onset of this disease has been described as early as 4 months[8] and most series report slightly more boys than girls. One pair of identical twins[3] and another set of siblings[9] have been reported. In the latter, the onset was in adult life.

"Violaceous" or "heliotrope" eyelid discolouration with periorbital edema has been noted frequently, as has similar discolouration of the malar eminences.[5, 10] Erythematous scaling dermatitis over the extensor surfaces of joints (particularly the knuckles) is considered pathognomonic,[3] although rarely a similar rash has been described in lupus erythematosus.[11] As the disease progresses, thickened skin and mottled hyper- and hypopigmentation may replace the original erythema,[4] and the skin overlying proximal and distal interphalangeal joints may become white and atrophic with prominent superimposed telangiectases.[5]

In addition to muscular weakness and pain, induration of the affected musculature provides evidence of myositis in most cases.[3] Contractures are common[3] and associated tiptoe gait has been described.[4] Muscle atrophy accompanying severe disease may be partly due to disuse.[4] In the most advanced cases muscle atrophy may be extreme and associated with pressure sores.[3] Weakness of palatal, pharyngeal and esophageal muscles may also occur and is of serious prognostic significance.[8]

Respiratory failure may be complicated by dysphagia, and aspiration of foreign material may be a prelude to death from pneumonia.[3]

Calcinosis has been found in from 29%[9] to 44%[3] of all cases, and in as many as 60% of those survivors followed up for more than two years.[3] Deposition of calcium may be diffuse or circumscribed and extrusion through the skin may be seen.[5] Calcinosis has been considered a favourable prognostic sign, possibly because its appearance usually indicates long-standing disease and thus survival beyond the critical acute phase.[3, 5, 12]

Abdominal pain[4] or melena[3] may signify gastrointestinal ulceration, which may occur independently of steroid therapy[3] and which is due to the same vascular lesion found in other organs,[4] namely, arterial and venous intimal hyperplasia with subsequent formation of fibrin thrombi and vascular occlusion. Such lesions constitute a second important cause of death.[4, 5]

In addition to malaise and anorexia, low-grade fever has occurred in nearly half of the cases.[3] Generalized pitting edema is occasionally found.[13] Splenomegaly has been reported in 15% of cases.[3] Carcinoma is associated with dermatomyositis in a significant percentage of adults,[14, 15] but reports of malignancy in children are limited to one case of leukemia and one case of lymphosarcoma. The clinical features of these two cases are described as atypical.[3, 5]

Within the range of disease described, any degree of involvement may be seen. The overall outcome in the series of 50 children of Cook and his associates[3] was 23 well, 15 dead and 12 either alive and crippled or not covered by adequate follow-up. From a review of 168 documented pediatric patients Bitnum *et al.*[1] found 33% recovered, 34% dead and 33% alive and crippled. Hanson and Kornreich[5] reported 13 of 17 cases doing well, two with severe disability and two dead. It would appear from several reports that the majority of fatalities occur within two years of onset.[3, 5, 13] Although fluctuations in the intensity of disease may be observed, reactivation once the process has become inactive is unlikely.[3]

Laboratory investigations rarely assist in diagnosis.[3] Although the value of biopsy has been stressed by some,[3, 4] others have reported non-specific findings.[16] Serum muscle enzyme levels,[5] particularly SGOT levels, have been found useful indications of activity of the myositis. Of the many drugs employed at one time or another for juvenile dermatomyositis, adrenocortical steroids have received the most attention. Some believe they offer no more than symptomatic relief,[3, 4] while others emphasize their value and claim a favourable influence on outcome of the disease.[5, 17]

The higher incidence of intestinal ulcers in children treated with steroids has been noted repeatedly.[3, 13] Other complications, including osteoporosis,[1] CNS changes[3] and aggravation of muscle weakness,[9] have also been stressed. Testosterone[13, 18] and other androgens[19] have been used in an attempt to stimulate muscle regeneration and also to delay osteoporosis.

Methotrexate has recently been found beneficial in one child and three adults.[20] Chelating agents may remove some calcium[21] from connective tissue. Surgical removal of calcium has helped in some instances.[3] Physiotherapy has been valuable.

The etiology of the condition is unknown and has been the subject of much speculation. The hypothesis that abnormal immune reactions may be responsible is not proved.[3] Bitnum *et al.*[1] suggest that agricultural factors may be of etiological importance; however, nine of our 13 patients are city-dwellers.

Discussion

The cases reported are similar to those described in other series. Diagnosis is based on clinical features. Enzyme changes, electromyography and skin and muscle biopsy only reinforce the characteristic clinical findings. In contrast to the multitude of laboratory abnormalities in systemic lupus erythematosus, there are but few in juvenile dermatomyositis, even in severe and acute phases. Enzyme levels can be used as an aid to clinical follow-up study, since they usually vary according to the degree of muscular damage. However, there are exceptions to this relationship. High levels of gamma globulin have been noted in acute phases.

A striking feature of this disease is the extreme variability of its natural history. This makes it difficult to draw valid conclusions from a relatively small series. Our results, namely, 11 living out of 13 patients with a minimum follow-up period of four years, appear better than those reported by Bitnum *et al.*[1] and Cook, Rosen and Banker,[3] and we would agree with Pearson[2] and Hanson and Kornreich[5] that steroids have a favourable influence on juvenile dermatomyositis.

We have repeatedly noticed prompt improvement in muscle strength and concurrent lessening of muscle pain when large doses of prednisone are given. These symptoms worsened when the dosage was reduced. In most cases a satisfactory balance between steroid dose, control of disease and side effects of therapy has been attained. Also, the self-limited nature of the disease has often allowed eventual discontinuation of the drug.

Severe problems have been encountered, however. One child died of fulminating disease while on prednisone, 60 mg. daily, and additional ACTH. Three children suffered multiple vertebral compression fractures with gross demineralization while on high steroid dosage, and three of the four children with gastrointestinal bleeding had been on steroids over a long period. It is conceded, however, that gastrointestinal bleeding may be a complication of dermatomyositis itself. We favour prednisone therapy for juvenile dermato-

myositis, but also recognize the risk of severe complications.

Methotrexate was given to two children with severe disease that had not responded well to prednisone, on the basis of a report of satisfactory improvement in four patients, one of them a juvenile.[20] No benefit was seen after seven months' and four months' treatment respectively. The use of other immunosuppressive agents will doubtless be reported, although the "auto-immune" nature of this disease appears less clear than in some other connective tissue diseases.

Résumé

La dermatomyosite juvénile

Les auteurs présentent 13 cas de dermatomyosite chez l'enfant. Une revue des dossiers hospitaliers (hôpitaux desservant 250,000 enfants) s'étendant sur la période de 10 ans au cours de laquelle ces enfants ont été vus, n'a pas permis de découvrir d'autres cas. Onze de ces 13 enfants sont vivants, dont sept sont considérés comme ayant bénéficié d'une rémission complète. Les six autres ont une maladie évolutive dont le degré de sévérité est variable. Les auteurs ont passé en revue la littérature mondiale sur cette pathologie infantile exceptionnelle et sont d'accord avec l'opinion générale que le tableau clinique de cette pathologie est très typique. Ils discutent certaines caractéristiques spéciales, dont l'exanthème, l'atteinte musculaire, les difformités, la calcinose et les hémorragies gastro-intestinales. Ils soulignent la valeur de la corticothérapie, mais reconnaissent l'existence de certaines complications sévères. Le méthotrexate, prometteur pour certains cliniciens, n'a pas eu d'effet favorable dans deux cas.

ADDENDUM

One child (Case 12), described as having severe, continuing disease, has subsequently died, with widespread skin breakdown, sepsis and respiratory failure.

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Sexual behaviour

The very language we use about sex — words like "urge" and "impulse" and "need" — seems to imply that it is a biological force which we can dam up, divert, or be carried away by — but which, whether we like it or not, is there. We must do something about it.

This conception of the nature of sexuality is not based upon research into animal and human sexual behaviour, because the great bulk of that research has in fact been done in the past decade or two. What's more, when we look at this research closely it becomes increasingly clear that the popular notion of sex as a biological force driving men and animals into sexual behaviour is seriously inadequate and even misleading, especially when it's applied to human beings. If you take the point which this research seems to be making, you've got to look at the possibility that human sexual behaviour is more in the nature of an acquired habit, taste, appetite or even addiction than it is an instinct as popularly understood.

This doesn't mean that biological factors do not influence sexual behaviour, because of course they do, and in quite a number of ways. But in accounting for the wide differences in sexual behaviour between people, biological factors are less important than learning experiences, than the personal experiences that people have had in the course of growing up.

If sexual desire is learned and cultivated, then obviously there is no biological standard of sexual behaviour against which to assess ourselves. Norms in this matter are completely social. If we want to ignore them, we can. The sexologists have sought to liberate us in the area of our sexual relationships. Now, to my way of thinking, liberation in sex means being able to take it or leave it. The biological-energy idea claims that it's impossible to leave it, but when we recognize that it is to a large extent learned, we begin to realize it is also a matter of choice whether we have it or not. — Derek Wright, Lecturer in Psychology, University of Leicester. From a BBC broadcast published in *The Listener*, 84: 99, 1970 (July 23).