C-reactive protein in childhood dermatomyositis

R. H. HAAS, R. F. DYCK,* V. DUBOWITZ, AND M. B. PEPYS*

*From the Department of Paediatrics and Neonatal Medicine (Institute of Child Health), and the *Immunological Medicine Unit, Department of Medicine, Royal Postgraduate Medical School, Hammersmith Hospital, London W12 0HS*

SUMMARY Serum levels of C-reactive protein (CRP) were determined in 9 patients with childhood dermatomyositis. Four children were seen during clinical relapse and all had serum CRP levels less than 1 mg/l. In addition direct immunofluorescent staining of muscle biopsies from 4 patients showed no evidence of CRP deposition in muscle tissue. Such patients appear to be able to produce CRP in response to acute infections, and it is suggested therefore that the pathological process in childhood dermatomyositis may not induce a significant CRP response.

C-reactive protein is the classical acute-phase reactant, and although produced nonspecifically in response to tissue injury there is evidence that the levels of CRP which are attained may vary significantly between different diseases.^{1–3} Dermatomyositis is characterised by extensive inflammation and muscle necrosis, but in the only case in which it has previously been reported serum CRP was not detected, albeit by an insensitive assay.⁴

We report here that among 9 cases of childhood dermatomyositis, 4 (cases 1, 2, 3, 4) seen during an active phase of their disease had serum CRP levels of less than 1 mg/l (Table 1). Three children (cases 6, 7, 8) had 'burnt-out' disease. One (case 7) was well apart from widespread calcinosis and no serum CRP was detected. When subsequently reassessed with healing local skin ulcers, the CRP level was 4 mg/l. The second child (also with calcinosis) (case 6) was well and had no detectable CRP, but the third patient (case 8) with a paronychia on one toe had an elevated CRP of 20 mg/l. Patient 9 had minimally elevated levels of CRP on 2 occasions, while patient 5, whose disease was well controlled by steroids, was clinically well with a CRP of 57 mg/l when seen for review as a day patient. A subclinical infection could not be ruled out, and a second CRP determination done 4 months later was 10 mg/l.

CRP was not detected by direct immunofluorescence in the muscle biopsies of the 4 children (cases 3, 4, 6, 7) tested or in normal muscle tissue. Complement profiles were within normal limits in all patients in whom they were measured.

The results demonstrate that in children active involvement of muscle tissue in the inflammation and necrosis of dermatomyositis is not associated with appreciable elevation of the serum CRP concentration. This is not apparently due to deposition of CRP in the damaged muscle, nor to inability to produce CRP, since at least one of the patients had a significant response to an intercurrent infection.

Our observations extend the range of inflammatory disorders in which even extensive tissue damage is associated with only a modest elevation of serum CRP. This is the case in both systemic lupus erythematosus^{4 8 10–13} and ulcerative colitis¹⁴ and contrasts sharply with the high CRP levels which are regularly seen in active cases of rheumatoid arthritis^{2 12 15} and Crohn's disease respectively.¹⁴ Although patients with uncomplicated SLE have been reported with high CRP values^{2 16} all the published results confirm that this is exceptional.^{13 17} Measurement of serum CRP is therefore a valuable aid in the diagnosis of intercurrent infection in febrile patients with SLE,¹³ and this may also be true in dermatomyositis.

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Correspondence to Dr M. B. Pepys, Immunological Medicine Unit, Department of Medicine, Royal Postgraduate Medical School, Du Cane Road, London W12 0HS.

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Table 1 Clinical details and C-reactive protein levels in patients with childhood dermatomyositis*

Patient	Age at study	Muscle pain†	Muscle tenderness†	Stiffness joint pain†	Rash†	Malaise†	Total score	Muscle strength‡ % normal	Prednisone mg/day	Comments	Creatine phosphokinase (normal <120 IU/l)	ESR mm in 1st h	CRP mg/l†
1 Male	10 yr 11 mo	0	1	0	1	2	4	41	5	Possible relapse	311	45	0
	10 yr 11 mo	1	1	2	1	3	8	32	15	Relapse	304	nd	0
2 Male	14 yr 10 mo	1	1	2	2	2	8	28	10	Relapse	857	45	0
	15 yr 2 mo	0	1	0	1	1	3	48	12.5	Remission, ? deep venous thrombosis	101	12	0
3 Female	5 yr	2	2	2	3	3	12	33	0	Relapse. No previous steroid therapy	990	30	0
4 Female	3 yr 8 mo	0	0	1	2	2	5	50	10	Relapse	87	30	0
5 Male	13 yr 8 mo	1	1	1	2	0	5	63	7.5	Recovering from acute relapse	56	21	57
	14 yr	0	0	1	1	0	2	88	10	Remission	61	23	10
6 Male	8 yr 11 mo	0	0	0	0	0	0	115	7.5	Inactive calcinosis, reducing steroids	83	25	0
7 Female	13 yr 2 mo	1	0	0	2	1	4	89	2.5	Burnt out disease, calcinosis, and contractures	97	32	0
	11 yr	0	0	1	2	1	4	69	0	Healing skin ulcers	91	45	4
8 Female	17 yr 11 mo	0	0	0	0	0	0	50	3.5	Burnt out disease, calcinosis, paronychia on toe	72	30	29
9 Female	15 yr 6 mo	0	0	2	1	0	3	26	0	Scleroderma features, pulmonary infiltrates	698	6	4
	15 yr 8 mo	0	0	0	0	0	0	36	0	Clinically well	562	11	8

*Diagnosis established by clinical features of muscle weakness, skin rash and malaise, together with muscle tenderness, fatigue, or calcinosis, supported by measurements of serum creatine phosphokinase, electromyography, and muscle biopsy.†

†Scores of 0–3 assigned for severity of each of the features shown.

‡Measured by electroimmunounassay. * Among normals 90% are <3 mg/l and 99% are >10 mg/l.*

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