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Commentary

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The three patients with steroid resistant dermatomyositis documented by Drs Yoshioka, Okuno, and Mikawa are all highly atypical of the usual childhood dermatomyositis, as the authors themselves point out. In two of them there was evidence of other collagen disease—systemic sclerosis (case 13) and rheumatoid arthritis (case 10). The remaining patient (case 2) had what sounds like a more classic dermatomyositis, but his response to prednisolone was odd. Usually there is evidence of clinical response in the way of improved general well being and loss of the characteristic misery, and improvement in muscle function within two to four weeks of starting treatment and the creatine phosphokinase activity, if initially raised (which only occurs in about half the cases), often lags behind, remaining raised at times, even after complete clinical resolution with tailing of steroids. For this reason the clinical course is a much more reliable guide to reducing treatment than the creatine phosphokinase activity. In their case 2, the response was completely paradoxical with a precipitous fall in the creatine phosphokinase activity without any apparent clinical improvement. Although steroid has a direct influence on creatine phosphokinase activity, it seems unlikely one could ascribe so noticeable a fall to a non-specific steroid effect. All steroids are likely to cause a steroid myopathy, but the 9- α -fluoro steroids even more so; so that their use of betamethasone would normally be contraindicated as a drug of choice for dermatomyositis.

With increasing build up of cytotoxic treatment in addition to continuing steroid this patient eventually showed some response. It is often assumed that lack of response to increasing treatment (or high dose initial treatment) indicates unresponsiveness, but the alternative possibility of iatrogenic steroid myopathy as a result of overtreatment is equally

likely. This was well illustrated by the recent case of an 11 year old girl referred to me with refractory dermatomyositis, who had initially responded well to treatment but subsequently became resistant to increasing doses of steroids with associated increasing generalised weakness together with swallowing and respiratory difficulties. She had become bed bound for 6 weeks and was considered to be terminal. She was on 70 mg prednisolone/day and had severe painful osteoporosis of her spine which made it almost impossible to move her. Over a period of five months we were able to gradually wean her off the steroids and get her ambulant again and as we reduced the steroids her strength steadily improved.

In the series of 29 of our personal cases reviewed by Miller *et al*¹ the group of patients having a low dosage of prednisolone initially had fewer relapses and less morbidity and were on steroids for a shorter period of time than those receiving higher dosage. Six of the 10 cases in the low dosage group made an uncomplicated recovery and were off treatment at least a year compared with only 1 of 18 in the second group, although the initial severity of the groups seemed comparable. Of the 12 patients in group 2 off steroids there were more severe problems with calcinosis and contractures and four were severely incapacitated.

Childhood dermatomyositis is almost uniformly responsive to steroid treatment. I think there is a good chance of remission with minimal risk of secondary complications from treatment with an initial low dosage schedule of prednisolone of 1 mg/kg daily and a reduction as soon as there is definite sign of clinical improvement (usually within two to four weeks) either in general wellbeing and loss of misery, or in increase in muscle power. This should be done very gradually and tailored to the needs of each patient, who should be closely monitored. In a 30 kg child, for example, I would start on 30 mg prednisolone/day and reduce by 5 mg/day at two weekly intervals till 20 mg/day and then by 2.5 mg/day at two weekly intervals till 10 mg/day and then by 1 mg/day at two weekly intervals. If at any point there is a suggestion of deterioration, manifesting itself either as increasing weakness or general misery, one should go back one step in the treatment and perhaps wait an extra two weeks before reducing this, or reduce at half the rate (alternate days instead of each day).

I reserve the use of additional drugs such as azathioprine, methotrexate, or cyclophosphamide for patients who are either incompletely responsive to steroid or who are difficult to wean off steroids. Alternate day steroid treatment is less effective and much slower for initial induction and should be

reserved for patients one cannot wean off steroid. If one cannot get beyond, say, 10 mg/day without apparent deterioration one could then try 20 mg on alternate days over a more prolonged period of tapering. If the child is miserable on the day without treatment one could give a small dose of, say, 2.5 mg on that day and gradually reduce the larger dose.

As in so many other medical disorders, one has to

ensure that the side effects and complications of the treatment of the dermatomyositis are not more crippling than the disease itself.

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