Pediatric Rheumatology

Poster presentation

Treatment of refractory juvenile dermatomyositis with tacrolimus

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Background

Corticosteroid and second line agents such as methotrexate have dramatically improved the outcome for patients with Juvenile Dermatomyositis (JDM). Nevertheless, some patients suffer persisting disease activity despite these treatments. Tacrolimus, an inhibitor of T-cell activation and proliferation, is one of the new therapeutic options for JDM. However, little is known about its efficacy in this patient group. We report the clinical course of three patients with refractory JDM who were treated with tacrolimus.

Patients

Three corticosteroid dependent children with extensive skin disease and severe muscle weakness were started on oral tacrolimus treatment and followed-up for 7–9 months. Patient 1: 11 year old boy, age of onset 6.1 years. Patient 2: 7 year old girl, age of onset 5.8 years. Patient 3: 9 year old girl, age of onset 4.4 years.

Results

All three patients showed impressive improvement of mainly the cutaneous lesions, and overall disease activity decreased along with the muscle enzyme levels (Figure 1), while corticosteroids could be tapered. All children became more physically active. None of the patients showed recovery of muscle strength, probably due to irreversible muscle damage related to the long-standing myositis.

Conclusion

Tacrolimus is an effective and safe second line agent in the treatment of chronic refractory JDM and improves the skin involvement substantially.

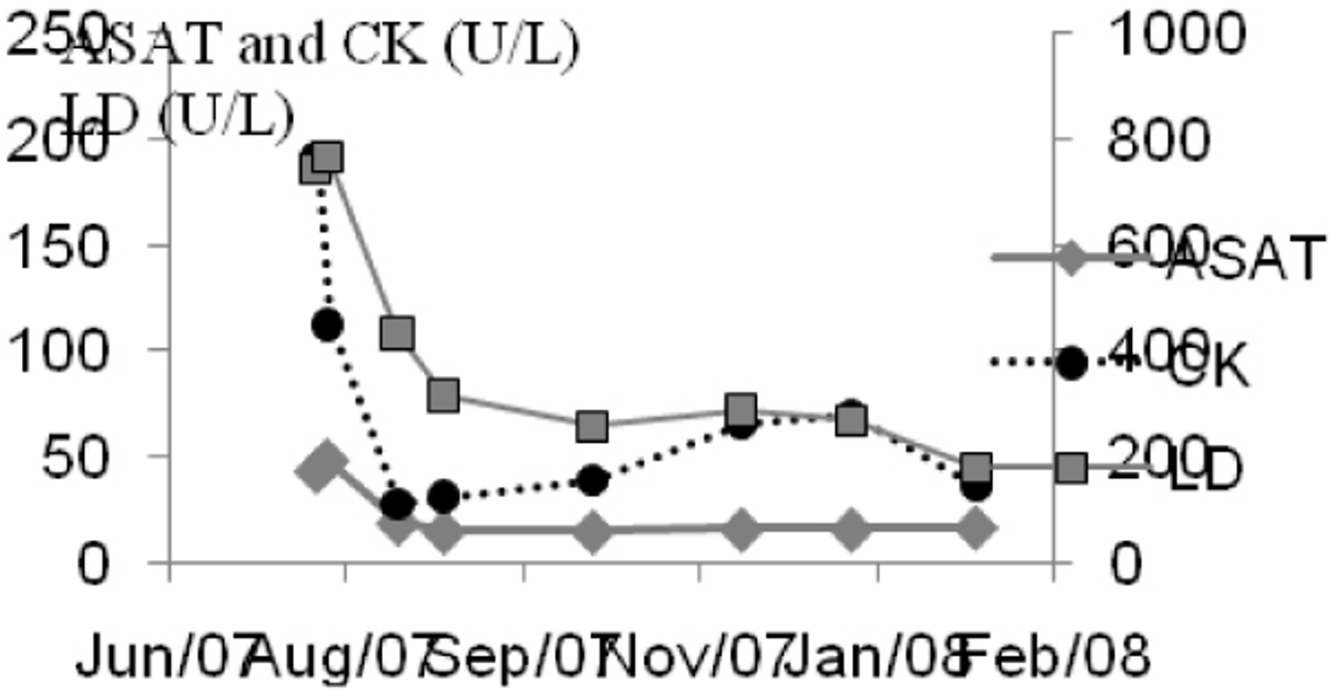

Figure 1
Course of muscle enzyme levels after the start of tacrolimus in patient 1, representative also for the other patients. Reference ranges: ASAT 0–30 U/L, LD 0–325 U/L, CK 0–145 U/L.