Juvenile dermatomyositis with tongue calcinosis and poor growth

A 9-year-old boy presented with an 18-month history of faltering growth, recurrent respiratory tract infections, persistent ‘aphthous’ ulcer on his tongue and ‘warts’ on his fingertips and soles.

He is the second offspring of a healthy unrelated Sri Lankan couple and past medical history was unremarkable. Baseline immunology and infectious disease screen were negative. On close questioning the patient had reduced energy levels, poor appetite and dysphagia of solids. His growth parameters were below the fifth centile, he had sustained central muscle weakness, a heliotrope rash on his eyelids and extruding calcinosis on his palms and soles, nail beds and tongue (figures 1 and 2).

He was clinically diagnosed with juvenile dermatomyositis (JDM) and underwent investigations to define disease severity. Muscle biopsy confirmed the diagnosis.

The patient was treated with steroids and methotrexate and surgical excision of the calcinosis on his tongue.

JDM is a rare autoimmune disease in childhood. It affects 1–4/106 children per year and females are affected twice as often as males. Median age at presentation is 7 years (range 0–16) and the disease is more frequent in Caucasians. Calcinosis is seen in 14% of children at presentation, while 30% of patients will develop it in the course of their disease. The presence of calcinosis has been linked with delayed diagnosis or inadequate treatment. It may present as a firm nodular dystrophic calcification of a previously injured area and is mainly seen in the extremities. Lesions may resolve or extrude towards the skin emptying ‘milky’ material. Treatment options include pamidronate, anti-tumour necrosis factor agents and surgical excision when pressure phenomena or disfigurement are present.

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