A Case of Severe Dermatomyositis in an African Woman

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A 39 year old black African woman woke up one day with a swollen face and neck. A few days later she developed neck and shoulder stiffness, a rash over the exposed area of her shoulders and scapulae, and swelling and weakness of the arms. Within 3 weeks the weakness had progressed and extended to her legs, which also became swollen, with tightness and sensitivity of the skin as well as muscle pain. She lost her mobility, her speech became nasal, she developed odynophagia, was soon unable to swallow and started drooling.

She was admitted to Princess Marina Hospital in Gaborone, Botswana, where a clinical diagnosis of severe dermatomyositis was made and treatment with intravenous methylprednisolone 1 mg/kg daily was begun. The patient was transferred to Bokamoso Private Hospital in Gaborone, where she was found to have non-pitting edema of her face and proximal limbs, with a slight erythema over the cheeks and peri-orbital regions, and a healed, desquamated rash on the upper chest. Proximal limb power was considerably reduced (2/5). Chest and abdominal examination were unremarkable, and chest X-ray and electrocardiogram were normal. Blood tests showed elevated aspartate aminotransferase (820 IU/L), alanine aminotransferase (294 IU/L), creatine phosphokinase (20,333 IU/L), and C-reactive protein (31 mg/L). Antinuclear antibodies were positive (1:160, speckled staining pattern) and anti-Jo antibodies negative. Platelets (95,000/µl) and albumin (26 g/L) were reduced.

A deltoid muscle biopsy was performed and histology showed predominantly endomysial and occasional perimysial chronic inflammatory cells comprising lymphocytes and focal plasma cells. There was only focal peri-fascicular atrophy. Evidence of skeletal muscle breakdown was noted. The CD4 [Figure 1] and CD8 [Figure 2] immunohistochemical stains showed a predominant CD4-positive lymphocyte population.

The patient received 1 g methylprednisolone daily for 3 days intravenously. After a percutaneous endoscopic gastrostomy tube was inserted, she was treated with prednisolone 1 mg/kg daily and started on a first dose of 7.5 mg methotrexate.

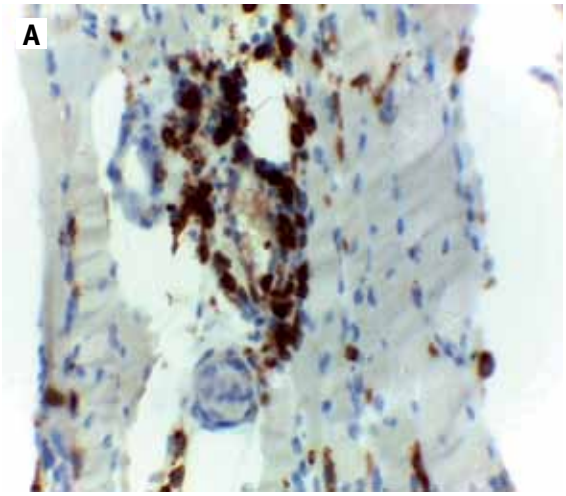

Figure 1. Perimysial, perivascular chronic inflammation (CD4 immunoperoxidase, 400x magnification)

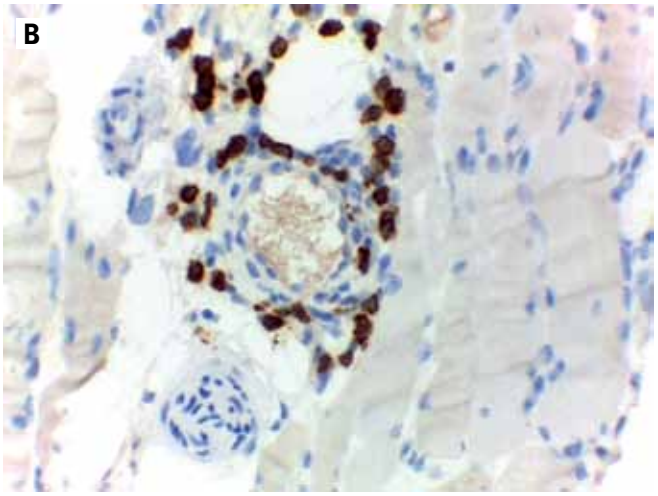

Figure 2. Perimysial, perivascular chronic inflammation (CD8 immunoperoxidase, 400x magnification)

One week later the patient had partly regained her muscle power. However, her neck muscles remained weak and there was no improvement in swallowing. She was therefore given intravenous immunoglobulins at 2 g/kg over 5 days. On the last day of the infusions she developed marked facial and peri-orbital edema and IVIG was stopped.

Blood test results (ALT 66 IU/L, AST 91 IU/L, CPK 595 IU/L, platelets 225,000/μl) and muscle power improved considerably over the following 4 weeks but swallowing and phonation did not and the patient had to continue receiving nutrition via a PEG tube. Methotrexate continued to be increased by 2.5 mg per week to a maintenance dose of 25 mg; prednisolone was continued at 80 mg daily.

At 4 months after the onset of symptoms, the patient's clinical condition has improved considerably; phonation is back to normal, she can now swallow and is regaining further power in the proximal limbs.

Dysphagia due to involvement of the pharyngeal muscles, the upper esophagus, or both, is reported in 32–84% of patients with myositis [1]. Aphagia (dysphagia to both liquids and solids) is observed in a minority of patients [2]. Our case demonstrates that aphagia can be reversible if appropriate treatment is instituted, as shown in another recently described case [3], and indicates that severe cases of dermatomyositis can occur in sub-Saharan Africa where the disease is reportedly rare [4,5].

IVIG = intravenous immunogloublin
ALT = alanine aminotransferase
AST = aspartate aminotransferase
CPK = creatine phosphokinase
PEG = percutaneous endoscopic gastrostomy

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