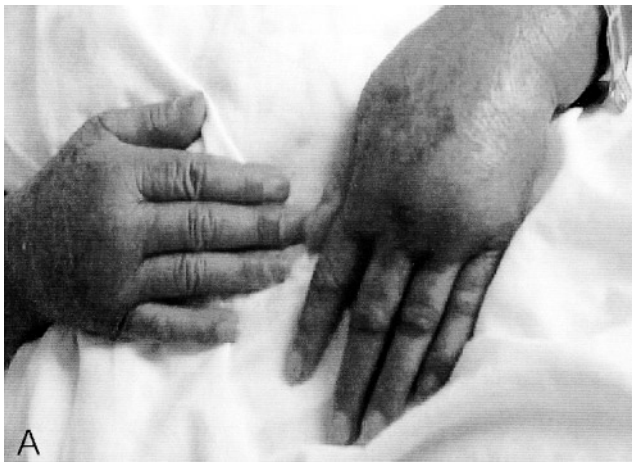
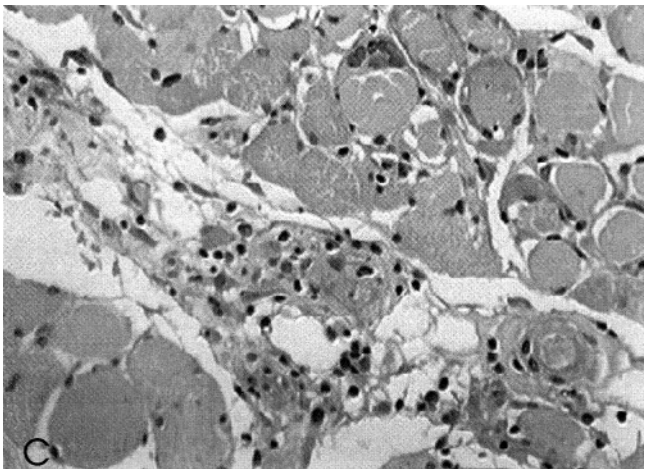
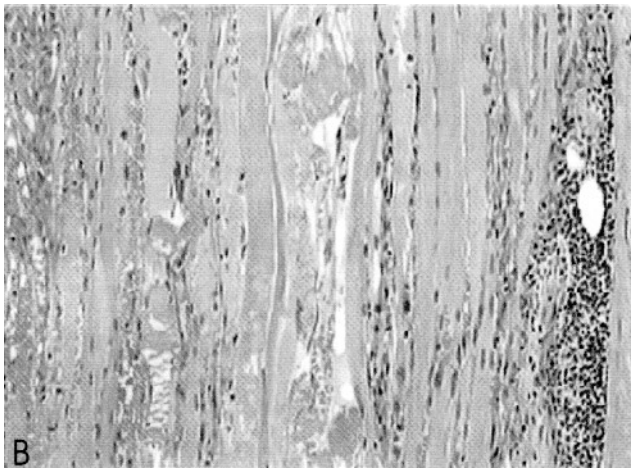
A Case of Edematous Dermatomyositis

KAMEL H. MROUE', MD, Fellow in Rheumatology, Department of Internal Medicine; NABIL H. SHARARA, MD, Assistant Professor, Department of Family Medicine; JEAN G. RBEIZ, MD, Professor, Department of Internal Medicine; THURAYYA K. ARAYSSI, MD, Assistant Professor, Department of Internal Medicine, American University of Beirut, Cairo Street, Beirut, Lebanon. Address reprint requests to Dr. T. Arayssi. E-mail: ta01@aub.edu.lb

A 78-year-old woman presented with a 10-day history of fatigue, upper extremity weakness, rash over her face and upper chest, and frank edema of both upper limbs. Physical examination showed erythematous lesions over her face and upper chest. Tenderness and weakness of the proximal muscles of both upper and lower extremities and significant non-pitting edema of both upper limbs were noted (Figure 1A). Complete blood count, erythrocyte sedimentation rate, and thyroid-stimulating hormone were within normal limits. SGOT and SGPT were elevated, and CPK was 5700 IU/l (normal range 15–195 IU/l). Antinuclear antibody was positive. Venous duplex scan of upper extremities showed no evidence of thrombosis. Electromyographic examination was compatible with an inflammatory myopathy of the proximal muscle groups. Biopsy of her left deltoid muscle showed extensive perifascicular muscle fiber necrosis with regeneration, and perivascular and interstitial lymphocytic infiltrate consistent with dermatomyositis (Figures 1B and 1C). Malignancy investigations including bilateral mammography, computed tomographic scan of chest,







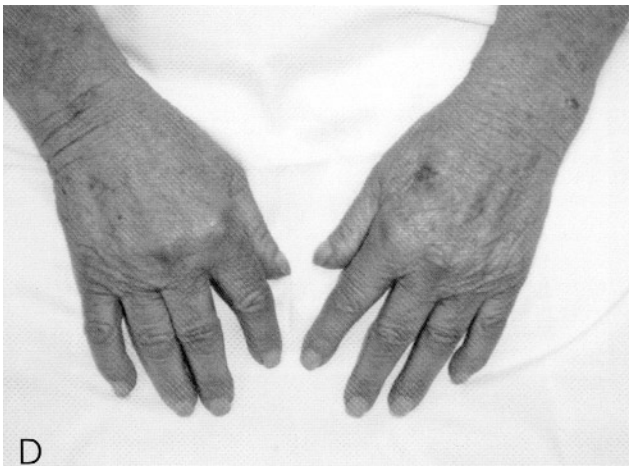


Figure 1A. Edematous dermatomyositis in a 78-year-old woman. A. Non-pitting edema seen on presentation. B and C. Biopsy of the left deltoid muscle revealing perifascicular muscle fiber necrosis, with regeneration and perivascular and interstitial lymphocytic infiltrate consistent with dermatomyositis. D. Resolution of edema 2 weeks after initiation of prednisone treatment.

abdomen and pelvis, and colonoscopy were normal. She was given prednisone 60 mg po daily and methotrexate 10 mg po weekly, with rapid improvement in her weakness, normalization of CPK, and resolution of the edema within 2 weeks (Figure 1D). One month later she developed pulmonary embolism and received anticoagulation, but died few days later.

Gross peripheral edema is a rare presentation of dermatomyositis, reported only in a few cases in the literature[1,2]. It clinically mimics deep vein thrombosis or eosinophilic fasciitis, which should be considered in the differential diagnosis[2,3]. It may be a hallmark of a distinct variant of such inflammatory myopathies. More cases are needed to establish guidelines for treatment and prognosis.

REFERENCES

1. Gorelik O, Almoznino-Sarafian D, Alon I, et al. Acute inflammatory myopathy with severe subcutaneous edema, a new variant? Report of two cases and review of the literature. Rheumatol Int 2001;201:163-6.
2. Andonopoulos AP, Gogos CA, Tzanakakis G. Subcutaneous edema: an 'unrecognized' feature of acute polymyositis. Rheumatol Int 1993;13:159-61.
3. Smyth AE, Bell AL, Crone M. Acute oedematous dermatomyositis [letter]. Ann Rheum Dis 2000;59:575.