

Connective Tissue Disease

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Toxic epidermal necrolysis-like acute cutaneous lupus erythematosus with CNS vasculitis

Premjit Vaiyavatjamai, MD, Division of Dermatology, Department of Medicine, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Ratchathewi, Bangkok, Thailand; Chanutwan Treewittayapoom, MD, Division of Dermatology, Department of Medicine, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Ratchathewi, Bangkok, Thailand; Poonkiat Suchonwanit, MD, Division of Dermatology, Department of Medicine, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Ratchathewi, Bangkok, Thailand

A 67-year-old male, a known case of discoid lupus erythematosus (DLE) and chronic obstructive pulmonary disease (COPD), presented with a gradual development of generalized, erythematous coalescent papules and plaques on trunk, extremities, palms and soles in 1 week. Some lesions were ulcerated with hemorrhagic crusts. He had fever for 1 month with progressively altered consciousness accompanying skin eruption manifested in 1 week prior to admission. Five days after hospital admission, the skin lesions developed into widespread dusky erythema with bullae and extensive erosions forming on previous lesions, involving more than 70% of body surface area. Conjunctivae, oral mucosa and genitalia were spared. Nikolsky's sign was positive.

He had DLE for 2 years, without systemic involvement. Antinuclear antibody (ANA) was negative. Hydroxychloroquine were prescribed and sun avoidance was suggested; however, he had been lost to follow-up for 18 months without any flare episode. He took oral theophylline and salmeterol-fluticasone inhalation for COPD.

Skin biopsy indicated subepidermal separation, superficial perivascular and periadnexal infiltration with lymphocytes, some melanophages and evidences of basal vacuolarization. Direct immunofluorescence test showed granular C3 deposition on dermoepidermal junction. Investigation demonstrated ANA of 1:80, nucleolar pattern; negative anti-Ro(SS-A); positive anti-La(SS-B); positive anti-dsDNA and positive lupus anticoagulant. C3, C4 and CH50 were 821 ug/ml(900-1400), 109 ug/ml(100-400) and 75%(100%), respectively. Laboratory showed leucopenia (3500 cells/ml) with normal absolute lymphocyte count, hypochromic microcytic anemia and normal platelets. Chest x-ray, urine analysis, renal and liver function tests were unremarkable. Septic workup revealed no pathogen. Lumbar puncture was not performed due to eroded skin. MRI brain showed evidences of multiple ischemic foci suspected from vasculitis. A diagnosis of toxic epidermal necrolysis (TEN)-like acute cutaneous lupus erythematosus (ACLE) was made based on the clinical, histological and immunological findings. After intravenous immunoglobulin(IVIg) infusion 2 g/kg in 5 days, skin lesions were dramatically improved in 5 days and resolution was observed in 3 weeks.

We present this case to highlight the rare presentation of vesiculobullous LE skin disorders resembling TEN. Early recognition and aggressive management are critically needed.

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Etanercept induced dermatomyositis

Michael Bernardi, MD, University of Louisville, Department of Medicine, Division of Dermatology, Louisville, Kentucky, United States; Jeffrey Callen, University of Louisville, Department of Medicine, Division of Dermatology, Louisville, Kentucky, United States; Jyoti Burruss, MD, University of Louisville, Louisville, Kentucky, United States

Introduction: Dermatomyositis (DM) is a connective tissue disease with well-described cutaneous manifestations. These cutaneous findings often precede a proximal myopathy.

To our knowledge, etanercept has never before been reported to induce DM. We report a patient who developed the features of DM soon after initiation of etanercept therapy.

Case: A 41-year-old woman with a history of psoriasis presented for evaluation of worsening disease. She was well controlled on etanercept 5 years previously but stopped this medication after 1 year for unknown reasons. Her previous medical history was significant for insulin dependent diabetes, hypertension, and steatohepatitis. Etanercept 50mg weekly and methotrexate 10mg weekly were initiated. One month later, after two doses of etanercept, she complained of a new rash on her chest as her more typical psoriatic plaques were improving.

On physical examination, well demarcated, scaly, erythematous plaques remained on the trunk with more prominent involvement of the eyelids. A violaceous eruption was noted in a photodistribution over the upper chest. Small erythematous papules were evident over the interphalangeal joints. Cuticular dystrophy was apparent. Muscle strength was noted at 4/4 proximally and distally in all extremities. Laboratory studies were significant for mildly elevated liver transaminases. Creatinine kinase of 809 (24-173 U/L) and aldolase of 17.6 (1.2-7.6 U/L) were increased. Anti-nuclear antibody was within normal limits. Biopsy was non-diagnostic. Methotrexate and etanercept were both discontinued.

Three weeks later she complained of increased pruritus and lower extremity weakness. Proximal muscle strength was noted at 2/4. Repeat biopsy showed perivascular lymphocytes and histiocytes, increased smudging of the dermal-epidermal junction, and mucin deposition in the upper dermis most consistent with collagen vascular disease. Computed tomography of the chest and abdomen was unremarkable. Systemic therapy with prednisone 60mg daily and methotrexate 10mg weekly was begun.

Conclusions: Although etanercept has been reported to exacerbate preexisting DM, to our knowledge it has never been shown to induce the disease. The induction of autoantibodies by etanercept has been previously documented, as has the onset of cutaneous lupus following initiation of therapy. We present a case of DM occurring shortly after commencing etanercept therapy and add it to the growing list of drug-induced cases.

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