

come more commonplace. All patients should be evaluated with bilateral mammography followed by a core biopsy if mammography findings are abnormal. Treatment may include modified radical mastectomy, axillary staging, radiation therapy, or systemic chemotherapies.

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Eczematous Dermatitis in the Setting of Hyper-IgE Syndrome Successfully Treated With Omalizumab

Hyper-IgE syndrome (HIES) is a rare primary immunodeficiency (<1 case per million population) characterized by elevated serum IgE levels, recurring staphylococcal skin abscesses, and pneumonias.¹ Patients with HIES may show peripheral eosinophilia, eosinophilic infiltration of the skin, and increased susceptibility to infection.² In newborns, it first manifests as an eruption³ that usually persists as a moderate to severe eczematoid dermatitis into childhood and adulthood. This multisystem disorder is associated with skeletal, facial, and dental abnormalities.¹ We report the first case to our knowledge of a patient with HIES with severe recalcitrant eczematous dermatitis successfully treated with high-dose monoclonal anti-IgE therapy.

Report of a Case. A 26-year-old woman with multiple autoimmune phenomena was referred for evaluation. She had severe unremitting eczematous dermatitis poorly controlled by tacrolimus and systemic steroids. Despite treatment, she experienced significant pain and pruritus that limited her activities. She reported dermatitis since birth and multiple cutaneous complications. Her medical history included viral meningitis at age 9 years and recurrent infectious bronchitis, pneumonias, and cystitis. She retained several of her primary teeth requiring extraction. She had controlled asthma and allergic rhinitis since childhood.

She was the only child of nonconsanguineous parents. Family history was notable for seasonal allergies in her mother and thyroid cancer and acute leukemia in 2 maternal aunts. In 1999, she had autoimmune hemo-

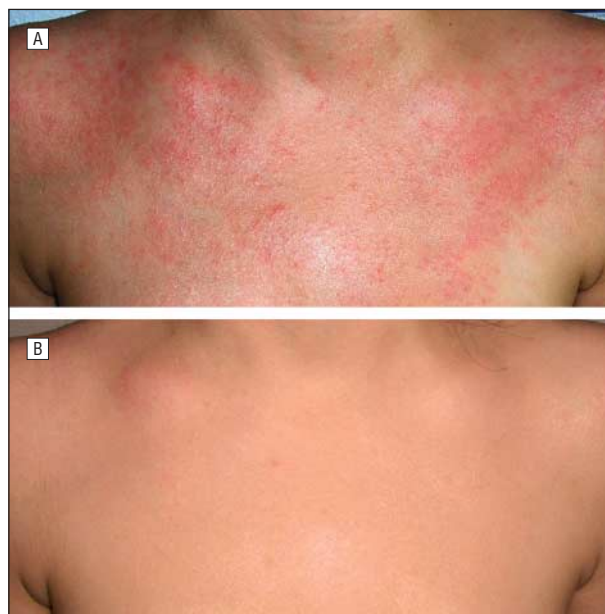


Figure. Clinical appearance of our patient before (A) and 6 months after stopping (B) monotherapy with omalizumab.

lytic anemia and was diagnosed as having hepatitis C viral infection. She then developed alopecia universalis and autoimmune hypothyroidism requiring treatment with corticosteroids and levothyroxine, respectively.

Physical examination revealed lichenified skin with extensive erythema and eczematous plaques hypersensitive to light touch, excoriations, and dry scaly plaques along the flexor surfaces of her upper and lower extremities, chest (**Figure**, A), and back. She exhibited hypertelorism, widened nasal root and alae, scoliosis, and joint hyperextensibility. Results of laboratory investigations revealed a differential eosinophil count of 9.2% (normal, 0%-5%) and IgE concentration of 1083 IU/mL.

Based on her medical history and HIES score⁴ (41 points), a diagnosis of HIES was made. We initiated treatment with omalizumab (recombinant monoclonal anti-IgE antibodies), 450 mg subcutaneously every 2 weeks. This is the maximum dose reported⁵ in the context of immunosuppressant limitations imposed by a history of hepatitis C infection and failure of conventional therapies. One month after initiating omalizumab treatment, her dermatitis and asthma were markedly improved. She attained complete remission after 3 months of treatment, and her eosinophil differential count decreased to 2.4%. She remained well 6 months after stopping treatment (**Figure**, B).

Comment. Omalizumab binds to the FcεRI receptor binding site on the IgE molecule thereby inhibiting its binding to its receptor on mast cells and basophils and their subsequent activation.⁵ Dosing is based on body weight and baseline serum IgE concentration not exceeding 700 IU/mL to administer at least 0.016 mg/kg/IU of IgE per milliliter of serum over a 4-week period.

The dermatitis associated with HIES is attributed to the increased IgE-stimulated mast cell release of hista-

mine. In light of omalizumab's success in treating various atopic diseases associated with elevated IgE levels,⁶ we attempted a trial of omalizumab. Although costly, omalizumab has proven beneficial in a patient with HIES and very limited therapeutic options.

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Successful Treatment of Eruptive Unilateral Angiokeratomas

We report a case of eruptive unilateral angiokeratomas with no underlying vascular abnormality. To our knowledge, only 1 other case report of unilateral eruptive angiokeratomas has been reported, and that case was isolated to the scrotum with an underlying vascular defect.¹ Other reports of eruptive angiokeratomas associated with cerebral hemangiomas and trauma have been documented, but none of these cases were unilateral.²⁻⁵

Report of a Case. A 51-year-old African American woman presented to our clinic with a 1-week history of eruptive lesions over her left hand. Physical examination revealed approximately 1 dozen 2- to 4-mm black papules on the dorsal and palmar surfaces of the left hand. These lesions were firm, nonblanchable, and nonpulsatile. The distribution could not be isolated to any particular dermatome. No axillary lymphadenopathy was present. The lesions were asymptomatic. New lesions continued to erupt daily. Two weeks after the initial appearance of these lesions, she developed approximately 50 new lesions on the palmar and dorsal surfaces of her left hand (**Figure 1**).

The patient's medical history was notable only for hypertension controlled with medication. The patient denied any personal or family history of similar lesions or any other vascular malformations. A full review of sys-



Figure 1. Patient's left hand before treatment showing both dorsal (A) and palmar (B) surfaces affected by eruptive lesions.

tems turned up no positive findings. There was no history of trauma, vasculitis, carcinoma, hypercoagulable disorders, or other autoimmune disorders. A biopsy specimen from the left palm showed the classic histologic features of an angiokeratoma.

Our patient was treated with 2 separate techniques: electrocautery and laser therapy. The patient did not tolerate electrocautery well, and therefore most of the lesions were treated with pulsed-dye laser. Complete resolution of the lesions was achieved with 2 treatments, 1 month apart, using a 585-nm pulsed-dye laser (SPTL-1b Vascular Lesion Laser; Candela Corp, Wayland, Massachusetts). During both treatment visits, double pulse stacking was used with a 300- to 500-microsecond pulse duration and a fluence of 5.5 J/cm² with a 5-mm spot size. The patient tolerated 85 pulses on the first treatment and 71 pulses during the second treatment with these laser settings. No external cooling mechanism or suction was used because the laser was not equipped with either mechanism. No residual scarring or dyschromia occurred. The patient was seen 3 months after the final treatment, and only a single resolving lesion remained. All other lesions had resolved (**Figure 2**). There was no recurrence after 10 months, and the patient was pleased with the aesthetic result.

Comment. The word *angiokeratoma* is derived from the Greek words for vessels, horn, and tumor. The mechanism and pathogenesis of these lesions is not known. Angiokeratomas are common benign superficial vascular mal-