P8375

Giant pyoderma gangrenosum with myiasis

Gonzalo Blasco Morente, PhD, Virgen de las Nieves Universitary Hospital, Granada, Spain; Aurelio Martín Castro, MD, Virgen de las Nieves Universitary Hospital, Granada, Spain; Carmen Martínez Peinado, PhD, Virgen de las Nieves Universitary Hospital, Granada, Spain; Cristina Garrido Colmenero, PhD, Virgen de las Nieves Universitary Hospital, Granada, Spain; Eliseo Martínez García, PhD, Virgen de las Nieves Universitary Hospital, Granada, Spain; José Aneiros-Fernández, PhD, Virgen de las Nieves Universitary Hospital, Granada, Spain; Salvador Arias Santiago, MD, Virgen de las Nieves Universitary Hospital, Granada, Spain

Introduction: Pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis characterized by painful, necrotic ulceration. It typically affects patients in the third to sixth decades of life, with almost equal incidence in men and women. PG occurs most frequently on the lower extremities, but it is exceptionally associated with myiasis. A case of PG with large size and myiasis is reported.

Case report: A 76-year-old woman with a history of hypertension and dyslipidemia referred recurrent painful ulcer on the legs for 5 years. No other systemic signs. Now she presented an ulcerated plaque of 20 × 11 cm with irregular edges and fibrinous center with 2 larvae in her left leg, In contralateral foot, she presented an ulcerated plaque of 3 cm with similar characteristics. Blood tests were normal, except for C-reactive protein: 10.2 mg/dL, autoantibodies were negative, and imaging test found no abnormalities. Histopathology was suggestive of PG. In microbiology fly larvae were identified, it was unable to classify species. Suspecting PG with myiasis we retired larvae and prescribed treatment with prednisone 90 mg every 24 hours and mycophenolate mofetil 500 mg every 24 hours, along with local treatment. Steroid dose was gradually decreased to 50 mg/day and mycophenolate was increased to 2 g/day. The patient experienced progressive improvement until complete resolution of the clinic in 2 months.

Discussion: PG is a rare disorder, included in the spectrum of neutrophilic and autoinflammatory dermatoses. Half of PG cases are seen in association with systemic disease, not found in our case. Mimickers include infection, vascular insufficiency ulcers, systemic vasculitis, autoimmune disease, cancer, and exogenous tissue injury, among others. Misdiagnosis can lead to considerable morbidity, specifically, unnecessary treatments, multiple ineffective surgeries, and possible amputation. To the best of our knowledge, we have not found an association between PG and myiasis described in the literature. Larval therapy is an effective treatment in debridement of ulcers, but the presence of unsterilized larvae can cause superinfection, so mechanical removal is recommended. Prednisone and cyclosporine have been mainstays of systemic treatment for PG, although increasing evidence supports the use of biologic therapies. Our patient improved with treatment with prednisone 1 mg/kg/day and mycophenolate.

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P8717

Reflectance confocal microscopy as a noninvasive diagnostic tool for Hailey–Hailey disease

Marta Kurzeja, MD, PhD, Department of Dermatology CSK MSW, Warsaw, Poland; Joanna Czuwara, MD, PhD, Department of Dermatology CSK MSW, Warsaw, Poland; Lidia Rudnicka, MD, PhD, Lidia Rudnicka, Warszawa, Poland; Malgorzata Olszewska, MD, PhD, Department of Dermatology Medical University of Warsaw, Warsaw, Poland

Reflectance confocal microscopy (RCM) allows noninvasive imaging of the epidermis and superficial dermis. The aim of the study was to evaluate the potential usefulness of RCM in diagnosing Hailey–Hailey disease (familial benign chronic pemphigus). Three patients with Hailey–Hailey disease were examined by RCM. Skin biopsies were taken at the site of RCM examination to evaluate possible correlations of RCM with histopathology. In all patients (3/3; 100%) the most sticking RCM feature was acantholysis resembling a dilapidated brick wall at the level of the granular and spinous layer. Other RCM features included: crust on the skin surface, epidermal disarray, intraepidermal clefts, and inflammatory cells in the epidermis and in the superficial dermis. Detailed analysis revealed a good correlation between RCM and histopathology findings. In conclusion, reflectance confocal microscopy examination is a promising noninvasive diagnostic tool for diagnosing Hailey–Hailey disease.

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P7664

Postpartum pemphigoid gestationis

Pamela Morganroth, MD, Henry Ford Hospital, Detroit, MI, United States; Chauncey McHargue, MD, Henry Ford Hospital, Detroit, MI, United States

Background: Pemphigoid gestationis is a rare vesiculobullous disorder associated with pregnancy. Although the disease typically presents during late pregnancy, it can also occur during early pregnancy or in the immediate postpartum period. We present a case of postpartum pemphigoid gestationis with extensive cutaneous disease.

Case report: A 24-year-old white female presented with a widespread intensely pruritic eruption that started 4 days postpartum. The patient had an extensive cutaneous eruption involving most of her trunk, including the periumbilical area, and all 4 extremities. Her skin lesions had multiple morphologies, including polycyclic vesicles overlying erythematous plaques and targetoid erythematous plaques with dusky centers. Biopsy of a vesicle showed a subepidermal vesicle with numerous eosniophils. Perilesional direct immunofluorescence showed linear C3 deposition at the dermoepidermal junction. Pemphigoid gestationis was diagnosed, and the patient was treated with oral prednisone, which resulted in dramatic improvement within days. The patient's full-term newborn baby did not have any cutaneous lesions.

Discussion: Although pemphigoid gestationis typically occurs during pregnancy, this case reminds us that it can also occur postpartum. Pemphigoid gestationis is particularly important to recognize because it is associated with an increased risk of Graves disease, premature delivery, and a small risk of skin lesions in the newborn. Patient counseling is also essential because pemphigoid gestationis usually recurs in subsequent pregnancies and may also recur with the menstrual cycle and with oral contraceptive use.

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Resolution of pemphigus foliaceus after hematopoietic stem cell transplant

Mary Pilcher, MD, West Virginia University, Morgantown, WV, United States; Rodney Kovach, MD, West Virginia University, Morgantown, WV, United States; Roxann Powers, MD, West Virginia University, Morgantown, WV, United States

Hematopoietic stem cell transplant (HSCT) has been used for many years to treat malignancies of the bone marrow and many genetic defects. In recent years, the concept of treating autoimmune disease with HSCT has gained recognition. Pemphigus foliaceus is a chronic autoimmune disease that is confined to the skin and is difficult to treat. Many patients respond to immunosuppressive drugs, such as prednisone, azathioprine, mycophenolate mofetil, and cyclophosphamide, but must be maintained on these drugs indefinitely to maintain remission. These patients are therefore subjected to the long-term side effects of these medications. We present a case of pemphigus foliaceus in which the patient obtained sustained remission without need for immunosuppressive medications after HSCT for another condition.

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