All specimens tested negative for microorganisms by Grocott-Gomori methenamine silver and acid-fast bacilli stains and were not consistent with lichen sclerosus or lichen simplex chronicus. Hydroxychloroquine therapy had previously failed for this patient, and she was therefore prescribed mycophenolate (500 mg, twice a day) by her pulmonologist. In addition, we recommended 3-times-daily application of topical clobetasol ointment and nightly hydrocortisone suppositories.

Discussion | Although it is rare, sarcoidosis may occur in other regions of the female genital tract, including the uterus, ovaries, fallopian tubes, cervix, placenta, and vaginal wall. The differential diagnosis of granulomatous disease of the vulva includes tuberculosis, Crohn disease, syphilis, foreign body reactions, and lymphogranuloma venereum.

Our case demonstrates TEE, a histologic phenomenon that, to our knowledge, has never been reported in a case of vulvar sarcoidosis. During TEE, the epidermis or the follicular epithelium undergoes a process of pseudohyperplasia to encompass the targeted material, which is eventually eliminated by maturing keratinocytes. Importantly, the epithelium does not undergo major structural remodeling and returns to normal after the targeted material is successfully removed. In the dermis of our patient’s vulva, the granulomas were the targeted material to be removed.

In summary, our patient presented with sarcoidosis of the vulva with histologic evidence of TEE. This case highlights that sarcoidosis can have an uncommon presentation, warranting extensive physical examination of patients presenting with possible sarcoidosis.

Stacey Watkins, BA
Aimen Ismail, BS
Kristopher McKay, MD
Kathleen Beckum, MD
Vlada Groysman, MD

Hypertensive Emergency, Matlike Telangiectasias, and Calciphylaxis in POEMS Syndrome

POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes) is a paraneoplastic syndrome caused by plasma cell dyscrasias. We report hypertensive emergency and acral matlike telangiectasias as novel manifestations of POEMS syndrome (POEMS). Also, to our knowledge, this is the first reported case of calciphylaxis and POEMS occurring in the absence of sclerotic bone lesions or a peripheral monoclonal gammopathy, highlighting the diagnostic challenges posed by this disease.

Report of a Case | A woman in her 30s with a 2-year history of distal polyneuropathy and matlike telangiectasias presented with a hypertensive emergency and peak blood pressure of 247/154 mm Hg. Workup revealed a serious pericardial effu-
sion, hepatosplenomegaly, ascites, shotty lymphadenopathy, and diffuse subcutaneous edema; the remainder of her workup findings were negative, including those for rheumatoid factor, antineutrophil cytoplasmic antibodies, antinuclear antibodies, extractable nuclear antigen antibodies panel, serum and urine protein electrophoresis with immunofixation, and brain magnetic resonance imaging. After blood pressure was brought under control, prednisone treatment was empirically initiated, and the ascites improved.

Over the next 4 months, the patient developed hemangiomas, amenorrhea, refractory ascites, hepatosplenomegaly, weight loss, and fatigue. On readmission, she was found to have hypothyroidism, diabetes mellitus, hypogonadism, and papilledema. Significant laboratory findings included thrombocytosis (platelet level peaking at 2045 × 10^9/dL) and transient renal insufficiency (creatinine level peaking at 3.04 mg/dL, then quickly decreasing). (To convert creatinine to micromoles per liter, multiply by 88.4.) Parathyroid hormone level was within normal limits, and the level of calcium phosphate product was 43.12 mg/dL (normal, <55.00 mg/dL).

Light chain concentrations, both λ and κ, were elevated at 35 mg/dL (normal, 0.57-2.63 mg/dL) and 4.2 mg/dL (normal, 0.33-1.94 mg/dL), respectively, and the κ/λ ratio was low, at 0.12 (normal ratio, 0.26-1.65). The finding of a blood test for human herpesvirus 8 (HHV-8) was negative. Results of bone scans and skeletal surveys were unremarkable.

A dermatology consult was requested to evaluate painful abdominal purpura of 2 weeks’ duration. The patient was cachectic with bulging flanks and lower extremity edema; tender purpuric retiform plaques and eschars were present on the abdomen (Figure 1), and telangiectatic patches were noted on the dorsal surfaces of the feet (Figure 2A). Scattered on her lips, trunk, and extremities were numerous hemangiomas. A biopsy specimen from the abdomen demonstrated subcutaneous intravascular calcification, consistent with calciphylaxis.

Bone-marrow biopsy revealed atypical megakaryocyte hyperplasia, and flow cytometry revealed a small, monoclonal, λ-restricted plasma cell population, which, together with her symptoms, physical examination findings, and laboratory results, were consistent with POEMS. The patient’s level of vascular endothelial growth factor (VEGF) was unremarkable, but she was taking prednisone. Weekly infusions with cyclophosphamide and dexamethasone were started, followed by marked general improvement and resolution of her telangiectasias (Figure 2B). Reducing treatment to every 3 weeks resulted in worsening ascites and calciphylaxis and plasma VEGF levels increasing to 1499 pg/mL (normal, 0-115 pg/mL). Resuming weekly infusions and adding bortezomib to her treatment regimen resulted in substantial improvement.

**Discussion** | POEMS has diverse cutaneous manifestations, including hyperpigmentation, sclerodermoid changes, acrocyanosis, acquired facial lipoatrophy, hemangiomas, hypertrichosis, Terry’s nails, and clubbing. While telangiectasias have been reported in POEMS, they are not well characterized. The acral matlike telangiectasias in this patient were striking, similar to those in systemic sclerosis. Calciphylaxis in POEMS has been reported, but in all 5 cases, it occurred concomitantly with a monoclonal gammopathy or sclerotic bone lesions. In cases of POEMS associated with diffuse bone-marrow infiltration, radiologic findings may be absent, and the monoclonal popu-
loration small, with no detectable M-protein. Diagnosis requires either a bone-marrow biopsy with flow cytometry or recognition of atypical megakaryocyte hyperplasia and plasma cell rimming around lymphoid aggregates.5 To our knowledge, our patient’s presentation with hypertensive emergency has not been reported in POEMS and may be connected to the proliferative effects of VEGF on the glomerular endothelium. POEMS can be associated with multicentric Castleman disease; in this setting the result of HHV-8 testing is usually positive.6

POEMS may be treated with alkylating agents, corticosteroids, immunomodulatory agents, and autologous stem cell transplantation. The proteasome inhibitor bortezomib has shown significant promise through NF-κB inhibition in plasma cells and possible direct effects on endothelial cells via inhibition of VEGF and angiogenesis.7

Meg R. Gerstenblith, MD
Kord S. Honda, MD
Erica Campagnaro, MD
Meg R. Gerstenblith, MD

Author Affiliations: Department of Dermatology, University Hospitals Case Medical Center, Case Western Reserve University School of Medicine, Cleveland, Ohio (Novoa, Honda, Gerstenblith); Department of Medicine, University Hospitals Case Medical Center, Case Western Reserve University School of Medicine, Cleveland, Ohio (Campagnaro).

Corresponding Author: Meg R. Gerstenblith, MD, Department of Dermatology, University Hospitals Case Medical Center, Case Western Reserve University School of Medicine, 1110 Euclid Ave, Lakeside Third Floor, Cleveland, OH 44106 (meg.gerstenblith@uhhospitals.org).


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Mapping Biopsy Procedure on Management of Severe Buruli Ulcer Due to Mycobacterium ulcerans, Subspecies shinshuense

Report of a Case | A healthy male teenager presented with a 2-month history of eruptions on the dorsum of his left hand, which showed diffuse painful erythema and a 2-cm ulcer. Results of blood tests were normal but for slightly elevated C-reactive protein levels (0.46 mg/dL). Culture results from the ulcer were negative for any general bacteria. Computed tomography showed neither gas production nor abscess formation. Histologic examination of the erythematous lesion revealed diffuse necrosis, and Ziehl-Neelsen (Z-N) staining showed many acid-fast bacilli in the subcutis. We diagnosed cutaneous mycobacterial infection.

Despite treatment with oral clarithromycin, 800 mg/d, and levofloxacin, 500 mg/d, the lesion expanded. Therefore rifampin, 450 mg/d, was added to the treatment regimen 7 days after the initiation of therapy. We performed debridement on day 8 (Figure, A). Although almost all the skin on the dorsum of his hand was debrided, the erythema further expanded. We performed a second debridement on day 25 and additionally excised the erythematous lesion toward the wrist, including the area labeled 8 in Figure, B, and debrided necrotic tissue. The lesion rapidly improved after the second debridement, although some finger extensor tendons gradually became necrotic, and reconstruction of tendons was needed. The patient showed no recurrence at 1-year follow-up.

At 4 weeks, growth of a yellow-white colony in Ogawa egg medium was observed. Analysis using DNA-DNA hybridization (Kyokuto, Japan) demonstrated Mycobacterium marinum bacteria. Polymerase chain reaction (PCR) analysis targeting insertion sequence IS24041,2 and pathogenic plasmid MUM001 gene sequences3 revealed Buruli ulcer caused by Mycobacterium ulcerans subspecies shinshuense, a rare mycobacterium phenotypically close to M ulcerans that produces a toxin resembling mycolactone.3-4 We note that M ulcerans subspecies shinshuense can also affect tendons and muscles, as in our case and at least 1 other case.5 Recommendations concerning the duration of antibiotic treatment and timing of surgical intervention for Buruli ulcer have been offered in preliminary guidelines issued by the World Health Organization, but there is no reliable evidence to determine the extent of surgical excision.

Discussion | Using a 3-mm punch, we performed multiple biopsies from 13 erythematous sites on our patient’s hand and wrist (Figure, A-D) before first debridement at points 1 through 6 (Figure, A) and before second debridement at points 7 through 13 (Figure, B-D). We retrospectively examined the relationship among clinical presentations, histologic findings (hematoxylin-eosin [H-E] and Z-N stainings), and results of PCR analysis for IS2404 (Figure, G) to demonstrate the features of sites that needed debridement (Table). From the clinical course, we determined that all sites of first debridement (samples 1-6) and the area around sample 8 of the second debridement needed surgical resection. We performed the second debridement to address severe postoperative functional limitations and in-

Letters

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