tion compliance and minimize dose reductions and discontinuation of the drug treatment.

Maider Pretel, MD, PhD
Mercedes Íñarrairaegui, MD, PhD
José Miguel Lera, MD
Leyre Aguado, MD, PhD
Miguel Angel Idoate, MD, PhD

Author Affiliations: Department of Dermatology, School of Medicine, University Clinic of Navarra, Pamplona, Spain (Pretel, Íñarrairaegui, Lera, Aguado); Department of Pathology, School of Medicine, University Clinic of Navarra, Pamplona, Spain (Idoate).

Corresponding Author: Maider Pretel, MD, PhD, Department of Dermatology, University Clinic of Navarra, Avda Pío XII s/n, 31080 Pamplona, Spain (mpretel@unav.es).

Published Online: March 26, 2014. doi:10.1001/jamadermatol.2013.6924.

Conflict of Interest Disclosures: None reported.


Systemic Sarcoidosis With Unique Vulvar Involvement

Sarcoidosis is a complex, multisystem disease with an unclear cause. Research suggests that the pathogenetic mechanism of sarcoidosis is dysregulation of the immune system in individuals with a genetic predisposition who are subsequently exposed to inciting environmental agents. Cutaneous sarcoidosis is often one of the earliest clinical signs of the disease and can be divided into 2 subclasses: specific and nonspecific lesions. Specific lesions are characterized by granulomas identified histologically and include macules, papules, plaques, annular lesions, lupus pernio, infiltration of scars, and subcutaneous nodules. Alternatively, nonspecific sarcoid lesions, including erythema nodosum, prurigo, or calcifications, are reactive inflammatory processes.

Report of a Case | A woman in her 40s presented with erythematous papules with perinasal and periocular distribution involving both the upper and lower eyelids and an atrophic plaque on the dorsal surface of her neck. The patient complained of vaginal changes including itching, burning, tearing, pain with intercourse, and a painful lesion in the perianal area. Examination revealed an erythematous scaly plaque on the mons pubis with atrophic, white, discolored patches in the vaginal area without evidence of tearing (Figure 1).

The patient presented with a 5-week history of productive cough with exertional dyspnea, and her chest radiograph revealed upper-lobe calcified granulomas but no bilateral hilar lymphadenopathy. The patient's pulmonary function tests revealed no airflow limitations, normal lung volume, and a mild reduction in the diffusing capacity of the lungs for carbon monoxide. The results of blood tests, including complete blood cell count, comprehensive metabolic panel, angiotensin-converting enzyme level, and erythrocyte sedimentation rate, were within normal limits.

Biopsies of the vulva and mons pubis revealed pauci-inflammatory nodular granulomas consistent with sarcoidosis. On examination of the epidermis, vaguely psoriasiform epidermal acanthosis with hyperkeratosis was found. Within the dermis, tissue biopsy specimens demonstrated numerous, well-circumscribed nodular granulomas with multinucleated giant cells and lacking significant numbers of neutrophils (Figure 2A). Most of the granulomas were pauci-inflammatory, but some had a cuff of lymphoplasmacytic infiltration. Finally, in areas of the epidermis there was transepidermal elimination (TEE) of the granulomas, a unique finding in the vulvar area of a patient with sarcoidosis.
(Figure 2B). 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 gammapathy, 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 (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 gammapathy, 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-

Author Affiliations: Department of Dermatology, University of Alabama, Birmingham (Watkins, Ismail, McKay, Beckum, Groysman).

Corresponding Author: Vlada Groysman, MD, Department of Dermatology, University of Alabama at Birmingham, 1530 Third Ave S, EFH 414, Birmingham, AL 35294 (VGroysman@cahabaderm.com).

Published Online: March 5, 2014.

Conflict of Interest Disclosures: None reported.