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Case presentation

Lupus miliaris disseminatus faciei

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Abstract

Lupus miliaris disseminatus faciei (LMDF) is a rare, inflammatory condition, which is characterized by red-brown and yellow-brown papules on the face, with characteristic involvement of the eyelids and with histopathologic findings of suppurative and granulomatous folliculitis and dermatitis. The etiology of this disease is not known, but retinoids, anti-inflammatory, immunosuppressive, and antimicrobial medications are utilized to treat the condition with variable results. We present the case of a patient with LMDF that has thus far been refractory to treatment.

Case synopsis

History: The patient is a 28-year-old man who presented to the Skin and Cancer Unit in November, 2014, with a papulopustular eruption on the face that initially had developed in June, 2014. The patient initially developed a lesion under his left eye. Two weeks later, he developed a febrile illness while traveling in Brazil; a week after this, he noted swelling of the face, which included the eyes and cheeks but without lip swelling, that resolved with the use of fexofenadine. He then developed a papulopustular eruption over the infraorbital skin, chin, and nose. A topical antifungal cream and topical glucocorticoids were prescribed but were ineffective. The patient presented to an outside dermatologist in July and doxycycline 100 mg daily and topical metronidazole gel were started for 30 days without improvement. The eruption continued to spread and a prednisone taper was initiated, with continued progression of the eruption. A biopsy specimen of the left lower eyelid skin showed a granulomatous dermatitis; a periodic acid-Schiff stain was negative. Minocycline was started without improvement, but the eruption flared after minocycline was discontinued. In September, isotretinoin 40 mg daily was started with an increase to 60 mg daily one month later. The eruption spread supraorbitally and became more prominent over the cheeks. A potassium hydroxide preparation showed a few *Demodex* mites and the patient was treated empirically with permethrin. A biopsy specimen from the right nasolabial fold skin showed subepidermal fibrin deposition, solar elastosis, and dilated, thin-walled blood vessels.

The patient returned to the Skin and Cancer Unit. The history was consistent with the above description with the addition that he denied flushing or flushing/flaring with alcohol or spicy foods. The patient was using Vanicream on the face and isotretinoin 60 mg daily. Review of systems was negative. Past medical history included eczema and seasonal allergies.

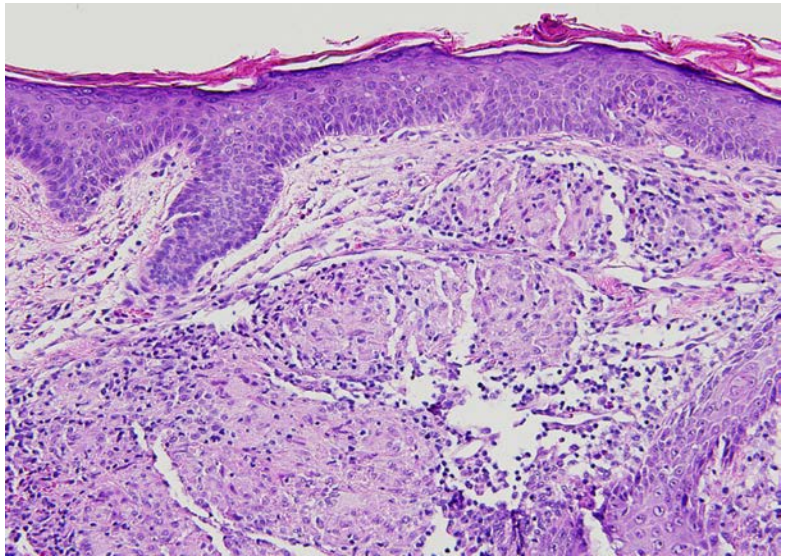
Considering the lack of improvement with the use of isotretinoin and the development of muscle and back pains since starting the medication, isotretinoin was discontinued. Doxycycline was prescribed to treat a diagnosis of granulomatous rosacea. The patient remained on doxycycline for approximately three months, initially 100 mg twice daily and then three times daily and also benzoyl peroxide wash (no improvement), sulfur precipitate 5% in calamine lotion (no improvement), azelaic acid (discontinued secondary

to eye and skin irritation), and pimecrolimus ointment (caused burning of the skin). The patient did not develop new lesions during this time, but there was minimal improvement while on doxycycline. A biopsy was obtained.

Physical examination: There were small, discrete and grouped, red-yellow papules, vesicles, and pink-yellow pustules, some umbilicated and some dome-shaped. Some of the papules were crusted and some coalesced over the upper eyelids, lateral canthi, lower lids, glabella, cheeks, temples, perinasal skin, upper lip, mental crease, and neck.

Laboratory data: A complete blood count with differential analysis and a hepatic panel were normal. A superficial wound culture showed no growth. Herpes simplex virus was not detected with polymerase chain reaction. QuantiFERON gold and human immunodeficiency virus were negative.

Histopathology: There is a disrupted and distorted follicular infundibulum within which are numerous neutrophils that are surrounded by a granulomatous infiltrate that is comprised predominantly of epithelioid and occasional multinucleated histiocytes. A periodic acid-Schiff stain with diastase and an acid-fast bacillus stain fail to show microorganisms. No polarizable foreign material is present.



Figures 1. Papulopustular facial eruption Figure 2. Epithelioid granulomatous infiltration in the dermis

Discussion

Diagnosis: Lupus miliaris disseminatus faciei

Comment: Lupus miliaris disseminatus faciei (LMDF) is a rare, inflammatory and granulomatous dermatosis of the face with many postulated etiologies. Although once associated with *Mycobacterium tuberculosis* infection owing to the frequent presence of central caseating necrosis on histopathologic examination, the use of polymerase chain reaction and culture have shown no evidence of *Mycobacterium* [1]. Similarly, lesions were once associated with sarcoidosis, but lack of other confirmatory features for sarcoidosis made this association less likely [2]. More recently, some have categorized LMDF as a form of granulomatous rosacea or a rosacea-like syndrome that is a distinct entity, owing to the commonality of symmetrically distributed and similarly appearing papules on the face that surround the pilosebaceous units [3]. Furthermore, a recent case series showed that *Propionibacterium acnes* DNA could be isolated in tissue samples from all nine cases of LMDF that were studied [4]. A literature review showed that most authors believe LMDF and rosacea to be similar yet distinct entities. Although the exact pathogenesis is unknown, many authors have suggested that granulomas in LMDF form in association with inflammation and destruction of pilosebaceous units [4, 5].

The incidence of LMDF is unknown but is thought to be rare, with a possible greater prevalence in Japan that is based on the reported cases [6]. It affects both genders although most cases have been in men. It occurs most commonly in young adults, such as our patient, although rare cases that involve the elderly [7] and children [8] have been reported.

LMDF presents as red-brown or yellow-brown papules, which eventually evolve into pustules and then pigmented scars upon involution and regression [9]. Although typically presenting on the face and neck with characteristic involvement of the lower

eyelids [4], extrafacial manifestations have been reported, with involvement of the axillae [10, 11], hands, legs, and genitals [12]. Some clinical manifestations are distinct from those of rosacea. As in our patient, LMDF patients do not experience flushing in response to spicy foods or alcohol [2]. Most patients experience a self-limited course, with lesions resolving in one to four years [2]. Despite the usual spontaneous resolution, no treatment has consistently been able to prevent the disfiguring scars that the disease produces [3].

Histopathologic features of most cases of LMDF include epithelioid granulomatous inflammation, with caseous necrosis that surrounds the pilosebaceous units [4]. However, other histopathologic findings have been described, with reports of several patients with sarcoid-like granulomas and a non-specific perifollicular lymphohistiocytic infiltrate [3].

Aside from LMDF and rosacea, the differential diagnosis also includes eczema herpeticum, molluscum contagiosum, sarcoidosis, and Crohn disease. Results of our patient's evaluation precluded a role for herpes simplex virus or *Mycobacterium tuberculosis*, and the patient had no systemic symptoms that were suggestive of a systemic disease. Considering the patient's travel history and febrile illness that preceded the onset of the eruption, an infectious trigger has not necessarily been ruled out. Additionally, our patient was noted to have *Demodex* mites on a microscopy specimen. *Demodex* mites are reported to be present at a higher density in the skin of rosacea patients although they may be normal colonizers of healthy patients. A role for these mites in the pathogenesis of rosacea has been discussed [13], but it is yet unknown if they have any association with LMDF.

Although no established treatment guidelines exist, many successful therapies have been reported in the literature. Oral glucocorticoids [14], dapsone [15], tetracycline [16], and tranilast [17] have been reported as successful treatments. Intramuscular glucocorticoids [18], isotretinoin [19, 20], and clofazimine [21] also have been shown to be efficacious. More recently, combination therapy has been utilized to improve the clinical response. Successful treatment of LMDF with combined oral metronidazole and topical tacrolimus ointment has been reported [22]. Oral dapsone combined with topical tacrolimus ointment also has yielded good results [3]. Finally, laser therapy also may have a role in the treatment of this disease as successful treatment of patients has been reported with a non-ablative fractionated 1,565 nm laser and a 1,450 nm diode laser [6, 23]. The continued availability of new potential treatments for LMDF may reduce patient morbidity and ultimately shed light on the pathogenesis of this complex entity.

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