Extrafacial nuchal lupus miliaris disseminatus faciei

Laura G. Pruitt, MD,[a] Carley O. Fowler, MD,[b] Robert N. Page, MD,[c,d]
Neil M. Coleman, MD,[c,d] and Roy King, MD[c,d]
Greenville, North Carolina and Knoxville, Tennessee

Key words: acne agminata; caseation necrosis; diascopy; granuloma; granulomatous disease; histopathology; lupus miliaris disseminatus faciei.

INTRODUCTION

Lupus miliaris disseminatus faciei (LMDF) is an uncommon granulomatous disease of unknown origin. It most commonly presents as yellow-brown to red papules on the central face and often involves the lower eyelids. Extrafacial manifestations have been reported but almost always coexist with facial involvement. Histopathology findings show epithelioid cell granulomas with variable caseous necrosis depending on the timing of the biopsy. We present a case of LMDF occurring solely on the posterior neck without facial involvement.

CASE REPORT

A 65-year-old white woman with a medical history of cutaneous squamous cell carcinoma of the arm and epidermal inclusion cysts presented with numerous small papules on the posterior neck that were present for several months before consultation. She denied any associated symptoms including pruritus, pain, or burning. She denied a history of flushing, recurrent lesions on her neck, lung disease, and infection with tuberculosis. Examination found a group of approximately twelve 2- to 3-mm dome-shaped yellow-brown papules on the posterior neck (Fig 1). Similar lesions were not observed elsewhere on the body. The differential diagnosis included milia, herpes simplex virus infection, and elastosis perforans serpiginosa. A 4-mm punch biopsy found a well-formed granuloma with central caseous necrosis accompanied by a perivascular lymphocytic infiltrate (Figs 2 and 3). The overlying epidermis was unaffected. Periodic acid-Schiff and Fite's acid-fast stains were negative for microorganisms. The patient declined treatment, and at follow-up 1 month later, the lesions were unchanged.

Abbreviation used:

LMDF: lupus miliaris disseminatus faciei

DISCUSSION

LMDF, also known as acne agminata, was first described in 1878,[1] and approximately 200 cases have since been reported in the literature.[2] It most commonly manifests as solitary or multiple, discrete, 1- to 3-mm skin-colored to yellow-brown to red papules involving the central and lateral aspects of the face and lower eyelids with occasional extension to the chin and neck. Diascopy of the lesions often finds an apple-jelly color.[3] Extrafacial lesions are not infrequent but almost always coexist with facial involvement.[4] There are 2 previous reports of isolated axillary LMDF[4,5] and 1 report of LMDF involving the neck and chest without facial involvement.[2] Solitary lesions on the antihelix and lower back have also been reported.[6]

The hallmark histopathologic feature of LMDF is an epithelioid cell granuloma with caseous necrosis.[3] However, the changes vary according to the age of the lesions at the time of biopsy. Early lesions typically lack granuloma formation, and only a nonspecific perivascular and periappendageal infiltrate of lymphocytes, histiocytes, and scattered neutrophils may be seen. As lesions become fully

From East Carolina University Dermatology[a]; Dermatology Associates of Knoxville[b]; Dermatopathology Partners of Knoxville[c]; and University of Tennessee Graduate School of Medicine at Knoxville, Department of Pathology.[d]

Funding sources: None.

Conflicts of interest: None declared.

This case was a poster presentation at the American Society of Dermatopathology meeting, October 27-30, 2016, Chicago, Illinois.

Correspondence to: Laura G. Pruitt, MD, ECU Dermatology, 517 Moye Blvd, Greenville, NC 27834. E-mail: pruittl16@ecu.edu.

JAAD Case Reports 2017;3:319-21.

2352-5126

http://dx.doi.org/10.1016/j.jdcr.2017.04.009

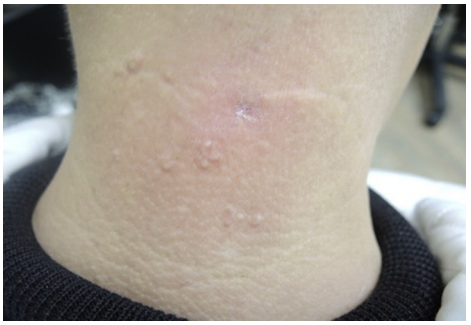

Fig 1. Lupus miliaris disseminatus faciei. Approximately twelve 2- to 3-mm dome-shaped skin-colored papules on the posterior neck. Telangiectasia is absent.

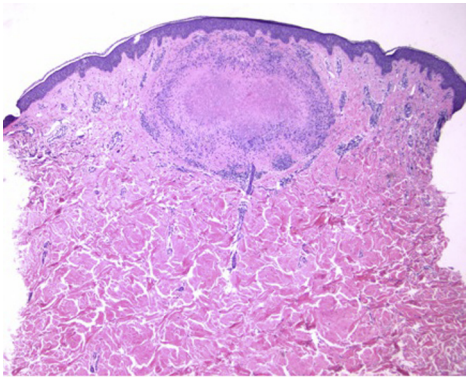

Fig 2. Histopathology of lupus miliaris disseminatus faciei. A well-formed granuloma with central caseous necrosis accompanied by a perivascular lymphocytic infiltrate. (Hematoxylin-eosin stain.)

developed, perifollicular epithelioid cell granulomas form. Three histologic stages have been described: epithelioid cell granuloma without caseous necrosis, epithelioid cell granuloma with neutrophilic abscess formation, and epithelioid cell granuloma with caseous necrosis. Perifollicular fibrosis with scattered lymphocytes and histiocytes are seen in late lesions.[7,8]

The etiology and pathogenesis of LMDF remains unclear. Originally, it was thought to be a reaction pattern to *Mycobacterium tuberculosis*; however, there is no evidence to date supporting this idea.[9] More recently, LMDF has been viewed as a type of granulomatous rosacea, as the granulomas in LMDF occur in association with pilosebaceous units, and epithelioid cell granulomas have been identified in some cases of rosacea.[10] However, LMDF occurs in different anatomic locations than rosacea; has no associated erythema, flushing, or telangiectasias; and has a self-limited course unlike rosacea. These significant differences make it difficult to accept this concept, and our case of exclusive posterior neck involvement supports the distinction

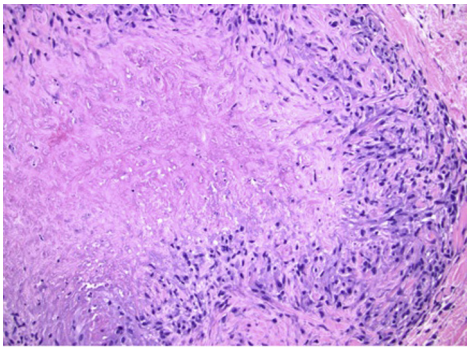

Fig 3. Histopathology of lupus miliaris disseminatus faciei. Caseous necrosis characteristic of well-developed lesions of lupus miliaris disseminatus faciei. (Hematoxylin-eosin stain.)

of LMDF from rosacea. LMDF has also been postulated to occur as a reaction to *Demodex folliculorum*, ruptured hair follicles, and ruptured epidermal inclusion cysts, but these associations have not been confirmed. LMDF has also been described as a micropapular form of sarcoidosis, but this diagnosis is easily ruled out with the history, physical examination, laboratory investigations, and chest radiograph.[11]

LMDF typically has a chronic but benign course with spontaneous involution in 12 to 24 months leaving behind small, pitted scars. Various treatments are reported in the literature, but specific treatment guidelines are lacking. Retinoid, anti-inflammatory, immunosuppressive, and antimicrobial medications as well as laser therapy have been used for the treatment of LMDF, but no single agent is consistently effective.[8,12,13] Tetracyclines are commonly used as a first-line treatment, but their efficacy in this disorder is often unsatisfactory. Treatment with oral prednisolone has been found to prevent scarring when initiated early in the disease course.[12] Given the rarity of LMDF and its tendency to spontaneously resolve, the effect of different therapies on the disorder is difficult to evaluate.

LMDF represents a diagnostic challenge given its highly diverse clinical presentations. Our patient's case represents an unusual clinical presentation of LMDF involving a localized area of the posterior neck without facial involvement and highlights the importance of characteristic histopathologic findings in establishing the diagnosis. This case also further supports LMDF as a distinct clinical entity from rosacea. We report this unusual case to remind clinicians that LMDF can occur in nonfacial areas without concurrent facial involvement.

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