Recurrent retroauricular cystic nodules: lichen planus follicularis tumidus

Thinh Chau[1*] BS, Nima Amini[2*] MD, Daniel B Eisen MD[3], and Maxwell A Fung MD[2,3]

[*]Authors contributed equally to this work

Affiliations: [1]School of Medicine, [2]Department of Pathology and Laboratory Medicine, [3]Department of Dermatology, University of California Davis, Sacramento, California, USA

Corresponding Author: Maxwell A. Fung MD, 3301 C Street, Suite 1400, Department of Dermatology, University of California Davis, Sacramento, CA 95816, Email: maxfung@ucdavis.edu

Abstract

Lichen planus follicularis tumidus (LPFT) is a rare subtype of lichen planus (LP) that has been most commonly described in middle-aged women. LPFT clinically manifests as recurrent cystic follicular nodules that preferentially involve the retroauricular area; concurrent classic LP lesions on the extremities and mucosal surfaces may also be present. Histologically, LPFT demonstrates epithelial-lined follicular cysts filled with orthokeratotic keratin surrounded by a dense lichenoid infiltrate. We present a case of a 67-year-old man with clinical and histopathologic findings consistent with LPFT and discuss differential diagnostic considerations for entities resembling LPFT. Lastly, treatment options for LPFT are reviewed.

Keywords: lichen planus follicularis tumidus, lichen planus, retroauricular cystic nodules, interface dermatitis, dermatopathology

Introduction

Lichen planus (LP) has many subtypes based on sites of involvement, clinical morphology, and histopathology. Of these, lichen planus follicularis tumidus (LPFT) is an unusually rare subtype first described by Belaich and colleagues in 1977 [1]. There have since been fewer than 20 reported cases in the literature. Most commonly occurring in middle-aged women, LPFT preferentially involves the retroauricular area and affected individuals may have concurrent classic LP lesions on the extremities and mucosal surfaces [2, 3]. Similar to classic LP, LPFT has been associated with systemic viral infection (hepatitis B and C) and T cell-mediated autoimmune disease (Hashimoto's thyroiditis), respectively [3, 4]. We herein report a case of a 67-year-old man presenting with recurrent retroauricular cystic nodules clinically and histologically consistent with LPFT. We review reported clinical and histopathologic findings of LPFT and discuss differential diagnostic considerations as well as treatment options.

Case Synopsis

A 67-year-old man was referred for evaluation of a nodule behind the right ear that had been present for greater than two years and previously biopsied twice, without specific diagnosis. The lesion was pruritic with occasional drainage. The patient reported having similar lesions behind both ears. Past medical history included a diagnosis of cutaneous squamous cell carcinoma (SCC) of the left retroauricular area from an outside institution. Physical examination revealed a 1.5cm, lichenified, hyperpigmented right retroauricular cystic nodule with a central punctum adjacent to a well-healed scar (**Figure 1**). A 1cm left retroauricular nodule with similar characteristics was also noted. There were also multiple smaller cystic nodules surrounded by comedones and double comedones on the zygomatic and occipital regions bilaterally. No extrafacial involvement was noted.

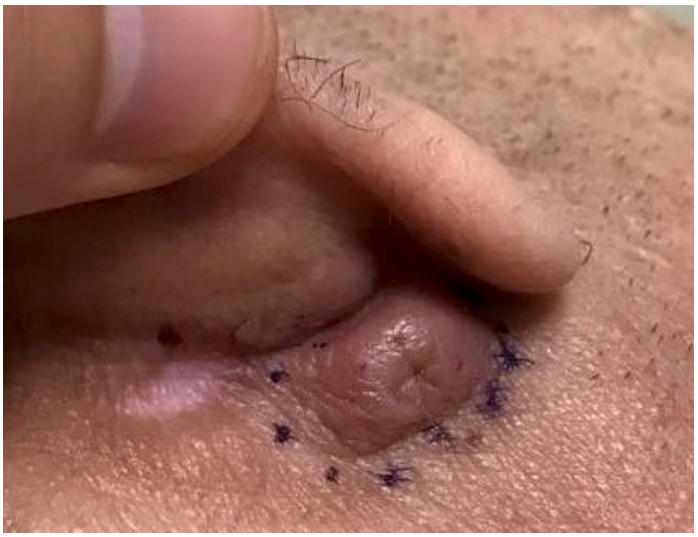

Figure 1. *Right postauricular cystic nodule with lichenification, hyperpigmentation, and central punctum adjacent to well-healed scar.*

Hematoxylin-eosin stained sections revealed epithelial-lined follicular cysts with irregular epithelial hyperplasia and hypergranulosis with jagged sawtooth-shaped rete ridges. The follicular cysts were surrounded by a dense lichenoid lymphocytic infiltrate with scattered eosinophils (**Figure 2**).

Case Discussion

Clinically, reported cases of LPFT document asymptomatic to intensely pruritic, red-violet plaques that contain multiple white-yellow cysts and comedones. Histologically, a dense band-like lichenoid infiltrate associated with epithelial hyperplasia and surrounding dilated follicular cysts containing orthokeratotic keratin is characteristic for this entity [1, 3, 5-7]. The differential diagnosis for LPFT includes: milia en plaque, steatocystoma multiplex, nevus comedonicus, Favre-Racouchot syndrome, follicular mucinosis, folliculotropic mycosis fungoides (MF) with cysts and comedones, and discoid lupus erythematosus (DLE). Despite their clinical resemblance, milia en plaque is characterized by extrafacial involvement while lacking the dense perifollicular lichenoid infiltrate characteristic of LPFT [8, 9]. Steatocystoma multiplex, which can be localized to the head and neck, features cysts with a crenulated eosinophilic lining and sebaceous lobules contiguous or adjacent to the cyst wall [10]. Nevus comedonicus, which occurs shortly after birth or in early childhood, has dilated follicular openings filled with lamellated keratin and devoid of hair shafts [11]. Favre-Racouchot syndrome is usually restricted to sun-exposed areas of the face and neck and exhibits marked solar elastosis on biopsy [12]. Folliculotropic MF and DLE require careful clinicopathologic correlation. In our case, the cystically dilated follicles with irregular epithelial hyperplasia and absent lymphocytic atypia, folliculotropism, and epitheliotropism excluded follicular MF. Our patient denied photosensitivity and his biopsy contained eosinophils while lacking increased interstitial dermal mucin deposition or basement membrane thickening, arguing against DLE. Direct immunofluorescence for

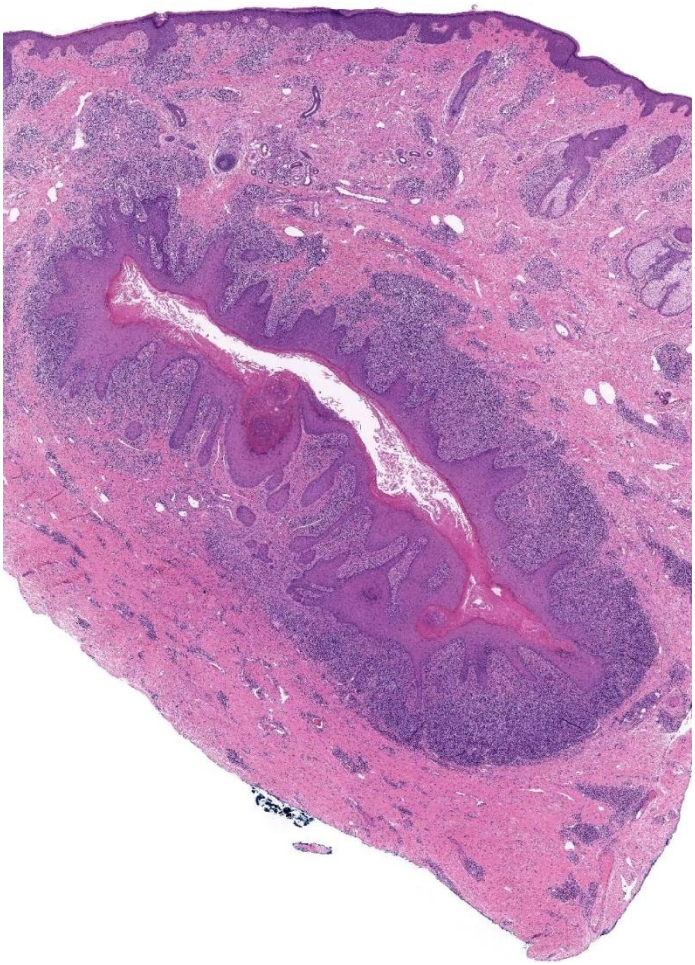

Figure 2. *Dense lichenoid lymphocytic infiltrate with scattered eosinophils centered around cystically dilated follicular epithelium with irregular acanthosis and hypergranulosis. H&E, 20×.*

Table 1. *Clinical and histologic features helpful in distinguishing lichen planus follicularis tumidus from other entities.*

	Clinical features	Histologic features
Lichen planus follicularis tumidus [1-8]	Middle age onset, Female > Male Red-violet plaque containing white-yellow cysts and comedones Predilection for retroauricular area, no extrafacial involvement Possible concurrent lichen planus lesions on extremities and mucosal surfaces	Keratin-filled follicular cysts Cyst wall exhibits epithelial hyperplasia with lichen planus features of hypergranulosis, jagged rete ridges Dense lichenoid perifollicular lymphocytic infiltrate
Milia en plaque [8, 9]	Middle age onset, Female > male Erythematous plaque containing white-yellow cysts Predilection for periauricular area, extrafacial involvement	Keratin-filled follicular cysts Sparse-moderate perifollicular lymphocytic infiltrate
Steatocystoma multiplex [10]	Adolescent-early adult onset, Female = male Firm, flesh-colored-yellow papules or nodules Autosomal dominant familial inheritance Commonly involves face, trunk, and extremities	Empty cysts with crenulated eosinophilic cyst lining Sebaceous lobules inside or attached to cyst wall
Nevus comedonicus [11]	Congenital-childhood onset, Female = male Grouped, dilated follicular openings containing black keratinaceous material Commonly involves face and neck Possible associated skeletal, ocular, central nervous system abnormalities	Dilated follicular ostia with lamellated keratin Absence of hair shafts
Favre-Racouchot syndrome [12]	Elderly onset, Male > Female Open and closed comedones on sun-damaged skin Chronic sun exposure, heavy smoking Predilection for sun-exposed periorbital, temporal, and neck areas	Keratin-filled cysts and dilated, plugged follicular infundibula Non-inflammatory background of marked solar elastosis
Folliculotropic mycosis fungoides [13]	Middle age-elderly onset, Male > Female Erythematous papules and plaques with follicular prominence, alopecia, comedones, cysts, or nodules Predilection for head and neck areas	Enlarged follicular infundibula with atypical lymphocytes exhibiting epitheliotropism Pautrier's microabscesses
Discoid lupus erythematosus [14]	Adolescent-adult onset, Female > Male Erythematous plaques with scale and follicular prominence, scarring alopecia Photosensitivity Predilection for sun-exposed face and neck, scalp areas, extrafacial involvement	Interface dermatitis with perivascular and periadnexal lymphocytic infiltrate Plugged follicles, increased dermal interstitial mucin, thickened basement membrane
Follicular mucinosis [13, 14]	Adult versus childhood onset, Female = Male Follicular papules within erythematous patches or plaques, associated alopecia (alopecia mucinosa), mucinous discharge Predilection for head and neck, extrafacial involvement Possible coexisting folliculotropic mycosis fungoides or discoid lupus erythematous in adult subset versus usually benign course in pediatric subset	Dilated follicular infundibula and cysts with marked mucin deposition Perifollicular lymphohistiocytic infiltrate with eosinophils

immunoglobulin deposition along the basement membrane (lupus band) or T cell receptor gene rearrangement analysis may be performed if there is high clinical suspicion for DLE or folliculotropic MF, respectively. These ancillary studies were not indicated in our work-up given the classic LPFT presentation, both clinically and histologically. Lastly, follicular mucinosis, whether primary or secondary to MF or DLE would feature follicular mucin deposition, which was absent in our case [13, 14]. Of note, this dark-complected patient reported a prior history of SCC of the left retroauricular area; one could speculate whether pseudocarcinomatous epithelial hyperplasia or SCC superimposed upon LPFT might have been present [15]. The differential diagnosis for LPFT is summarized in **Table 1**.

Although spontaneous resolution of lesions has been documented, LPFT is predominantly a chronic, relapsing condition that is resistant to treatment [5]. Recurrence occurs frequently following surgical excision, as in our patient's case. Reported therapies include high potency corticosteroids, prednisone, isotretinoin, and cyclosporine for severe disease [3, 4].

Conclusion

LPFT is a rare variant of LP with distinctive clinical and histologic findings. Familiarity with this rare variant may facilitate earlier diagnosis and screening for LP involving other sites as well as associated systemic disorders such as viral hepatitis and autoimmune thyroiditis.

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