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Lichen planus pigmentosus and lichen planopilaris

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Abstract

Lichen planus is an inflammatory skin condition with characteristic histopathological findings that has many clinical variants. Recently, there have been 25 cases reported in the literature of lichen planus pigmentosus (LPPi) that preceded the development of frontal fibrosing alopecia (FFA), which is a variant of lichen planopilaris (LPP). We review the literature regarding LPPi, LPP, and FFA and present a case of a 40-year-old African American woman with LPPi and LPP.

Case Presentation

PATIENT: 40-year-old woman

DURATION: Three years

DISTRIBUTION: Scalp, face, neck, and upper extremities

HISTORY: A 40-year-old African American woman who presented to Center for Women's Health for a three-year history of dark pigmentation, which started on her arms and spread to face and neck. She denied a history of skin conditions. She stated that the condition was mildly pruritic at times. She had tried hydroquinone and azelaic acid without improvement. She also reported a history of hair loss over the last few years although she could not recall the exact time frame when it began. The hair loss predominantly affected her frontal scalp and a few areas of her occipital scalp. She did report having permanent waves and extensions in the past but denied braids or dreadlocks. She also reported losing hair from her eyebrows and upper extremities. Past medical history included anemia, prolactinoma, and fibroids.



Figure 1. Neck with ill-defined, dark-brown-to-grey macules and patches with no epidermal changes.



Figure 2. Frontal scalp with recessed hairline with notable follicular dropout and few areas of follicular hyperpigmentation.



Figure 3. Occipital scalp with localized patch of decreased hair density with notable follicular dropout.

PHYSICAL EXAMINATION: On the upper extremities, neck, and face were ill-defined, dark-brown-to-grey macules and patches with no epidermal changes (**Figure 1**). The frontal scalp had a recessed hairline with notable follicular dropout and few areas of follicular hyperpigmentation. (**Figure 2**). The occipital scalp also had a localized patch of decreased hair density with notable follicular dropout (**Figure 3**).

LABORATORY DATA: Prolactin was 53.3 ng/mL. Adrenocorticotrophic hormone was normal. Antinuclear antibody and anti-topoisomerase 1, anti-Ro/SSA, anti-La/SSB, and anticentromere antibodies were negative.

HISTOPATHOLOGY: There is a decreased number of terminal anagen follicles from the scalp. There is a sparse, perifollicular, lymphocytic infiltrate. Perifollicular fibrosis with clefting between the stroma and follicular epithelium is evident (**Figure 4**).

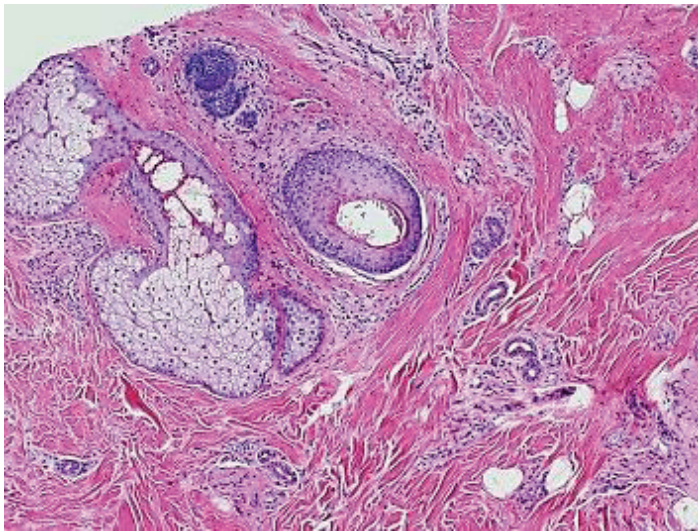


Figure 4. Biopsy from scalp showing decreased number of terminal anagen follicles and sparse, perifollicular, lymphocytic infiltrate. Perifollicular fibrosis with clefting between the stroma and follicular epithelium is evident.

A biopsy specimen from the neck shows a superficial, perivascular, lymphocytic infiltrate with numerous melanophages within the papillary dermis (**Figure 5**). Melan-A and PNL2 immunostains highlight the normal number and distribution of basal melanocytes.

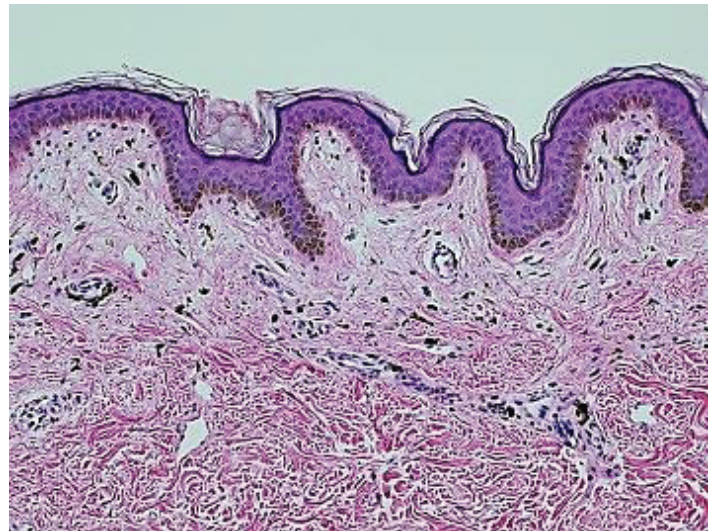


Figure 5. Biopsy from neck showing a superficial, perivascular, lymphocytic infiltrate with numerous melanophages within the papillary dermis. Melan-A and PNL2 immunostains highlight the normal number and distribution of basal melanocytes.

DIAGNOSIS: Lichen planus pigmentosus and lichen planopilaris

Discussion

Lichen planus (LP) is an inflammatory skin condition with characteristic clinical and histopathological findings that has many clinical variants. We shall focus on two such variants and their recently described association.

Lichen planus pigmentosus (LPPi) is an uncommon variant of LP, which was first described in a series of Indian patients in 1974 [1]. While it can affect all races, it has a predilection for darker-skinned individuals. The incidence and etiology remain unknown although some have proposed that viral infections, particularly hepatitis C virus, states of impaired carbohydrate metabolism; drugs; topical agents, such as mustard oil, alma oil, henna dye; and sun exposure are possible triggering factors [1-4]. LPPi typically affects middle-aged adults and presents with dark-brown-to-slate-gray macules and patches that are symmetrically distributed in sun-exposed areas. It commonly affects the head and neck [2, 5]. The pigmentation pattern is usually diffuse, but reticular, blotchy, unilateral linear, and perifollicular patterns have been described [6]. Unlike classic LP, the lesions of LPPi tend to be asymptomatic although pruritus and a burning sensation have been reported [3]. LPPi may have

a protracted clinical course. Treatments that have been reported with varying success are: topical and oral glucocorticoids, tacrolimus ointment, acitretin, skin-lightening creams, and neodymium:yttrium-aluminum-garnet laser [2, 5, 7-9]. Patients should practice strict photoprotection. The histopathologic features of LPPi are epidermal atrophy, vacuolar degeneration of the basal layer, perivascular and a band-like infiltrate in the upper dermis although it can be sparse, and dermal melanophages [3,9]. Melanin incontinence may be the only feature in older lesions.

Lichen planopilaris [LPP] is a variant of LP that also can be classified as a primary lymphocytic cicatricial alopecia [10]. There is a predilection for Caucasian and Indian populations, and it has a female predominance, with a female-to-male ratio of 1.8:1 [2]. It more commonly occurs in adults but has been reported in children. The exact etiology remains unknown, but it may be related to an inflammatory reaction that is mediated by T-lymphocytes, which are directed at follicular antigens. It typically affects the vertex and parietal scalp and presents with white, atrophic or scarred patches of alopecia. Perifollicular erythema and scale, keratotic follicular papules, and loss of follicular ostia commonly are observed [10-11]. Patients frequently report itching, burning, scaling, tenderness, and increased shedding.

Frontal fibrosing alopecia (FFA) is a subtype of LPP that affects the frontal hairline and leads to a symmetric, progressive, band-like hair loss, and scars of the frontotemporal scalp. It predominantly affects postmenopausal women. In addition to hair loss from the scalp, 52% of FFA patients also have eyebrow hair loss, which can occur both before and after frontotemporal recession [7, 12-13]. LPP and FFA are indistinguishable on histopathologic examination. However, some argue that findings of FFA are less severe [14]. Histopathologic features of an active lesion are a lichenoid, lymphocytic infiltrate that affect the isthmus and infundibulum, with relative sparing of the lower portion of the hair follicle. There is loss of sebaceous glands and arrector pili muscles with perifollicular lamellar fibrosis. With disease progression, the follicular unit will be completely destroyed and replaced by thick

fibrous tracts [10-11].

The incidence of cutaneous or mucosal LP with LPP and FFA is reported to be 17-28% and 8%, respectively [7, 10, 15]. Recently, there has been an association described between LPPi and FFA. In 2014, the first case report of LPPi with FFA, was described in a middle-aged, post menopausal Indian woman. Since then, a case series of 24 patients in South Africa, who presented with LPPi that preceded FFA, has been published. The majority of the patients were pre-menopausal (64%), African (91%), and women (95%). The lag time from LPPi to FFA was 14 months, with a range of six to 36 months [16]. These 25 cases show that LPPi perhaps can serve as a harbinger of FFA, particularly in African patients. Thus for these patients, who present with LPPi, it may be useful to evaluate them for hair loss and follow them more closely.

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