Lichen Planopilaris Developing at the Margins of a Sebaceous Nevus of Jadassohn: Co-existence or Etiologic Association?

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Abstract

Sebaceous nevus (SN), is a skin hamartoma, combining a variety of epidermal, follicular, sebaceous, and apocrine abnormalities. Although usually present at birth, it may become apparent later in life appearing as a yellowish-brown, verrucous plaque with alopecia. SN is implicated with secondary tumors arising on the hairless plaque during the adulthood. Lichen planopilaris is a common, primary lymphocytic scarring alopecia of unknown etiology, characterized by lichenoid/interface perifollicular lymphocytic infiltrate occurring at the level of the infundibulum and the isthmus. Here, we present a case of a 48-year-old Caucasian male with lichen planopilaris lesions developing at the periphery of a preexisting SN. Our case raises the question, whether the development of lichen planopilaris was coincidental, or is it indicative of an etiologic association between the lichen planopilaris and SN.

Key Words: Alopecia, dermoscopy, epidermal nevus, lichen planopilaris, pathogenesis, sebaceous nevus

Sebaceous nevus (SN) is a skin hamartoma combining a variety of epidermal, follicular, sebaceous, and apocrine abnormalities.\(^1\) A prevalence of 0.1%–0.3% has been reported for SN among newborns; 0.68% among biopsy specimens; and 0.5%–1% among dermatologic patients.\(^2,3\) Although usually present at birth, it may become apparent later in life. Generally, it is located on the scalp, appearing as yellowish-brown, waxy, and hairless plaque. During puberty, the lesion becomes thickened and verrucous. During adulthood, secondary tumors, benign and malignant, may develop. Occasionally, SN, especially when extensive, may be associated with the central nervous system, skeletal, optical, or cardiovascular abnormalities in the context of SN syndrome.\(^4\)

A 48-year-old Caucasian male was referred to us for an alopecic plaque of the scalp dating back to childhood. The lesion had increased in size and had become symptomatic a few months before. The patient was in good general health and his personal and family history was unremarkable. On clinical examination, a yellowish, moderately elevated, oval, hairless plaque, 14 mm in diameter, located on the left crown area of the scalp, was observed. At the periphery of the plaque, there were white or purple-red eruptions [Figure 1a]. The patient reported that the lesion had recently became intensely itchy, forcing him to scratch. The rest of the scalp was normal. Skin and mucous membrane examinations were negative. Dermoscopy revealed a central yellow-orange, elevated, verrucous area. At the periphery, there were sites of white atrophic skin with the absence of follicular openings, perifollicular scale, and purple-red perifollicular erythema [Figure 1b]. Two biopsy specimens one from the central part of the lesion and one from the periphery were obtained by 3-mm punch. Histologic examination of the central part showed the typical features of SN. In the peripheral part, mild perifollicular fibrosis and a lymphocytic inflammatory infiltrate involving the upper hair follicle, were observed [Figure 2]. Based on the clinical, dermoscopic, and histological grounds, the diagnosis of lichen planopilaris (LPP) developing at the margins of a preexisting SN was made.
SN is a well-recognized cause of alopecia. In a retrospective study of 345 cases of childhood alopecia, SN was the most common cause (13.2%) in newborns, together with aplasia cutis.\(^5\) In the present patient, SN showed the signs of peripheral expansion that histologically exhibited features of LPP. To the best of our knowledge, this is the first-reported case of LPP-associated with SN. Literature review revealed a single case of LPP-like changes arising within an epidermal nevus.\(^6\) In addition, a linear inflammatory variant of epidermal nevus referred to as inflammatory linear verrucous epidermal nevus (ILVEN), does exist, exhibiting histopathologic similarities to psoriasis.\(^7\) Some authors suggest that these two conditions share a common pathogenesis. There is evidence that interleukins 1 and 6, tumor necrosis factor, and intercellular adhesion molecule-1 are upregulated in ILVEN, similar to psoriasis.\(^8\)

LPP is a common, primary lymphocytic scarring alopecia, characterized by the lichenoid/interface perifollicular lymphocytic infiltrate, occurring at the level of the infundibulum and the isthmus.\(^9\) Although its etiopathogenesis is still unclear, there is evidence that a T-cell mediated immunologic reaction to a yet unknown antigenic stimulus may underlie LPP.\(^10\)

Our case raises the question of whether the development of LPP was coincidental, or it is indicative of an etiologic association between LPP and SN. The strict localization of LPP at the margin of the nevus without evidence of a more widespread involvement, suggests that local factors related to the SN may play an important role. The example of ILVEN also supports the notion that epidermal nevi may harbor immunologic inflammatory reactions. These reactions may be of the psoriasiform type, as in the case of ILVEN, or possibly of the lichenoid type, as in this case.

**Declaration of patient consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**
There are no conflicts of interest.

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