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Scalp porocarcinoma and lichen planopilaris

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Abstract

Porocarcinoma (PC) is a malignant neoplasm arising from the intraepidermal ductal portion of the sweat gland duct. Lichen planopilaris (LPP) is a not so rare variant of cutaneous lichen planus (LP) with a preferential involvement of hair follicles, consisting of a chronic lymphocytic inflammation, leading to cicatricial alopecia. A 42-year-old woman, recently diagnosed with HIV infection, was referred to our clinic because of an alopecic patch of 6 years' duration. In the upper region of the alopecia a 1.5cm nodule was noticed, which the patient stated had started growing soon after the appearance of the hair loss. Biopsy of the alopecia margin confirmed the diagnosis of LPP, whereas biopsy of the nodule revealed an infiltrating tumor consistent with the diagnosis of PC. We present a scalp PC emerging in a background of LPP in an HIV patient. We do not know the role, if any, HIV infection and LPP played in this particular case. Immunosuppression and HIV have been implicated in the etiology of PC. However, her HIV diagnosis was made after the appearance of the scalp nodule. We did not find any association between LPP and PC in the literature. Even though an association by chance cannot be excluded, this deserves further investigation.

Keywords: porocarcinoma, lichen planopilaris, HIV

Introduction

Porocarcinoma (PC) is a malignant neoplasm arising from the intraepidermal ductal portion of the eccrine sweat duct glands, representing 0.005% to 0.01% of all malignant epithelial neoplasms [1]. LPP is a not-so-rare variant of cutaneous lichen planus (LP) with a preferential involvement of hair follicles, consisting of a chronic lymphocytic inflammation, leading to cicatricial alopecia [2]. It is the most frequent cause of adult primary scarring alopecia. We present an HIV positive patient with scalp PC emerging in a background of LPP.

Case Synopsis

A 42-year-old woman was referred to our clinic because of pruritus, erythema, and scales in the vertex of the scalp for the past 6 years, with progressive development of an alopecic patch. In the upper left region of the hair loss a 1.5cm nodule was

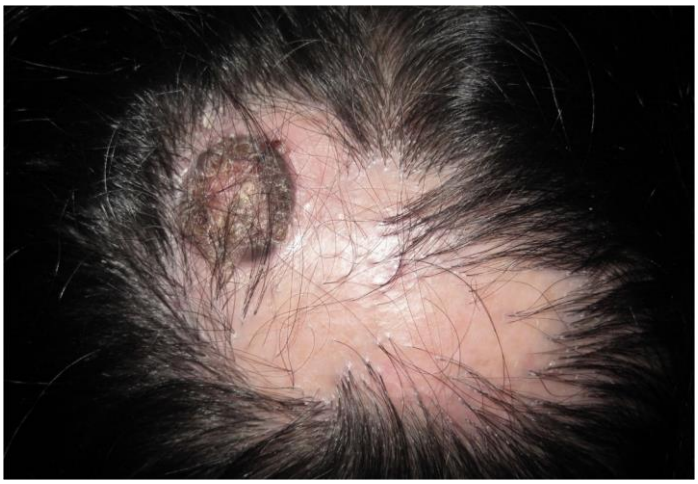

Figure 1. *Atrophic skin with no follicular ostia surrounded by hair-bearing scalp with mild erythema and scales, in the context of LPP. In the upper left region of the pseudopelade a 1.5cm hyperkeratotic nodule is seen.*

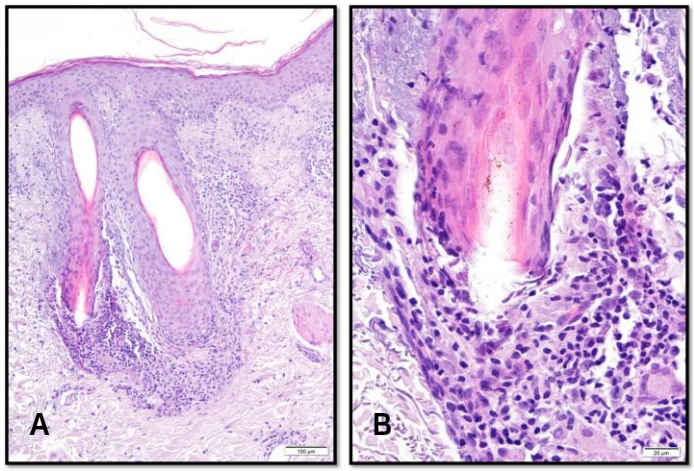

Figure 2. ***A)*** *Epidermis with minimal orthoqueratotic hyperkeratosis and dermis with moderate lymphocytic inflammatory infiltrate surrounding hair follicles.* ***B)*** *The follicular epithelium shows exocytosis of lymphocytes associated with vacuolar degeneration and apoptosis of basal keratinocytes, 40×. Direct immunofluorescence essays showed no immunoreactivity for IgM, IgG, Ig A, C3c, C1q or fibrinogen excluding the possibility of lupus.*

noticed (**Figure 1**) The patient stated it had started growing soon after the alopecia appearance. No other relevant mucocutaneous lesions were noticed. HIV infection had been diagnosed one year before presentation but was currently without treatment as she refused it. Her medical history was also positive for previous hepatitis B virus (HBV) infection, latent undetermined syphilis, and dyslipidemia. Biopsy of the alopecia margin confirmed the clinical diagnosis of lichen planopilaris (LPP), (**Figure 2**). Another biopsy was performed in the nodule revealing an infiltrating tumor composed of pleomorphic cuboidal cells with nuclear hyperchromasia and significant mitotic activity. The tumor cells were arranged in broad columns that extended from the lower epidermis into the dermis. Large irregularly shaped/sized ductal spaces were also present, consistent with the diagnosis of PC with apocrine differentiation (**Figure 3**). The nodule was fully excised and the patient started treatment with hydroxychloroquine 400 mg/day and pioglitzone 15 mg/day as well as topical betametasone solution every other day to the erythematous, scaly skin. The patient stopped oral treatment on her own one year later. The hair loss has been stable since the LPP diagnosis and the patient is still tumor free after 4 years of follow-up (**Figure 4**).

Case Discussion

Although rare, PC is the most commonly encountered malignant skin tumor of sweat gland origin [3]. Porocarcinomas occur more often in the

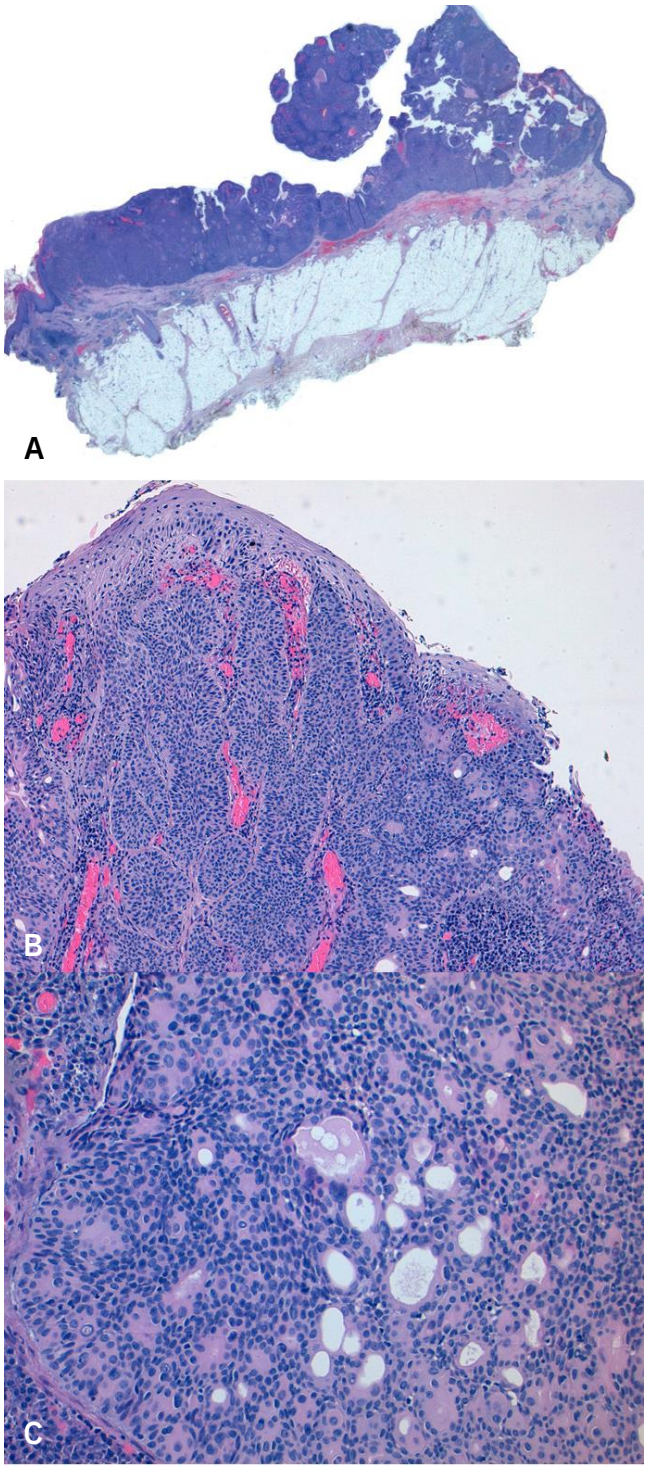

Figure 3. *An infiltrating tumor composed of pleomorphic cuboidal cells with nuclear hyperchromasia and important mitotic activity consistent with the diagnosis of porocarcinoma (a = 5x; b= 20x; c = 40x)*

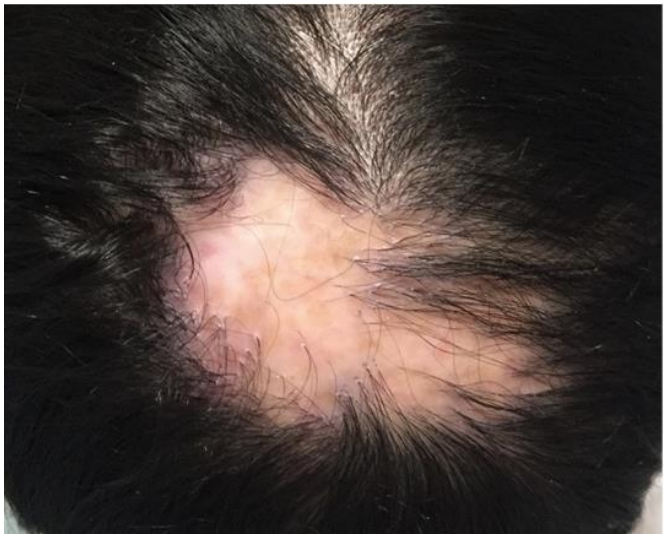

Figure 4. *Patient after 4 years of follow-up - pseudopelade is stable since LPP diagnosis and patient is tumor free.*

sixth to eighth decades of life and have been reported slightly more often in females [4]. Pathogenesis is still unknown, but it has been associated with burns, trauma, radiotherapy, prolonged exposure to ultraviolet radiation, immunosuppression, HIV, sarcoidosis, and malignant diseases [5]. Sometimes, there is a history of long duration and this may be explained by the fact that some PC arise from its benign counterpart, eccrine or apocrine poroma. Additionally, PC has been reported as arising in pre-existing seborrheic keratoses, nevus sebaceous, and Bowen disease [5]. PC has a great propensity to act aggressively. About 20% of PC will recur and the estimated prevalence of regional lymph node metastases ranges from 19% to 30% [1, 6]. Surgical resection is the mainstay of treatment [5].

The etiology of LPP is unknown, despite a suspected autoimmune origin mediated by T lymphocytes targeting follicular antigens. The process may be triggered by drugs, contact allergens, and virus or other infectious agents in predisposed subjects [2]. In this case, the patient's history of HBV infection or even syphilis could have played a role. Furthermore, the HIV associated immunosuppression could have contributed to the LPP stability. Perifollicular erythema and scaling are commonly the first clinical findings of LPP. These signs are then replaced by atrophic scarring related to inflammation [7, 8]. The classical form of LPP most commonly involves the vertex, but disease may also be present on the other hair-bearing sites [8]. An association with other forms of LP can be found in about 25% of LPP patients [8]. Thyroid disorders and other autoimmune diseases may also be seen in association with LPP [7, 9]. Additionally, squamous cell carcinoma can develop in longstanding LPP lesions [2].

Conclusion

We present a scalp PC emerging in a background of LPP in an HIV patient. We speculate that PC in this case could have arisen in a pre-existing lesion, explaining the 6 years' evolution. We do not know the role, if any, HIV infection and LPP played in this particular case. Immunosuppression and HIV have been implicated in the PC pathogenesis. However, the HIV diagnosis was made after the scalp nodule appeared.

We did not find any association between LPP and PC in the literature. Even though an association by chance cannot be excluded, this deserves further investigation.

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