Squamous cell carcinoma originating in folliculitis decalvans

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Abstract

Folliculitis decalvans is a chronic and progressive scarring alopecia at the vertex and occipital scalp with a predilection for middle-aged men. Squamous cell carcinoma is an exceedingly rare complication of folliculitis decalvans, reported in 5 cases to date. Herein, we present a case of squamous cell carcinoma of the scalp in a patient diagnosed with recalcitrant folliculitis decalvans and review the clinicopathologic characteristics of all reported cases in the literature.

Keywords: folliculitis decalvans, scarring alopecia, squamous cell

Introduction

Folliculitis decalvans is a chronic scarring alopecia characterized by neutrophilic inflammation. Clinically, folliculitis decalvans presents as follicular pustules, perifollicular erythema, and loss of follicular ostia, most commonly at the vertex and occipital scalp regions. Therapy with oral antibiotics is aimed at reducing inflammation and progressive hair loss. We report an immunosuppressed patient with cutaneous squamous cell carcinoma of the scalp and a history of folliculitis decalvans.

Case Synopsis

A 38-year-old man with a history of renal transplant and immunosuppression presented with painful lesions of the scalp that had been present for two years. He had been previously diagnosed with folliculitis decalvans, which had not responded to doxycycline, amoxicillin, or topical therapies, including corticosteroids and antibiotics.

The scalp exam revealed focal bogginess of the vertex scalp as well as a pink, shiny atrophy plaque with alopecia, few papulopustules, and one ulceration on the right vertex scalp (**Figure 1**). Initial biopsy demonstrated cicatricial folliculitis with Bowenoid dysplasia of the hair follicle epithelium (**Figure 2A**). Three repeat biopsies were performed and histopathologic assessment of two of the lesions was consistent with squamous cell carcinoma (**Figure 2B**). Histology of the third lesion demonstrated findings consistent with proliferative actinic keratosis. The sites of repeat biopsies are shown in (**Figure 3**).

He was treated with Mohs micrographic surgery with a final defect of 7.7cm x 4.8cm. There was no clinical sign of recurrence at follow-up one month later.

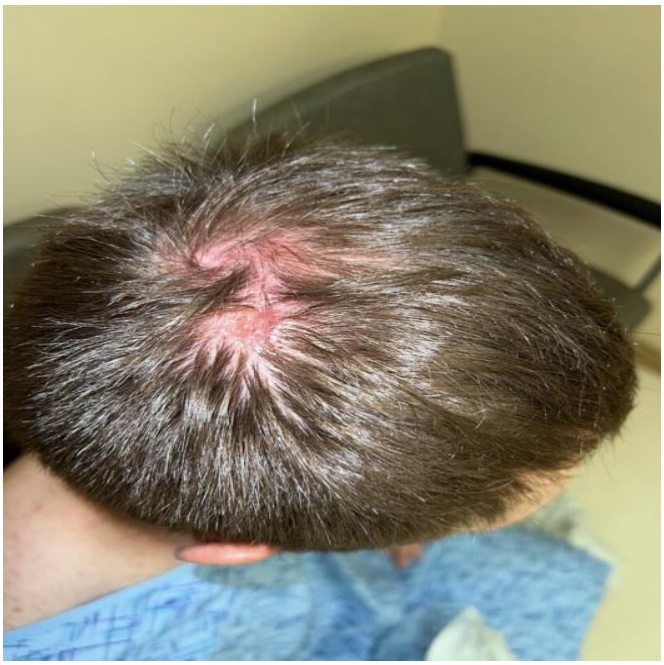

Figure 1. *Squamous cell carcinoma of the scalp. Focal bogginess of the vertex scalp. Papulopustules and one ulceration of the right vertex scalp within a pink, shiny, alopecic plaque.*

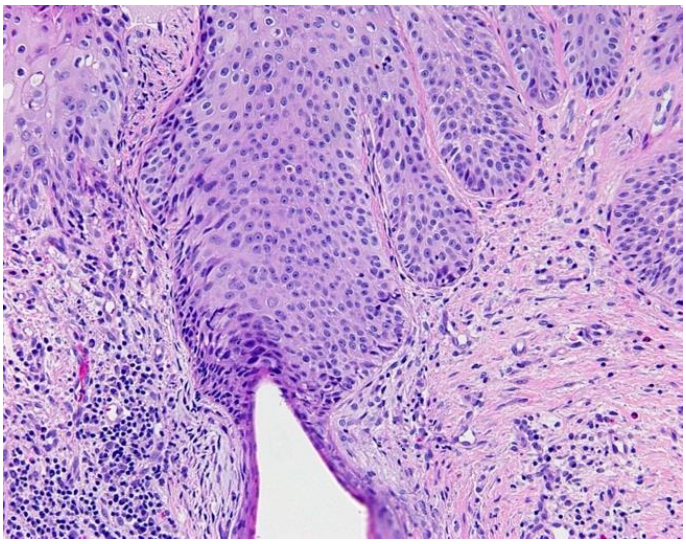

Figure 2A. *Histopathological findings. Initial biopsy demonstrating Bowenoid dysplasia of the follicular infundibulum with perifollicular desmoplasia and lymphoplasmacytic inflammation.*

Case Discussion

Chronic folliculitis decalvans presents clinically as tufted folliculitis along with follicular papules, pustules, and scarring [1]. Histopathology examination of folliculitis decalvans demonstrates a neutrophil-predominant infiltrate within the follicular infundibula expanding into the dermis as the disease progresses [1,2]. Older lesions contain mixed inflammatory infiltrate with giant cells enclosing hair bulbs and dermal fibrosis [2]. The first-line therapy for folliculitis decalvans is oral tetracyclines, aimed at decreasing the inflammatory cascade [1].

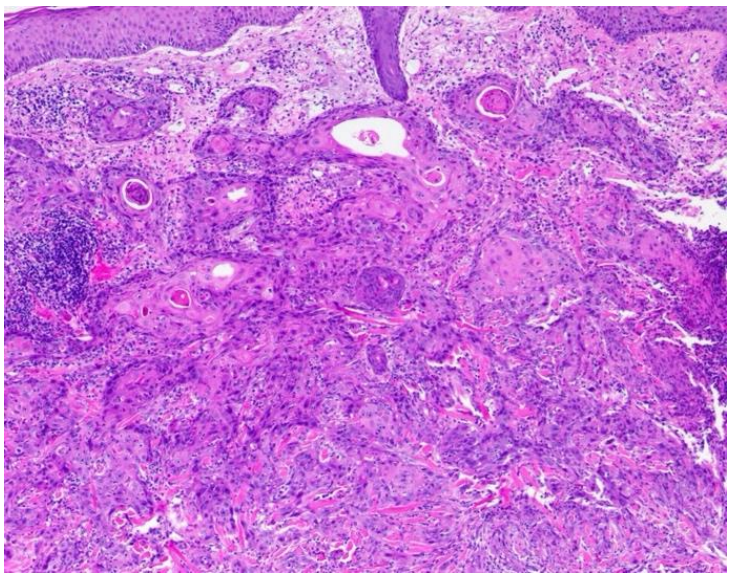

Figure 2B. *Second biopsy demonstrating squamous cell carcinoma.*

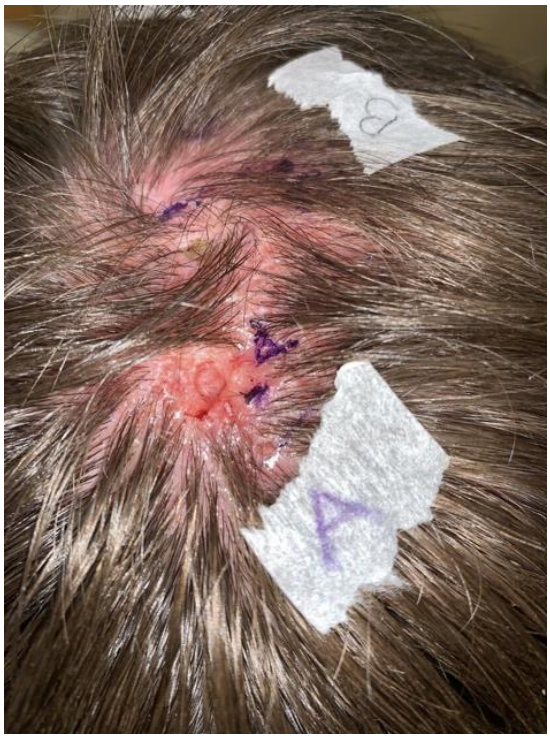

Figure 3. *Repeat of biopsy sites.*

Although cutaneous squamous cell carcinomas typically arise at sites susceptible to ultraviolet radiation-induced deoxyribonucleic acid damage, squamous cell carcinoma carcinogenesis may also occur in the setting of chronic inflammation, such as

in the case of Marjolin ulcers [3]. This is typically associated with burn injuries but has also been reported as a complication of chronic inflammatory dermatologic conditions such as hidradenitis suppurativa [3]. Cutaneous squamous cell carcinoma arising in the setting of folliculitis decalvans is rare, with a total of 6 reported cases (including the current case) to date (**Table 1**) [4,5,6,7,8]. All cases occurred in men at median age 49 years (range, 38-66 years) with long-standing, refractory folliculitis decalvans (range, 8-26 years) of the scalp [4,5,6,7,8]. None of the previously reported cases occurred in immunosuppressed patients. Two patients underwent oral isotretinoin therapy within two months prior to metastatic squamous cell carcinoma diagnosis [4,7]. In all cases, treatment involved surgical excision. Metastatic cases (three out of 6) were additionally treated with adjuvant radiation, chemotherapy with cemiplimab, and/or lymph node dissection [4,5].

Notably, our patient's transplant history and subsequent immunosuppression posed a significant, well-known risk factor for squamous cell carcinoma and was likely to have been the main contributor to squamous cell carcinoma carcinogenesis. Additionally, the chronic inflammatory state in the setting of folliculitis decalvans may have contributed to squamous cell carcinoma development.

Squamous cell carcinoma arising in the setting of chronic inflammation has a worse prognosis compared to squamous cell carcinoma associated with ultraviolet exposure, with metastasis in up to 40% and mortality in 21% of cases in the former [3,9]. Metastasis to the neck, cervical, and occipital lymph nodes occurred in 50% of squamous cell carcinoma associated with folliculitis decalvans [4,5,8]. Squamous cell carcinoma related to immunosuppression also has a higher risk of metastasis and mortality [10]. Squamous cell carcinoma arising in the combined setting of chronic inflammation related to folliculitis decalvans and immunosuppression may have an even higher risk of poor outcomes, though to our knowledge, no studies have demonstrated this.

Conclusion

The current case highlights the possibility of squamous cell carcinoma arising from sites affected by folliculitis decalvans in the setting of immunosuppression. Given the limited evidence indicating a high rate of metastasis in squamous cell carcinoma associated with folliculitis decalvans, surveillance of patients with folliculitis decalvans for changing skin lesions at the affected skin sites is warranted to ensure early diagnosis and prompt therapy.

Potential conflicts of interest

The authors declare no conflicts of interest.

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Table 1. *Summary of demographics, clinical presentation, histologic examination, treatment, and outcome of squamous cell carcinoma associated with folliculitis decalvans.*

Case	Age (Years)	Gender	Latency (Years)	Race/ Ethnicity	Clinical Presentation	Comorbidities	Immuno-suppressed	Histopathology	Treatment	Outcome	References
1	38	Male	2	Caucasian	Focal bogginess of the vertex scalp. Pink, shiny atrophic plaque with alopecia, few papulopustules, and one ulceration on the right vertex scalp.	None	Renal transplant, immunosuppressants (tacrolimus)	Squamous cell carcinoma, hair fiber granuloma, infundibular/isthmus hourglass desmoplasia. Perifollicular and perivascular plasmocytic inflammation.	Excision	No recurrence of metastasis at 3 months	CR = current report
2	40	Male	"Long-standing"	African-American	10 cm exophytic, verrucous-like mass with cutaneous horns	None	No	Squamous cell carcinoma, dermal scarring, mixed inflammation, follicular plugging and rupture, extravasated hair shafts	Excision	Metastasis to occipital lymph nodes	[8]
3	44	Male	8	N/A	8 mm rapidly enlarging nodule within area of scalp alopecia	Basal cell carcinoma	No	Squamous cell carcinoma infiltrating into the reticular dermis. Dermal fibrosis with heavy lymphoplasmacytic inflammation.	Excision	N/A	[6]

4	45	Male	"Long-standing"	N/A	Pink plaques and scattered tufts of hairs with 2.5 cm pink and boggy, exophytic nodule draining serosanguineous material	Former smoker	No	Squamous cell carcinoma	Excision and bilateral neck lymph node dissection	Metastasis to four right posterior neck lymph nodes	[5]
5	52	Male	>20	Hispanic	Scarred pink plaques with hemorrhagic crusting and tufting of hairs	Sebaceous carcinoma	No	Endophytic squamous cell carcinoma with basilar atypia and neutrophilic aggregates. Lichenoid inflammation, dermal fibrosis, foreign body giant cells, and pseudoepitheliomatous hyperplasia.	Excision, adjuvant radiation, cemiplimab	Metastasis to cervical lymph nodes. Dramatic reduction with cemiplimab.	[4]
6	66	Male	26	Caucasian	4 cm ulceration with erythematous, scaly edge and hemorrhagic crust and 3 cm nodules with cutaneous horn	N/A	N/A	Squamous cell carcinoma dense fibrosis, lymphocytic infiltrate with suppurative foci	Excision	N/A	[7]