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POROKERATOSIS OF THE FACE: REVISITING THE UNCOMMON IN DISSEMINATED SUPERFICIAL ACTINIC POROKERATOSIS

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Abstract
Porokeratosis is a rare disorder of keratinization characterized by annular lesions with raised keratotic borders and cornoid lamellae on histopathology. Disseminated Superficial Actinic Porokeratosis (DSAP) is its most common variant; however, facial involvement is uncommon and may be misdiagnosed.We report a case of a 32-year-old male presenting with a six-month history of hyperpigmented lesions on the face and neck, associated with chronic sun exposure and inadequate photoprotection. Clinical evaluation, dermoscopy, and histopathology confirmed DSAP. Treatment with topical retinoids and calcipotriol, combined with strict sun protection and daily moisturization, led to marked clinical improvement.This case highlights the importance of recognizing DSAP in atypical sites and the need for individualized treatment strategies. Early diagnosis and optimal management are crucial in preventing progression and improving patient outcomes.
Keywords- DSAP, cornoid lamellae, dermoscopy,

INTRODUCTION

Porokeratosis comprises a group of disorders of keratinization, characterized by annular macules or plaques with hyperkeratotic borders and atrophic centres. Disseminated Superficial Actinic Porokeratosis is the most frequently observed variant, which affects sun-exposed areas such as the forearms and legs [1,2]. Facial involvement is particularly rare and may mimic other dermatoses like actinic keratosis, complicating diagnosis[2]. DSAP usually presents in middle-age with cases reported from adolescence to late adulthood, and shows a slight female predominance [3,4]. The pathogenesis of DSAP involves genetic mutations and environmental triggers, particularly chronic ultraviolet (UV) exposure and immunosuppression [5]. While DSAP is generally benign, there is a minimal risk of progression to malignancy, most notably into squamous cell carcinoma, highlighting the need for precise diagnosis and appropriate management[6]. Management of DSAP, particularly with facial involvement, includes photoprotection, topical agents such as imiquimod, retinoids, and 5-fluorouracil, alongside procedural options like cryotherapy, photodynamic therapy, and laser treatments [7,8]. Fractional CO_2 laser therapy has also been shown to be effective, particularly in facial DSAP, with a reduced risk of scarring [9]. Oral retinoids are also effective in extensive or refractory cases[10]. Emerging approaches like chemical peels and combination therapies (e.g., tretinoin with calcipotriol) show promising results but require careful consideration to balance efficacy and cosmetic outcomes[7,11]. This article highlights the rare presentation of Disseminated Superficial Actinic Porokeratosis and the importance of patient education in achieving optimal outcomes.

Case Report

A 33-year-old male came to the outpatient department with a six-month history of multiple hyperpigmented lesions predominantly affecting the face and neck. Initially confined to the neck, the lesions gradually spread to the face over time. The patient reported no associated symptoms such as itching, pain, or burning. He denied any history of similar dermatological conditions or symptoms such as fever, weight loss, or photosensitivity. The patient's medical history was unremarkable, with no chronic illnesses, malignancies, or prior use of

immunosuppressive therapies. Additionally, there was no significant family history of similar conditions. Lifestyle factors revealed frequent sun exposure due to outdoor activities and an absence of regular sunscreen use, indicating insufficient photoprotection. On physical examination, there were multiple well defined hyperpigmented to brown macules with pale centre, surrounded by well-demarcated hyperkeratotic borders. There were no signs of erythema, inflammation, scaling, or crusting. Dermoscopy revealed a well-defined annular areas with a prominent white track along the hyperkeratotic borders and a central zone of brown pigmentation, indicative of atrophic skin (Figure 4). Biopsy from a lesion on the face showed the hallmark feature of porokeratosis, the cornoid lamella (Figure 5). Considering the patient's history, clinical features, dermoscopic findings, and histopathology, a diagnosis of Disseminated Superficial Actinic Porokeratosis (DSAP) of the face was made. The patient was treated with tretinoin 0.025% cream and calcipotriol 0.005% ointment, to be used on alternate nights, along with daily moisturization and strict sunscreen use. This treatment was well-tolerated and led to noticeable improvement in the appearance of the lesions over time.

DISCUSSION

Porokeratosis is a rare group of keratinization disorders presenting as annular lesions with raised, hyperkeratotic borders and an atrophic centre. The major types include porokeratosis of Mibelli, linear porokeratosis, punctate porokeratosis and Disseminated Superficial Actinic Porokeratosis (DSAP)[1]. DSAP is the most common subtype, predominantly affecting sun-exposed parts of the body such as the forearms and legs, with facial involvement being rare [2]. Facial involvement is reported in less than 15% of cases, complicating diagnosis as lesions may resemble other dermatoses such as actinic keratosis or seborrheic keratosis [2]. DSAP typically presents as reddish-brown macules or patches with atrophic centres and elevated keratotic borders [3]. A definitive diagnosis relies on histopathological confirmation, with the hallmark feature being the **cornoid lamella**, a thin column of parakeratotic cells overlying a focal area of thinned out epidermis and hypogranulosis, reflecting abnormal keratinocyte proliferation and differentiation [4].

The etiology of DSAP involves genetic and environmental factors. Mutations in the *mevalonate kinase* (MVK) and *FDPS* genes disrupt lipid metabolism, leading to keratinocyte dysfunction and lesion formation[5]. Ultraviolet (UV) radiation is a key environmental trigger, as lesions predominantly develop in sun-exposed areas [6]. Additionally, immunosuppression is a risk factor, further increasing susceptibility to DSAP in certain individuals [7].

Managing DSAP, particularly on the face, is challenging due to cosmetic concerns and the disease's chronic, recurrent nature. Treatment options range from topical agents to procedural interventions. Topical treatments, including 5-fluorouracil and imiquimod, have shown variable efficacy. Riad et al. reported that imiquimod is effective in reducing lesion size and improving facial lesion appearance [5]. Noborio et al. highlighted the benefits of fractional CO_2 laser therapy, which significantly improves lesion appearance while minimizing scarring, making it suitable for facial DSAP [9]. Procedural options, such as cryotherapy and CO_2 laser ablation, are effective for localized lesions but carry a risk of scarring, especially in sensitive facial areas Non-invasive treatments like photodynamic therapy have also been explored, but Dhillon et al. and Kim et al noted inconsistent results depending on lesion characteristics [2,8].

Our patient in this case was treated with tretinoin (0.025%) cream and calcipotriol (0.005%) ointment, applied on alternate nights, along with a daily moisturizer to mitigate irritation and sunscreen for photoprotection. This regimen aligns with findings by Severson et al., who documented a 91% reduction in lesion count after six months of combined tretinoin and calcipotriol therapy in refractory DSAP cases [12]. Similarly, Nakamura et al**.** highlighted the synergistic benefits of calcipotriol combined with other agents like adapalene, offering better outcomes than monotherapy [11].

Emerging treatments, such as chemical peels with glycolic acid (50%) and salicylic acid (25%), have also shown promise in managing DSAP. Lang et al. documented significant lesion improvement after three cycles of chemical peels, with minimal adverse effects and high patient satisfaction [7]. These treatments not only reduce lesion visibility but also offer cosmetic advantages, making them a valuable addition to DSAP management. Regardless of the chosen treatment, strict photoprotection remains crucial to prevent new lesions and slow disease progression [6].

Although DSAP is benign, long-standing lesions carry a small but notable risk of malignant transformation, particularly into squamous cell carcinoma. Sasson and Krain emphasized that lesions with significant hyperkeratosis and prolonged duration are more prone to malignancy [6]. Regular follow-up is necessary to monitor for malignant changes and manage emerging lesions effectively. Early detection and appropriate management are essential, not only to address clinical symptoms but also to mitigate cosmetic concerns, especially in cases with facial involvement.

Review of Literature

Several original studies have explored facial DSAP through case reports and interventional analyses. These include evaluations of topical therapies such as imiquimod, tretinoin, calcipotriol, and adapalene, as well as procedural interventions including CO_2 laser therapy and photodynamic therapy. Below is a summary of original published literature on facial DSAP.

Authors	Year	Title	Journal	Key Findings
Riad H et al.	2013	Disseminated Superficial Actinic Porokeratosis on the Face Treated with Imiquimod 5% Cream	Case Reports in Dermatology	Facial DSAP treated with imiquimod 5% cream showed remission and no relapse after 2 years.
Lang BM et al.	2019	Effective Treatment of DSAP with Chemical Peels – Customary Treatment for a Rare Disease	Journal of Dermatological Treatment	Chemical peels (glycolic and salicylic acid) effective in refractory DSAP, including facial involvement.
Severson KJ et al.	2024	DSAP Treated with Tretinoin and Calcipotriene	JAAD Case Reports	Dual topical therapy with tretinoin and calcipotriene yielded near-complete resolution in resistant DSAP.
Kim HS et al.	2011	PDT Combined with CO_2 Laser in Facial DSAP: Report of Two Cases	Annals of Dermatology	PDT combined with CO_2 laser showed significant improvement in facial DSAP with minor hyperpigmentation.
Noborio R, Morita A	2011	Split-face Trial of CO_2 Laser and Tacalcitol in Facial DSAP	Journal of Dermatology	CO_2 laser achieved better lesion clearance than tacalcitol; sustained results without scarring.
Dhillon KS et al.	2015	DSAP with Facial Involvement: A Case Report	International Journal of Advances in Medicine	Single session of CO_2 laser effectively managed facial and limb DSAP in a 30-year-old male.
Varala S et al.	2021	A Case Series of Disseminated Porokeratosis	Clinical Dermatology Review	Familial DSAP cases with facial lesions; underscores genetic and UV roles.
Cataldo K et al.	2014	Calcipotriol and Adapalene Therapy for DSAP	Indian J Dermatol Venereol Leprol	Combination of calcipotriol and adapalene was well tolerated and led to remission of facial DSAP.

CONCLUSION

Disseminated Superficial Actinic Porokeratosis (DSAP) with facial involvement is a rare manifestation that poses diagnostic and therapeutic challenges. This case of a 33 year old male highlights the need for clinical vigilance in recognizing atypical presentations. Accurate diagnosis requires histopathological confirmation, and treatment should be individualized, incorporating options like topical agents, laser therapy, or chemical peels alongside strict photoprotection. Regular follow-up is critical to monitor for malignant transformation, emphasizing the importance of early detection and timely intervention to prevent complications.

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- Conflict of Interest- None.

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Figure 1- DSAP involving the face

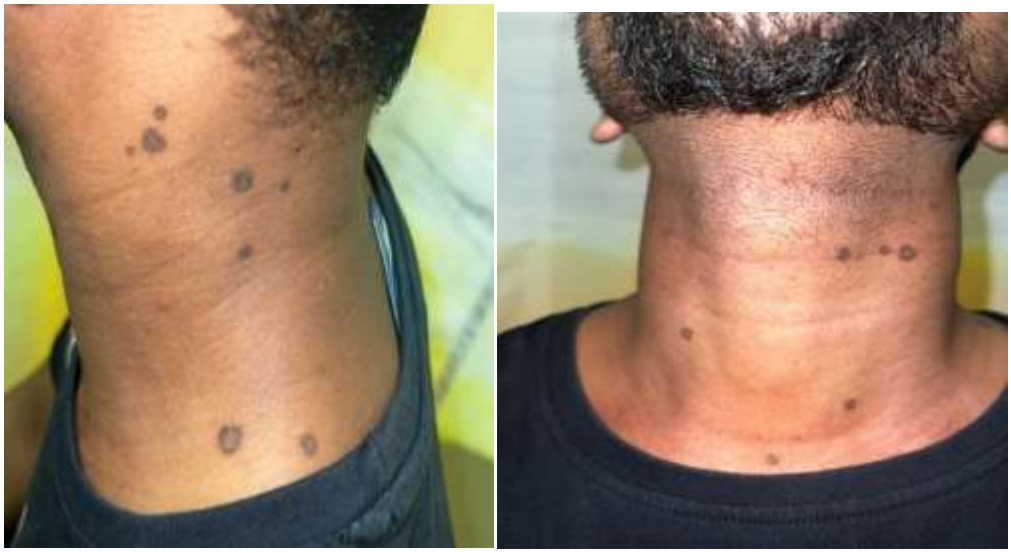

Figure 2,3- DSAP involving the neck

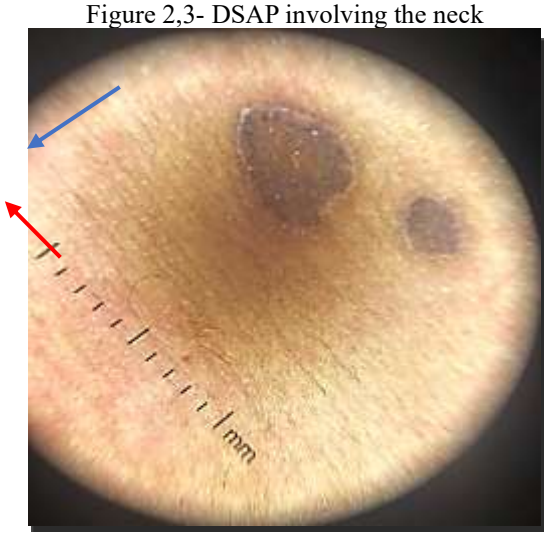

Figure 4-Dermoscopy (DermLite 5) showing hyperkeratotic rim (blue arrow), atrophic centre (red arrow)

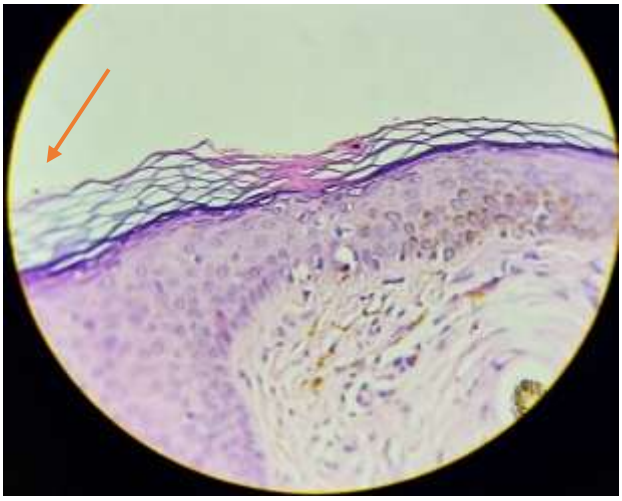

Figure 5- Cornoid lamellae on histopathology(orange arrow)