## CLINICAL VIGNETTE

# Disseminated Superficial Actinic Porokeratosis

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### *Clinical History*

A 72-year-old female has history of cutaneous disseminated superficial actinic porokeratosis (DSAP), and history of DM type 2, HTN, Hyperlipidemia, GERD. She has psoriasis and psoriatic arthritis which began at age 47. She noted her DSAP lesions at age 58. Biopsy was done but results could not be obtained. She was followed by dermatology and flare ups of DSAP were associated with arthritis flares. She characterized these flareups as intense itching and tenderness of her lesions. Lesions were primarily on her arms and legs.

She had several treatments for psoriasis and psoriatic arthritis through the years. She used etanercept for seven months with no significant response. She tried methotrexate for 4 months but had hair loss. She was on adalimumab for 21 months but stopped due to loss of efficacy. She then tried infliximab and abatacept which did not help at all. Secukinumab worked well for her psoriasis and helped with DSAP, but with little impact on her arthritis. Sulfasalazine was added for arthritis, but was stopped for gastrointestinal toxicity. She was not able to remain on secukinumab due to insurance restrictions. She also had insurance restrictions with tofacitinib, upadacitinib and certolizumab. Methotrexate was restarted but her transaminitis worsened to three to four times the upper limit of normal and resolved with discontinuation of methotrexate. She is currently on abatacept monitoring for response to this treatment.

Topical treatments continued through all these years with betamethasone and calcipotriene ointment. She also uses compounded preparations with zinc and clobetasol. Previously used clobetasol, fluocinonide, triamcinolone, desonide, tretinoin 0.05% cream and 5fluorouracil without relief and light therapy which did not help.

Patient's maternal uncle had psoriasis. No other family members had psoriasis or DSAP. She does not smoke or drink ETOH. She reports no recreational drug use.

Physical exam showed a well-developed woman in no apparent distress. Skin exam showed mild scalp flaking and erythematous, scaly papules and plaques on chest, abdomen, back, bilateral arms, thighs, lower legs. There are circular macules with raised rim on forearms, dorsum of hands, thighs and lower legs. (Figures 1 and 2).

Hands were without acute swelling or redness. There was tenderness in the left carpometacarpal joint, wrists, and bilateral olecranon. Right shoulder had anterior pain on active range of motion. Knees, ankles and feet were normal.

Labs showed normal CBC, CMP and CRP. ESR was mildly elevated at 33. RA, cyclic citrulline antibody and HLA B27 are negative. X-rays of hands and feet showed no erosions or evidence of inflammatory arthropathy.

### *Discussion*

Disseminated superficial actinic porokeratosis (DSAP) is a genetic skin disease of disordered keratinization causing dry, scaly patches. It is considered precancerous. This occurs in sun-exposed areas of the skin. A lack of awareness about DSAP may result in misdiagnosis and inappropriate management of this condition.[1]

It is an autosomal dominant inherited disorder with no clear pathophysiologic mechanism. Postulated mechanisms besides genetics include ultraviolet (UV) radiation, trauma, infection, and immunosuppression (as in transplant patients).[2,3] These lesions can be mistaken for solar keratoses and other skin disorders including seborrheic keratosis, psoriasis, discoid eczema, annular lichen planus or other rarer dermatological conditions.[3] Females are slightly more affected, more commonly with onset between 30 to 40 years old. The legs, forearms, shoulders, and back are most affected with the face, palms, and soles usually spared.[4]

Skin lesions of DSAP are usually maculopapular with raised borders and can be pruritic. They can also be asymptomatic. There is no standard therapy, but several treatment regimens have evolved, and include topical diclofenac, retinoids, vitamin D analogs, 5-fluorouracil, imiquimod, cryotherapy, photodynamic therapy, statin, and laser therapy. There is a 7.5 to 10% risk of malignant transformation to squamous cell carcinoma or basal cell carcinoma.[5,6]

The diagnosis of DSAP is usually clinical with help of dermoscopy. It can be missed by skin biopsy if the specimen does not include the rim, or is poorly oriented, or does not include the horny ridge.[7]

Patients should be educated on sun protection. This includes wearing long sleeves, skirts, and slacks and applying sunscreen.

Patients with frequent sun exposure should undergo regular skin checks and family screening if DSAP is present.

***Conclusion***

In conclusion, DSAP is a rare disease that can be confused with other dermatologic conditions. Recognizing the illness is important due to risk of malignant transformation. Lesions occur in sun-exposed areas and do not respond to conventional therapy, warranting dermatologic evaluation.

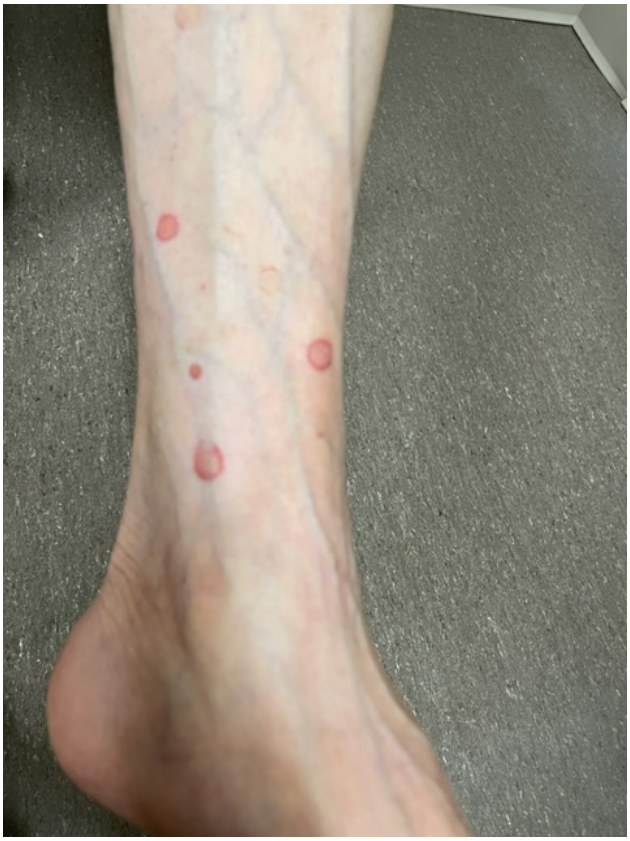

*Figure 1*

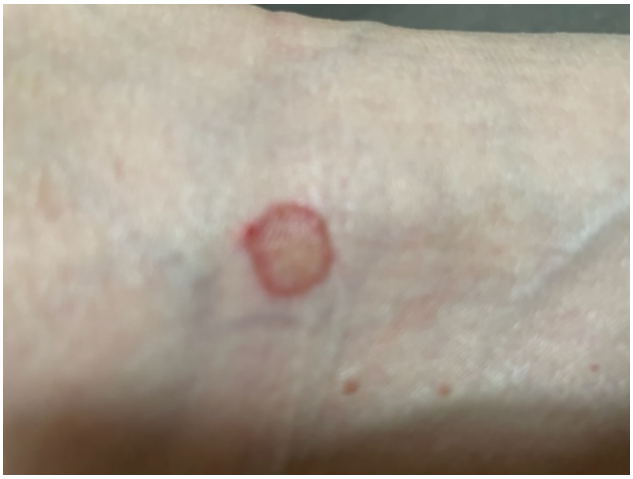

*Figure 2*

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