

Disseminated Superficial Actinic Porokeratosis: Case report

Montes-Flores Zyanya Lorely¹, Luna-Salazar Arely², Lucía-Achell Nava¹, Rosas- García Mariela Rosario², Pastrana-Otero María Fernanda², Dominguez-Navarrete Xymena², Dominguez-Navarrete Renata²

¹Centro Médico Nacional 20 de noviembre

²Clínica de Oncodermatología UNAM

ABSTRACT

Background: Disseminated superficial actinic porokeratosis is a rare disorder that presents abnormal clonal keratinization of unknown etiology. Frequently associated with UV radiation, genetic factors, and immunosuppression. It manifests clinically as solitary or multiple annular plaques surrounded by a hyperkeratotic border. Various clinical forms have been described, including disseminated superficial actinic porokeratosis, the most common subtype and represented in the following clinical case.

Clinical case: A case of a 62-year-old male is presented who presents with dermatosis characterized by annular erythematous squamous plaques spread to the hands, chest and upper and lower extremities and chest with collarette scales of chronic evolution. A clinical diagnosis is made, confirmed by classic histopathology and dermoscopic findings.

Conclusions. Disseminated superficial actinic porokeratosis is rare, difficult to diagnose and similar to multiple pathologies; This is why it is important to recognize them clinically and achieve timely treatment of potential malignancy that they present.

KEYWORDS: Porokeratosis, hyperkeratosis, cornoid lamella

ARTICLE DETAILS

Published On:
18 June 2024

Available on:
<https://ijmscr.org/>

INTRODUCTION

Porokeratosis is a rare, autosomal dominant, variably penetrant disorder of abnormal clonal keratinization of unknown etiology.

It manifests clinically as solitary or multiple annular plaques surrounded by a hyperkeratotic border. Various clinical forms have been described including disseminated superficial actinic porokeratosis, disseminated superficial porokeratosis, porokeratosis of Mibelli, linear porokeratosis, eruptive disseminated porokeratosis. 2

Porokeratotic lesions are progressive and have malignant potential, especially large and long-lasting lesions, as well as causing pruritus and cosmetic problems for some patients. 2,3

CASE REPORT 62-year-old male, native and resident of Mexico City, Mexico, administrator. History of insulin

resistance in treatment with Metformin and Semaglutide. COPD of 15 years of evolution treated with inhaled corticosteroids.

She presents dermatosis of 2 years of evolution with initial diagnosis of psoriasis, in treatment with topical steroids without improvement. Subsequently on dorsum of hands and dissemination to extremities and thorax, asymptomatic. Physical examination revealed disseminated dermatosis on the back, left shoulder and upper and lower extremities, bilateral with a tendency to symmetry characterized by multiple oval and round erythematous squamous plaques of 5 mm to 1 cm, surface with atrophic center and edges with collarette scale, hyperkeratotic with chronic asymptomatic evolution (Fig 1 ABCD and Fig 2 AB).

Disseminated Superficial Actinic Porokeratosis: Case report

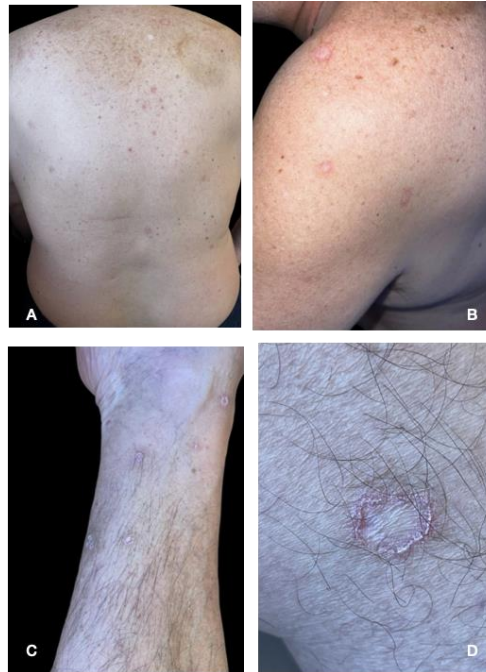


Figure 1. A,B,C,D

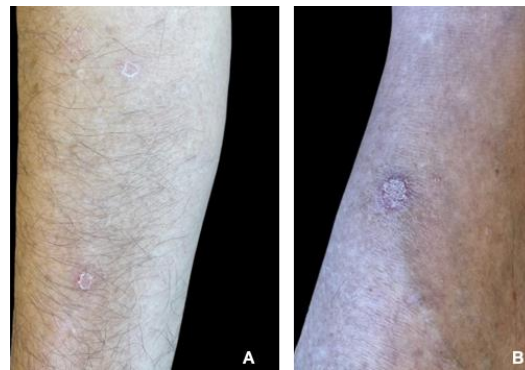


Figure 2.A, B

Dermoscopy non-melanocytic lesion characterized by a double whitish-whitish keratotic border, in the center red

dots, reddish lines and some lesions with a homogeneous central area (Fig. 3. A and B).

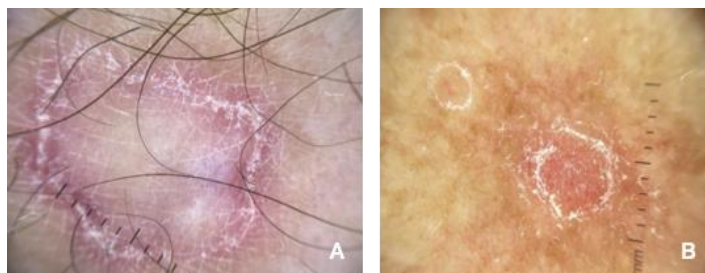


Figure 3. A, B

A skin biopsy was performed and reported: Photomicrography of skin panoramic section, where we observed compact lamellar stratum corneum and presence of lamella corneide: column of stratum corneum containing parakeratosis, vacuolated keratinocytes and absence of granular layer, discrete flattening of the interpapillary processes in dermis with normal adnexa and presence of lymphohistiocytic

Disseminated Superficial Actinic Porokeratosis: Case report

infiltrate in dermis (Fig 4: A and B).

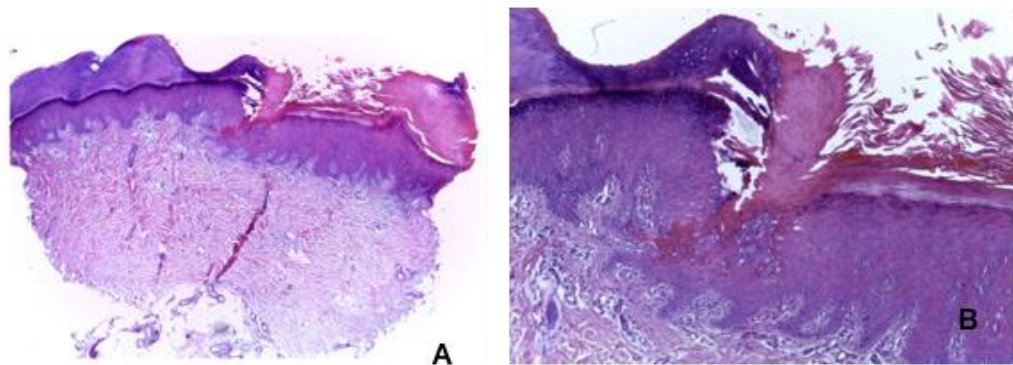


Figure 4. A, B

DISCUSSION

Disseminated superficial actinic porokeratosis is the most common form of porokeratosis, directly related to photoexposure.

Described as an autosomal dominant genodermatosis with loss of function in the following chromosomes: 12q23-2-q24-1, 12q24-1-q24-2, 15q25-1-q26-1, 1p31-3-q31-1 and 16q24-1-q24-3. 2,4,5

Recently, mutations have been found in the mevalonate kinase (mvk) gene, blocking the mevalonate pathway causes mitochondrial dysfunction in keratinocytes that produce type 1 keratin, associated with protection against UV radiation, and this dysregulation causes keratinocyte apoptosis.

The malignant potential is due to overexpression of the p53 gene, and occurs mostly in immunosuppressed or elderly patients associated mainly with Bowen's disease, epidermoid and basal cell carcinoma. This risk of malignancy occurs in 6.9 to 11.4% of patients. 2,4, 6

It starts in the fourth decade of life with predominance in women. It manifests with multiple small annular plaques that converge in skin-colored or hyperpigmented plaques with a keratotic border, distributed symmetrically on the back, extensor side of the extremities and shoulders. They are usually asymptomatic and cause pruritus in up to 15% of patients, with exacerbations following photoexposure. 2,3,5

Diagnosis is based on clinical findings, supported by histopathological study. The classic row of parakeratotic cells defined as a cornoid lamella, with hypogranulosis and occasionally, dyskeratotic cells, keratinocytes with vacuolization and eosinophilic spongiosis may be noted. In the dermis, perivascular inflammatory infiltrate. It is a classic sign of this entity but not pathognomonic. 2,8

Dermoscopy of these lesions reveals a brown center of atrophy surrounded by a scaly annular border.

In a study performed by Nicola et al, the following characteristics were determined in 6 patients: whitish border at the periphery of the lesion "volcanic crater lines seen from above", homogeneous atrophic central area, brown dots or globules, linear vascular structures crossing the lesion or

pinpointing the lesion. UV dermoscopy highlights the hyperkeratotic border as a "diamond necklace". 2

Proposed treatments include in the first instance the use of emollients and strict photoprotection. The following have also been used: 5-fluorouracil, imiquimod, vitamin D analogs, topical diclofenac, retinoids, 5-fluorouracil, cryotherapy, topical and oral retinoids, with variable results. 5

In our case, calcipotriol, general skin care and photoprotection were initiated, with partial response. New treatment proposals have been described such as the use of topical cholesteryl/lovastatin as well as CO2 laser. 7

CONCLUSION

Disseminated superficial actinic porokeratosis is a keratinization disorder, difficult to diagnose and regularly presented in photoexposed areas. In the case presented, a male patient with late onset of the lesions was diagnosed by clinical, dermatoscopic and histopathological features. This is a very important entity to recognize due to the risk of malignization.

REFERENCES

- I. Vargas-Mora P, Morgado-Carrasco D, Fustà-Novell X. Porokeratosis: A Review of Its Pathophysiology, Clinical Manifestations, Diagnosis, and Treatment. *Actas Dermosifiliogr* (Engl Ed). 2020 Sep;111(7):545-560. English, Spanish. doi: 10.1016/j.ad.2020.03.005. Epub 2020 May 13. PMID: 32401728.
- II. Nicola A, Magliano J. Dermoscopy of Disseminated Superficial Actinic Porokeratosis. *Actas Dermosifiliogr*. 2017 Jun;108(5):e33-e37. English, Spanish. doi: 10.1016/j.ad.2015.09.025. Epub 2016 Mar 23. PMID: 27015657.
- III. Florat GD, Viera CF, Rodríguez VKM, et al. Porokeratosis actínica superficial diseminada: reporte de caso y revisión de la literatura .

Disseminated Superficial Actinic Porokeratosis: Case report

- Dermatología Cosmética, Médica y Quirúrgica. 2019;17(1):42-45.
- IV. Logeshwari J, Patel ND, Varala S, Krishna AV. A case series of disseminated porokeratosis. Clin Dermatol Rev 2021;5:213-6.
- V. Das A, Vasudevan B, Talwar A. Porokeratosis: An enigma beginning to unravel. Indian J Dermatol Venereol Leprol 2022;88:291-9.
- VI. Waqar, M. U., Cohen, P. R., & Fratila, S. (2022). Disseminated Superficial Actinic Porokeratosis (DSAP): A Case Report Highlighting the Clinical, Dermatoscopic, and Pathology Features of the Condition. *Cureus*. <https://doi.org/10.7759/cureus.26923>
- VII. Tomsitz,D, Biedermann , T, Successful treatment of disseminated superficial actinic porokeratosis with topical 2% cholesterol/2% lovastatin cream: a case series with 7 patients. JEADV 2022; 36, e1–e79
- VIII. Quiroz-Valencia N, Vargas Bermudez MJ. Disseminated Superficial Actinic Porokeratosis. Actas Dermosifiliogr. 2016 Dec;107(10):858. English, Spanish. doi: 10.1016/j.ad.2015.07.022.