Dermatology Online Journal

Dermatology Online Journal
UC Davis

Letter: Disseminated superficial porokeratosis and pyoderma gangrenosum

Nicola di Meo MD, Catharina Fluehler, Valentina Perkan, Giusto Trevisan MD
Dermatology Online Journal 16 (10): 15

Piazza Ospedale 1, Trieste, Italy. nicodimeo@hotmail.it

Abstract

Disseminated Superficial Actinic Porokeratosis (DSAP) is usually triggered by sun exposure. In some cases sun exposure is not essential and this skin disease is related to immunosuppression. Many associated diseases are described in the literature. We report a clinical case of a patient affected by pyoderma gangrenosum, who developed DSAP.

Introduction

Disseminated superficial actinic porokeratosis (DSAP) is an inherited skin disease characterized by many uniformly small, annular, anhidrotic, keratotic lesions developing during the third or fourth decade of life on sun-exposed areas of skin [**1**]. Usually it develops during the summer months and disappears or improves during the winter. In some cases immunosuppression may favor its development and sun exposure is not always essential [**2**]. We report a clinical case of a patient treated for pyoderma gangrenosum, without a history of sun exposure or familiar skin disease. He developed DSAP in association with the occurrence of pyoderma gangrenosum and/or its treatment.

Clinical case

An 80-year-old man in good clinical condition was admitted with a large ulcer induced by a trauma on his left leg 10 days before.

This lesion was described as worsening rapidly to a dimension of 15 cm x 12 cm. The boarders were sharp; the edges were undermined and granulation tissue with purulent exudate was noted. The ulcer was painful without itch. The patient was afebrile but his leukocyte count was reported as 30000 cell/mm^3, with an absolute neutrophilia (Figure 1).

After ruling out the presence of infection and vasculitis, we considered the diagnosis of pyoderma gangrenosum and started a systemic therapy with clofazimine 100 mg/day by mouth and methylprednisolone 40 mg/day I.V. for two weeks. We then added to the therapy the antibiotics, teicoplanin 200 mg/day I.V. and piperacilline 2 g and Tazobactam 250 mg/day I.V.

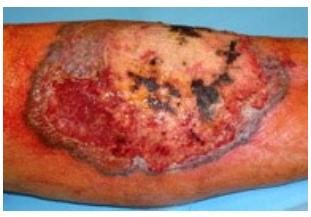

Figure 1

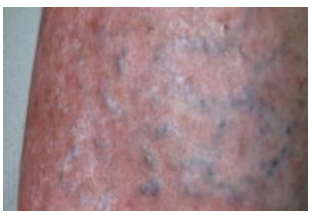

Figure 2

The patient was worked-up for underlying conditions, such as inflammatory bowel disease, rheumatological, hematological disease, and malignancy, but extensive testing yielded negative results.

After surgical and medical therapy the ulcer resolved and the patient continued oral therapy with methylprednisolone (Figure 2). The steroid was tapered by 4 mg/ week and clofazimine was continued at a dose of 100 mg daily.

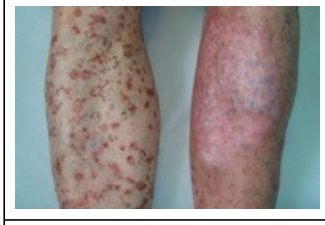

Figure 3

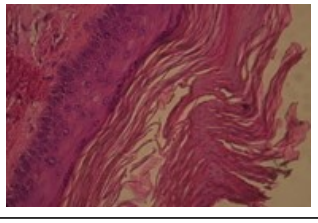

Figure 4

When the patient returned for follow up two weeks after his initial hospital discharge, he exhibited many small light brown variable-sized macules and patches with a threadlike border on the extensor surfaces of the arms and legs. These lesions involved only his extremities, sparing the trunk and the face. There was no itch or other cutaneous symptoms (Figure 3). There was no history of sun exposure and no family history of skin diseases.

superficial porokeratosis showing hyperkeratosis with the typical characteristics of cornoid lamella, a parakeratotic column without a granular layer (Figure 4).

This new clinical condition was treated with nonaggressive topical treatment and resolved rapidly, in a few weeks. Despite the reduction of clofazimine 50 mg on alternate days, to date he has not developed any recurrence of DSAP or pyoderma gangrenosum. The patient is seen in follow up every 6 months.

Discussion

Porokeratosis represents a group of epidermal cornification diseases, with an autosomal dominant transmission. However, there are many sporadic episodes of porokeratosis linked to spontaneous mutation. Clinically we can recognize five types of porokeratosis. Disseminated superficial actinic porokeratosis is an inherited disease described by Chernosky in 1966 that is characterized by the eruption on sun exposed areas of many small round brownish lesions surrounded by a hyperkeratotic margin. Many studies and case reports emphasize that sun exposure isn't always essential in the development of this clinical condition. Sometimes autoimmune diseases or their immunosuppressive treatments have been recognized as trigger factors in its development. In the literature there are cases of Sjogren syndrome [**4**], dermatomyositis [**5**], scleroderma [**6**], and rheumatoid arthritis [**7**] followed by DSAP. In our case we found a timely association with pyoderma gangrenosum. Hence, we suggest that the immunosuppressant treatment of PG could have played a role in the eruption of our patient's DSAP, but we can't exclude a role for an immune mechanism surrounding the pyoderma gangrenosum.

References

1. Chernosky ME, Freeman RG.Disseminated superficial actinic porokeratosis (DSAP). Arch Dermatol. 1967 Dec;96(6):611-24. [**PubMed**]

2. Bencini PL, Tarantino A, Grimalt R, Ponticelli C, Caputo R Porokeratosis and immunosuppression. Br J Dermatol. 1995 Jan;132(1):74-8. [**PubMed**]

3. Lederman JS, Sober AJ, Lederman GS. Immunosuppression: a cause of porokeratosis? J Am Acad Dermatol. 1985 Jul;13(1):75-9. [**PubMed**]

4. Terranova M, Amato L, Massi D, Fabbri P. Disseminated superficial actinic porokeratosis in a patient with Sjögren syndrome. Skinmed. 2003 Nov-Dec;2(6):390-1. [**PubMed**]

5. Monteagudo-Sánchez B, Ginarte M, Durana C, Labandeira J, de las Heras C, Cacharrón JM. Porokeratosis in a patient with dermatomyositis. Actas Dermosifiliogr. 2006 Dec;97(10):650-2. Spanish. [**PubMed**]

6. Kluger N, Guilpain P, Leclerc-Mercier S, Emilie S, Guillevin L, Mouthon L. Superficial porokeratosis of the lower limbs during systemic scleroderma Presse Med. 2009 Jan;38(1):146-8. [**PubMed**]

7. Jung JY, Yeon JH, Ryu HS, Youn SW, Park KC, Huh CH. Disseminated superficial porokeratosis developed by immunosuppression due to rheumatoid arthritis treatment. J Dermatol. 2009 Aug;36(8):466-7. [**PubMed**]