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Coexistence of porokeratosis of Mibelli with nail dystrophy to disseminated superficial porokeratosis

Ouédraogo Nomtondo Amina, Ouédraogo Sidnoma Muriel, Tapsoba Gilbert Patrice, Andonaba Jean Baptiste, Niamba Pascal, Traoré Adama

ABSTRACT

Introduction: Porokeratosis is a group of rare hereditary or acquired disorders of epidermal keratinization characterized by keratotic lesions with an atrophic center and a prominent peripheral ridge, the histological hallmark of which is the cornoid lamella. Many clinical variants have been identified. Although different types of porokeratosis have been previously reported to coexist in a single individual, such combinations are rare. We report coexistence of porokeratosis of Mibelli with nail dystrophy to disseminated superficial porokeratosis in a black African male.
Case Report: A 40-year-old male, living in Ouagadougou consulted for lesions showing hyperkeratosis evolving since his childhood. His fater had also similar lesions. Lesions started on his feet with a progressive extension to the rest of the body. Cutaneous examination revealed hyperpigmented plaques of 1–10 cm of diameter with central atrophy and distinct keratotic ridge. Keratotic lesions were located on feet, legs, thighs, hypogastria, buttock, and external genital organs. Lesions showing hyperkeratosis in the trunk were more superficial and less hyperkeratosis. Pachyonychia was seen on the two big toes. Histopathology showed parakeratotic column or cornoid lamella surrounding vacuolized keratinocytes with a granulous layer absent. Based on the clinical manifestations and histological features, diagnosis of porokeratosis of Mibelli associated with superficial disseminated porokeratosis was made. A treatment based on electrocoagulation, local keratolytic was given with improvement of keratotic lesions of buttocks.
Conclusion: The originality of our case resides in the coexistence of porokeratosis of Mibelli with nail dystrophy and disseminated superficial porokeratosis in a black African male.

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Keywords: Black African man, Cornoid lamella, Nail dystrophy, Porokeratosis

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INTRODUCTION

Porokeratosis is a group of rare hereditary or acquired disorders of epidermal keratinization characterized by keratotic lesions with an atrophic center and a prominent peripheral ridge, the histological hallmark of which is the cornoid lamella [1].

Many clinical forms have been individualized, the local forms include porokeratosis of Mibelli, palmoplantar porokeratosis, linear porokeratosis, punctuated porokeratosis, and the diffused forms with disseminated superficial porokeratosis (DSP), disseminated actinic porokeratosis (DAP), and disseminated palmoplantar porokeratosis [1, 2]. Even if there is a coexistence of various clinical forms [3], the one between porokeratosis of Mibelli and disseminated superficial form is rarely described. We report coexistence of porokeratosis of Mibelli with nail dystrophy to disseminated superficial porokeratosis in a black African male.

CASE REPORT

A 40-year-old African male, living in Ouagadougou (Burkina Faso), similar lesions, consulted at the Dermatology Department of Raoul Follereau Center for lesions showing hyperkeratosis evolving since his childhood, starting on the back of the foot with a progressive extension to the rest of the body. His father also had similar lesions. In addition, the patient was cisealing the most hyperkeratosis lesions on the buttocks and blunting them in order to be able to sit.

Cutaneous examination revealed hyperpigmented plaques of 1–10 cm of diameter with central atrophy and distinct keratotic ridge. Keratotic lesions were located on feet (Figure 1), legs, thighs, buttocks, external genital organs (EGO) (scrotum, penis) (Figure 2), and hypogastria (Figure 3). The lesions of the trunk (chest, back) were more superficial and less hyperkeratosis (Figure 4).

It existed pachyonychia nail dystrophy of the two big toes, dividing into two parts the ungueal tablets with crenelated aspect of their borders, forming a ventral pterygium (Figure 1). The rest of the examinations were normal.

Histopathology showed a parakeratotic column or cornoid lamella surrounding vacuolated keratinocytes with a granulous surface absent, compatible with the

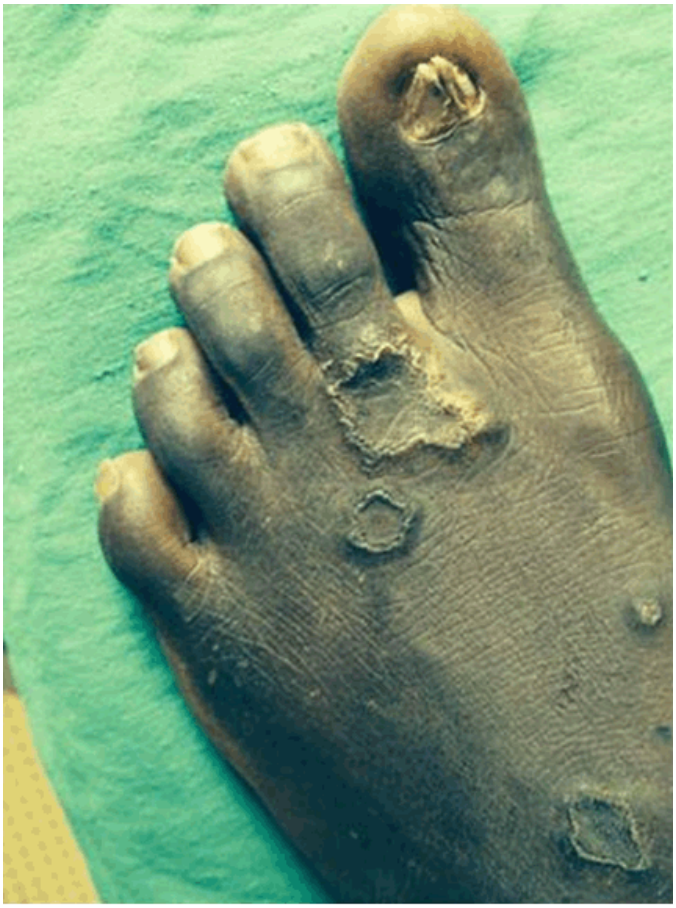

Figure 1: Porokeratosis of Mibelli lesions on the feet.

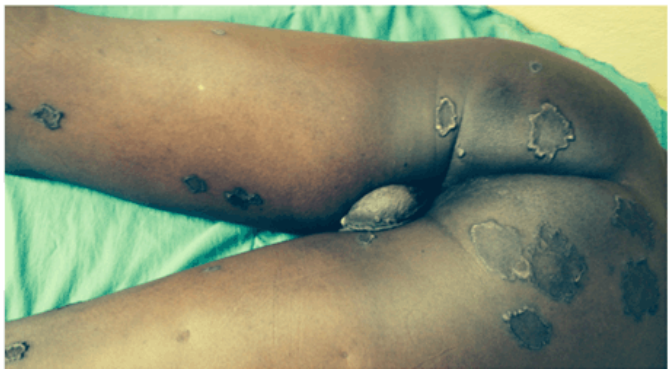

Figure 2: Porokeratosis of Mibelli lesions on buttocks, scrotum, thighs.

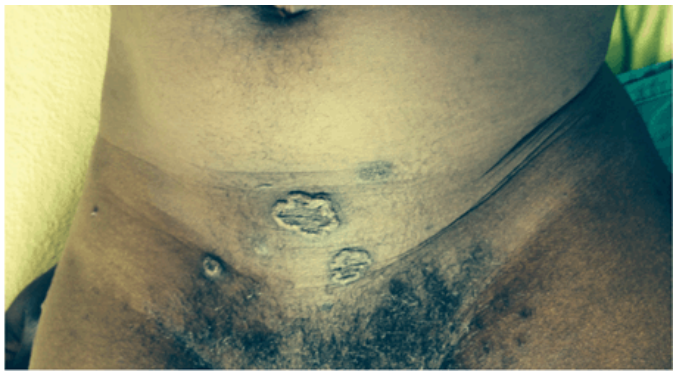

Figure 3: Porokeratosis of Mibelli lesions on hypogastria.

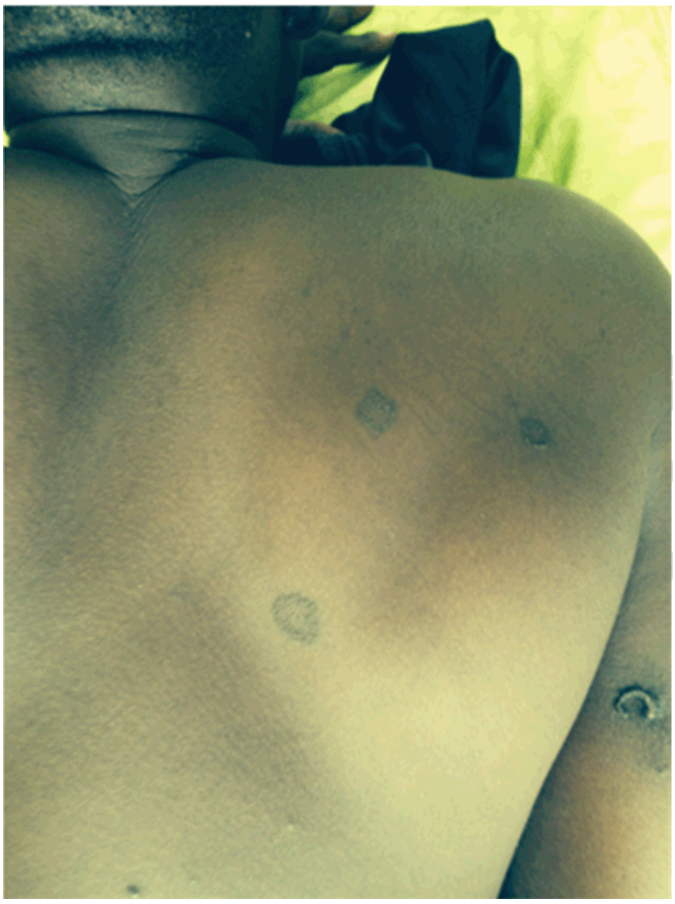

Figure 4: Superficial disseminated porokeratosis on the back.

diagnostic of porokeratosis. No malignant degeneration was noticed.

Retroviral serology was negative. Hemogram, kidney and hepatic function were normal. Chest radiography and ultrasonography of the abdomen was normal.

A treatment based on local keratolytic (30% salicylic vaseline then topic retinoid) was given. Electrocoagulation of the most prominent spelling lesions of the buttocks and the EGO had improved possibilities for sitting posture.

DISCUSSION

Described since 1893 by Mibelli, the etiology of porokeratosis is not totally clarified even if the most admitted etiology would be the expansion of a moving clone of epidermal keratinocytes, which could be transmitted genetically [1].

Porokeratosis will appear mainly to people of phototype I to IV, the Italians being particularly at risk [1, 2]. They are more rare for the phototype V and VI, and disseminated superficial forms being more frequently described on this phototype [4, 5].

The diagnosis of porokeratosis in our patient was made on the following arguments:

- In literature, porokeratosis of Mibelli was more reported in male [1, 2].
- Evolution since childhood and the existence of similar lesions to his father.
- The clinical appearance of the lesions showing hyperkeratotic lesions, the raised border and atrophic center [1, 2].
- Histopathology showed a cornoid lamella, absent granular layer below the invagination and mild inflammatory infiltrate in the upper dermis.

However, this case fits with more than one form of porokeratosis. In fact, it presents the characteristics of porokeratosis of Mibelli: hyperkeratotic lesions, very warty, hard with atrophic center at the low part of the body, mainly on buttocks, hypogastria, EGO, inferior members are hyperkeratosis, associated to nail dystrophy [6]. The nails dystrophy identified in our patient although something rare is described in porokeratosis of Mibelli at atrophy type, dystrophy and pterygium [7]. In addition, our patient presents some more superficial lesions at the arms, the back, the chest and the neck. It is then a coexistence of two clinical forms of porokeratosis for the same patient, mainly porokeratosis of Mibelli and disseminated superficial porokeratosis (DSP).

Even if the coexistence of various clinical forms of porokeratosis were reported, the most frequently described was about the superficial disseminated form and the linear form, the one between porokeratosis of Mibelli and DSP is less frequent [8, 9]. The lesions not being in the exposed areas and the absence of an immunodeprimed area (kidney transplantation, treatment by corticotherapy, or immunosuppressor, infection HIV, etc.) did not allow us to keep a disseminated actinic porokeratosis. There was no lesion of palmar and plantar areas excluding the palmoplantar form. The non-linear disposal, which did not follow the Blaschko's lines, was also pushing for the linear form [1, 2].

The treatment of porokeratosis is difficult. Local treatments (with vitamin D, acid salicylic, 5 fluoro uracil, imiquimod, dermocorticoids), general treatments (retinoid, methotrexate), and physical treatments (cryotherapy, dynamic phototherapy, electrocoagulation, the dermabrasion, the laser, the chirurgic) were notified with various results (at palliative aim and non-curative) [1, 2]. In our patient, topical keratolytic treatment with 30% of salicylic acid then topical retinoid had poor result. An electrocoagulation of the lesions in buttocks and EGO had improved the patient possibilities to walk and sit without pains.

An increasing of the number and the size of the lesions do the evolution of porokeratosis through a slow but progressive aggravation. Our patient noticed a progressive extension of the lesions. Malignant degeneration of porokeratosis is rare and appears in 7.5% of the cases in a minimum of 35 years [1, 2]. Face to the painful lesions, and the duration of evolution of disease (more than 35 years), malignant degeneration was discussed, then

rejected by the clinical and histopathology aspect which did not showed any malignant transformation.

CONCLUSION

The originality of our case resides in the coexistence of porokeratosis of Mibelli with nail dystrophy and disseminated superficial porokeratosis in a black African male.

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Author Contributions
Ouédraogo Nomtondo Amina – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Ouédraogo Sidnoma Muriel – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Tapsoba Gilbert Patrice – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Andonaba Jean Baptiste – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
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Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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