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 father 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.
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 father 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.

Keywords: Black African man, Cornoid lamella, Nail dystrophy, Porokeratosis
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 ungual 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
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 topical 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 nonlinear 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, democorticoids), general treatments (retinoid, methotrexate), and physical treatments (cryotherapy, dynamic phototherapy, electrocoagulation, the dermabrasion, the laser, the chirurgical) 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.

Acknowledgements
We are thankful to Dr. Maodo Ndiaye for his support

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
Niamba Pascal – 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
Traoré Adama – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

REFERENCES
Edorium Journals: An introduction

Edorium Journals Team

About Edorium Journals

Edorium Journals is a publisher of high-quality, open access, international scholarly journals covering subjects in basic sciences and clinical specialties and subspecialties.

Invitation for article submission

We sincerely invite you to submit your valuable research for publication to Edorium Journals.

But why should you publish with Edorium Journals?

In less than 10 words - we give you what no one does.

Vision of being the best

We have the vision of making our journals the best and the most authoritative journals in their respective specialties. We are working towards this goal every day of every week of every month of every year.

Exceptional services

We care for you, your work and your time. Our efficient, personalized and courteous services are a testimony to this.

Editorial Review

All manuscripts submitted to Edorium Journals undergo pre-processing review, first editorial review, peer review, second editorial review and finally third editorial review.

Peer Review

All manuscripts submitted to Edorium Journals undergo anonymous, double-blind, external peer review.

Early View version

Early View version of your manuscript will be published in the journal within 72 hours of final acceptance.

Manuscript status

From submission to publication of your article you will get regular updates (minimum six times) about status of your manuscripts directly in your email.

Our Commitment

Six weeks

You will get first decision on your manuscript within six weeks (42 days) of submission. If we fail to honor this by even one day, we will publish your manuscript free of charge.*

Four weeks

After we receive page proofs, your manuscript will be published in the journal within four weeks (31 days). If we fail to honor this by even one day, we will publish your manuscript free of charge and refund you the full article publication charges you paid for your manuscript.*

Favored Author program

One email is all it takes to become our favored author. You will not only get fee waivers but also get information and insights about scholarly publishing.

Institutional Membership program

Join our Institutional Memberships program and help scholars from your institute make their research accessible to all and save thousands of dollars in fees make their research accessible to all.

Our presence

We have some of the best designed publication formats. Our websites are very user friendly and enable you to do your work very easily with no hassle.

Something more...

We request you to have a look at our website to know more about us and our services.

We welcome you to interact with us, share with us, join us and of course publish with us.

CONNECT WITH US

* Terms and condition apply. Please see Edorium Journals website for more information.

This page is not a part of the published article. This page is an introduction to Edorium Journals and the publication services.