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Dermatological Manifestations of Connective Tissue Diseases in Black People

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Abstract:

Objectives: This study aims at contributing to provide a better knowledge of dermatological manifestations of Connective Tissue Diseases (CTDs) in dark-skinned patients. Though they appear visible and unsightly on black skins, cutaneous manifestations are often misleading and not really known by clinicians.

Methods: This is a cross-sectional and descriptive study base on the patient's medical records concerning those consulted and admitted at the department of Dermatology of the Teaching Hospital of Treichville (Abidjan, Côte d'Ivoire), for CTDs over the time period of 10 years, from January 02, 2002 to December 31, 2011. We included in this study, dark-skinned patients consulted or admitted in the department for Connective Tissue Diseases.

Results: Over the period of ten years, 42 499 patients were consulted for various dermatoses. Of those patients, we found 364 cases of CTDs about 0.8% of cases. In frequency order, these CTDs were as follows: Cutaneous Lupus Erythematosus (0.46%), Systemic Scleroderma (0.26%), Dermatomyositis (0.07%), Systemic Lupus (0.05%) and Sub-acute Lupus (0.0%); as a rule, dermatological manifestations were similar to those in white people. However, we have some particular aspects related to erythematous lesions, Raynaud's syndrome and pigmentary disorders which take misleading aspects in black people.

Conclusion: The dermatological manifestation of CTDs in black people is polymorphous and moreover misleading. In our environment of poverty and where immunological examinations are very costly, a good knowledge of these clinical signs could guide biological and immunological evaluation and contribute to early diagnosis.

Keywords: Clinical manifestations; Connective Tissue Diseases; Black skin

Introduction

Connective Tissue Diseases (CTDs) are inflammatory autoimmune diseases which may affect several organs simultaneously including the skin [1]. These diseases require most often a multidisciplinary approach. In this approach, the dermatologist has a crucial role to play given that cutaneous, mucous membrane and skin appendage manifestations often are preceded by other conditions several months or years [2,3]. Few studies related to systemic diseases have been conducted on the dark skin.

This work aims at contributing to provide a better knowledge on the clinical manifestations of CTDs on dark skin which takes at times misleading aspects. As specific objectives, we seek to:

1. Determine the frequency of CTDs in our Department;
2. Identify the different dermatologic physical signs in our patients;
3. And describe the most frequent signs and/or clinical particularities in black patients.

Methods

We conducted a cross-sectional and descriptive study base on the patient's medical records concerning those consulted and admitted at the department of Dermatology of the Teaching Hospital of Treichville (Abidjan, Côte d'Ivoire), for CTDs over the time period of 10 years, from January 02, 2002 to December 31, 2011.

This Department is the reference center for the treatment of cutaneous diseases over the national territory.

During the time period of this study, 42,499 patients with various types of dermatoses were consulted in our Department. Of those patients, 481 were identified and labeled as cases of CTDs based on their medical records.

Data were collected by a survey fact sheet with a questionnaire including socio-demographic characteristics, clinical description of lesions and the results of laboratory analysis.

The lack of materials in our hospital and the financial difficulties of our patients did not allow us performing immunological examinations to diagnose such autoimmune diseases.

We selected these criteria for different autoimmune dermatoses as follows:

1. For Acute Lupus Erythematosus, the presence of at least four criteria of the American College of Rheumatology (ACR 1997) was required, of which compulsorily the positivity to the native DNA antibody;
2. For Sub-acute Lupus, discoid Lupus and Scleroderma, in addition to clinical dermatologic manifestations, the data of the

histological examination of cutaneous lesions should be in the records.

Sub-acute lupus was diagnosed when confronted with discrete hyperkeratosis with lesser abundant perivascular and periadnexal infiltrations.

Chronic lupus erythematosus was diagnosed when confronted with a very pronounced hyperkeratosis orthokeratosic forming corneous plugs in follicular openings.

Systemic scleroderma was diagnosed in front of major and minor criteria of the ACR and confirmed by histologic examination revealing a cutaneous sclerosis.

1. Regarding dermatomyositis, cutaneous and muscular conditions should compulsorily be associated with a considerable elevation of muscular enzymes: creatine-phosphokinase, Aldolase and Lactate dehydrogenase (LDH) and transaminases.

Due to the high costs of muscular biopsy and electromyogram, these examinations were not prescribed in the department as routine examinations for dermatomyositis diagnosis.

Incomplete records were not included in this study, Likes records which did not contain the variables to be studied and records with no laboratory examination results likely to confirm the diagnosis of CTDs. As well, the records of Caucasian and Asian patients were not included. Of the overall 481 patients recorded initially and considered as cases of CTDs, only 364 cases met the eligibility criteria.

Results

Hospital frequency of CTDs

Over the time period of study, we received in consultation at the department of Dermatology 42,499 patients with various dermatoses. Of them, 364 Cases of CTDs were diagnosed with about 0.8% of cases.

These CTDs were distributed as follows: 196 cases of chronic Lupus Erythematous 0.46%), 113 cases of Systemic Scleroderma (0.26%), 30 cases of Dermatomyositis (0.07%), 23 cases of Systemic Lupus Erythematosus (0.05%) and 2 cases of Subacute Lupus (0.0%).

Socio démographic characteristics of patients

Age:

The average age of our patients was 35.5 years and varies from 18 to 72 years old.

Sex:

Women were more concerned than men, respectively with 65.3 % of cases and 34.7% of cases. The sex-ratio was 2.1 in favor to women.

Dermatological Manifestations

Dermatological manifestations ate illustrated in table 1.

Physical Dermatologic Signs	Number (n)	Frequencies (%)
Chronic Lupus Erythematosus (CLE)		
Discoid Lupus Plaques	156/196	79.5
Scarring Alopecia	84/196	42.8
Systemic Scleroderma		
Skin Induration	113/113	100
Sclerodactyly	108/113	95.5
Vilitigo	69/113	61
Pigment Disorder		
Raynaud's syndrome	04/113	3.5
Dermatomyositis		
Eyelid Edema	30/30	100
Heliotrope	Aug-30	26.6
Gottron's Papules	Oct-30	33.3
Poikiloderma	Feb-30	6.6
Systemic Lupus Erythematosus (SLE)		
Discoid Lesions	20/23	86.9
Alopecia	16/23	69.5
Buccal Erosions	Jul-23	30.4
Erythematous Malar Rash	Apr-23	17.3
Extensive Cutaneous Necrosis	Feb-23	8.6
Photosensitivity	Feb-23	8.6
Sub-acute Lupus		
Erythematosus annular Plaques on cheeks (annular-polycyclic type)	02-Feb	100

Table: Physical dermatologic manifestations identified in Patients over the study period. **Note:** PS: One patients presented one and more physical dermatologic signs.

Discussion

The different aspects of CTDs are well documented in the Anglo-Saxon and European literatures, namely with regards to epidemiological, clinical and therapeutic aspects. Contrary, in Sub-Saharan Africa, there is a scarcity of studies on these autoimmune dermatoses.

We think that this lack of study in our context is linked to the complexity of the cutaneous manifestations of these CTDs on black skin.

Hospital frequencies and socio-demographic aspect of CTDs

In studies conducted in black patients, the hospital frequency of those CTDs remains low [4]. As a matter of fact, in this study

conducted in Abidjan, the overall hospital frequency of CTDs over ten years of follow-up was 0.8% of cases. Most of CTDs were observed in our practice. Women around thirty were more affected than men. The sex ratio was 2/1. The CLE was the most frequent form with 0.46% of cases followed by systemic scleroderma with 0.26% of cases and Dermatomyositis (DM) with 0.07% of cases

We think that in our studies, these low frequencies could probably be explained by underestimated cases of CTDs due to diagnosis difficulties related to the skin color.

Dermatological manifestations

CLE was the first CTD observed in the Department with 0.46% of cases. Regarding its clinical aspects, the classic signs reported in the literature are well limited atrophic erythemato-squamous plaques associated with a scarring alopecia [5-7]. Discoïd plaque was the most observed dermatologic sign in our study with 79.5% of cases. This frequency was also observed by other authors in Italy and in Tunisia respectively 70% and 73% of cases [6,7]. It was followed by scarring alopecia with 42.8% of cases in our study.

The classic triad of discoid lesion including erythema, central atrophy and hyperkeratosis, was less characteristic in our study. Thus, in black people, the peripheral erythema typically found on the plaque will rather appear as a misleading greyish hyperchromic edging. Moreover the central atrophy on dark skin, takes an achromic aspect which could misleadingly be considered as a vilitigo (Figure 1).

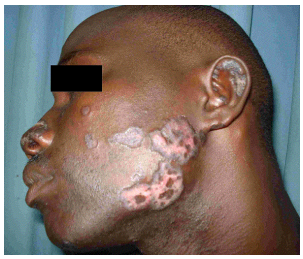

Figure 1: Lesion with hyperpigmented border

These atypical characteristics namely peripheral hyperchromic edging and the vilitigo aspect of central location constitute the particularity of the discoid lupus plaque on dark skin and should not be underestimated.

Scarring alopecia which represented the second manifestation of the CLE in our study, have no clinical particularity apart from its visible and unaesthetic character. This unaesthetic aspect could be explained by the important contrast it causes on black skin (Figure 2).

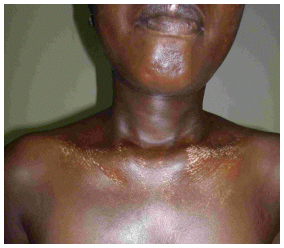

Figure 2: Scleroderma pigmentary disorder vitligo

In our series, systemic scleroderma was the second CTDs with 0.26% of cases. The most reported clinical manifestations in the literature are the Raynaud's syndrome. This sign may precede by several months or years the other signs and the cutaneous sclerosis which is progressive [8,9].

Raynaud's syndrome is seldom reported in CTDs on dark skin [10]. In our series, its hospital frequency was 3.5% against 40% of cases in Togo [11]. However, the diffuse sclerosis was the most observed sign in the studies carried out in the black African region. In our series, it was about 100% of cases, and it was less in Togo about 79.9% of cases.

The relative scarcity of the Raynaud's syndrome in the studies in black Africans could be explained on the one hand by the difficulties to identify this acrosyndrome and on the other hand, by the climatic conditions in our regions which are in general hot countries.

On black skin, the diagnosis of the Raynaud's syndrome cannot be undertaken easily by inspection, but it rather leans on a good questioning and a meticulous fingertips examination.

The questioning will check for the occurrence of digital pain during contact with cold (frozen matter, refrigerator) and the physical examination will check for the presence of finger pulp ulcer [12]. These signs will, in the firsthand lead to the suspicion of a previous occurrence of Raynaud's syndrome in the dark-skinned patients.

The high incidence of diffuse sclerosis in our patients could be linked to the absence of social security. This accounts for the fact that most of our patients go for consultation at a later stage of the disease. This diffuse cutaneous sclerosis does not display any clinical particularity.

For scleroderma in dark-skinned patients, lenticular achromia (Figure 3) which seems to be a very strong characteristic sign should moreover be recognized. In our series, it was found in 61% of cases and it was less in the study of Pitché in Togo about 53% of cases (in 7 patients over 13). Its clinical manifestations associate hypo and hyperpigmentation. These signs are mostly located on the chest and limbs where they create hypochromic, punctiform and lenticular zones. These lesions seem to be great diagnostic value of the systemic scleroderma in black patients; in as much as this vilitigoid lenticular aspect was not found in the review of literature in white skin patients [12].

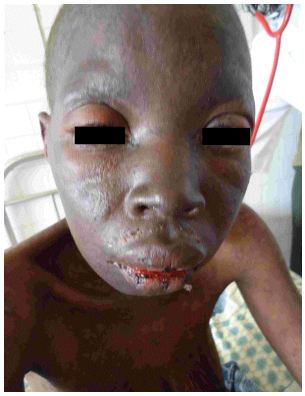

Figure 3: Dermatomyositis oedema more visible than heliotope

In our study, the frequency of dermatomyositis (DM) was 0.07% of cases. It is known that cutaneous lesions may precede by several months or years muscular manifestations [13] hence their importance. On Caucasian skins, these cutaneous lesions are typical and easily recognized by physician front of erythema associated with periorbital edema which is a very characteristic sign of the disease [14]. In our study, heliotrope rash was seldom found. It was reported in 26.6% of cases. In point of fact, heliotrope rash which is quite a visible sign on Caucasian skin seems to be hidden by the dark coloration of the skin. In our patients, it is mostly periorbital edema which constituted the most important manifestation. It was found in 100% of cases. It is an elastic and painless edema which constitutes on dark skin the key physical sign of DM (Figure 4).

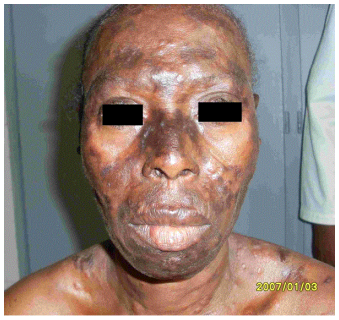

Figure 4: Systemic lupus erythematous- Butterfly rash with grey macules

It is worth calling for the clinician's attention on the fact that, any eyelid edema in dark-skinned patients without trauma and insect bite should evoke in the firsthand a dermatomyositis. This sign should therefore lead to the research of muscular weakness associated and the confirmation of the disease by the muscular enzyme dosage. And where it possible to carry out an electromyogram and a muscular biopsy to perform histological examination.

Gottron's papules found on the back of the hand were the second most observed clinical manifestations in our study about 33.3% of cases. On black skin, these lesions take a hypochromic aspect contrasting with the dark color of the skin explaining their easy identification.

Acute Cutaneous Lupus Erythematosus and Subacute Cutaneous Lupus were very rare in our study. They represented respectively 0.05% and 0% of cases. We think that the scarcity of these two affections should be linked to the difficulty of their diagnosis on dark skin.

Regarding lupus erythematosus, clinical manifestations were dominated by discoid lesions in 86.9% of cases and alopecia in 69.5% of cases. These data were in concordance with those in the study of Kombaté and peers on systemic lupus in Togo with about 87.5% of cases for discoid lesions, 56.2% of cases for malar rash and 43.7% of cases for alopecia [15].

Classic erythema in vespertilio of the SLE which is very characteristic on white skin [16,17] takes a misleading hyperchromic aspect on black skin. The difficulty of recognition of this characteristic sign could account for its underestimation observed in the studies conducted in black African [18].

As well, we seldom observed in our study, SLE clinical signs such as buccal erosion in 30.4% of cases and the photosensitivity in 8.6% of cases. Given that the diagnosis of the SLE is not easy on black skin, it would not be unnecessary to establish a clinical-immunological profile which would help to its diagnosis. Thus, the SLE profile in our study was characterized by:

1. The scarcity of malar rash, of buccal erosion and photosensitivity;
2. The high frequency of discoid lesions and alopecia;
3. And with regards to immunological aspects, the presence of native DNA anti-body

With regards to sub-acute lupus, its clinical aspect was dominated by annular erythematous plaques. These lesions are easily assimilated to a dermatomycosis on dark skin. In our environment, it is the histological examination that will allow us to orientate the diagnosis.

Conclusion

The dermatological manifestation of CTDs on black skin is polymorphous and moreover misleading. In our context of poverty where immunological examinations are very costly, a good knowledge of such clinical signs could guide biological and immunological evaluation and contribute to early diagnosis.

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