

## Scleroderma in a Black African Subject: A Study of 217 Cases in Cote d'Ivoire

BR Aka<sup>1</sup>, A. Diabaté<sup>1\*</sup>, M. Kaloga<sup>2</sup>, HS Kourouma<sup>2</sup>, B.Vagamon<sup>1</sup>, I. Gue<sup>1</sup>

<sup>1</sup>Dermatology Department, University Hospital of Bouaké, Côte d'Ivoire

<sup>2</sup>Dermatology Department, University Hospital of Treichville, Abidjan, Côte d'Ivoire

\*Corresponding author: Diabaté Almany, Dermatology Department, University Hospital of Bouaké, Côte d'Ivoire, Tel: 0022507985142; E-mail: docalmamy@yahoo.fr

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### Abstract

**Objective:** The goal of this study was to document the epidemiological, clinical, therapeutic and evolutionary profile of scleroderma in hospitals in Côte d'Ivoire.

**Method:** This was a descriptive study of the cases of patients suffering from scleroderma and received in the dermatology departments of Treichville and Bouake in Côte d'Ivoire from January 1983 to December 2015.

**Results:** The study of 217 cases of scleroderma revealed an increase in the number of scleroderma to 0.08% from 1983 to 2015. They predominated in women with a sex ratio (F/M) of 1.66. The mean age of patients was 25.35 years for localized scleroderma and 33.70 years for systemic scleroderma. Eighty point five (80.5%) of the subjects were of low socio-economic level. Seventy point eight (70.80%) of the patients consulted after a year of their disease. The most common clinical manifestations in the systemic scleroderma were the confetti skin bleaching and the Raynaud's syndrome. The extra-cutaneous manifestations were existing and varied. As for the localized scleroderma, three clinical aspects were found: morphea in plate, morphea in band, saber-like shape. The diagnosis in the majority of cases was done clinically. Therapeutically, the first-line treatment always remains the unsaponifiable of avocado oil and soya bean that give better results.

**Conclusion:** The results of this study confirm the scarcity of scleroderma in hospitals in Côte d'Ivoire and the clear predominance of females. It poses the difficulty of its management, which explains the many patients lost from sight.

**Keywords:** Scleroderma; Black skin; Côte d'Ivoire

### Introduction

Scleroderma is a connective tissue that is characterized by an over-activation of fibroblastic cells, lymphocytes and endothelial cells, resulting in an infiltration of the skin and the mucous membranes with young collagen that explain the sclerosis [1-4]. It includes two disease entities with a very different prognosis: mainly the localized scleroderma (LS), with benign progression and good prognosis, and the systemic scleroderma (SS) with cutaneous lesions and visceral involvement which constitute a morbidity factor and a poor prognosis [5,6]. The scleroderma in black Africa has the same clinical characteristics as those described in the literature [3-6], but its management is hampered in Africa due to the difficulties of access to care, but also to psychological and cultural contexts. In Côte d'Ivoire, there is little available data on scleroderma. The aim of this study was to determine prevalence of the scleroderma by report with other dermatosis encountered in the hospitals of Côte d'Ivoire, to describe sound epidemiological characteristics, to identify various clinical aspects of the disease, and to make a report of the progress of the disease in the treatment when this element is known in hospitals in Côte d'Ivoire.

### Materials and Methods

We carried out a descriptive study on the files for patients seen in consultation and/or in hospitalization in the dermatology departments of Treichville and Bouaké in Côte d'Ivoire. This study covered the period from 1 January 1983 to December 2015. We included in our study patients received for an LS or SS on the basis of the criteria of the ACR [7]. The Para-clinic examinations were requested on the basis of the functional symptomatology, except histology, blood count and lung radiography that were systematic. Also, no patient was immunologically tested. The development was considered favourable when the lesions were stationary and unfavourable when the lesions were increased in volume or the extension report showed an involvement of the viscera.

### Results

During the 32 years, 217 of the 257,168 patients received in dermatology departments had suffered from scleroderma, where from prevalence of 0.08% at the hospital. The patients' ages varied from 10 to 65 years, with an average of 30.7 years. Adolescents and young adults constituted the majority of the patients (63.10%). The predominant age group for individuals with localized scleroderma was 11 to 30 years and systemic scleroderma 21 to 40 years. The female predominance is clear with a sex ratio (F/M)=1.66. No matter the type of scleroderma, the female sex is predominant. We recorded 66.5% of LS and 33.5% SS. Most of the patients (76.40%) consulted after a year of disease

progression. The main symptoms observed (Table 1: overall results) were confetti skin bleaching (63.4%) (Figure 1). Cutaneous sclerosis (21.3%), hyperchromic maculae (14.1%) and the phenomenon of Raynaud (1.2%). The lesions were found in the limbs (44.3%), the head (16.1%), the bust (39.3%) and generalized in 10.5% (Figure 2). In the SS, the extra-cutaneous manifestations were dominated by digestive affections (52.5%) as well as pulmonary (17.9%), articular (16.5%), cardiovascular (11%), renal (1.2%) and bone (0.9%). All aspects of the Morphea were noted, dominated by plaque morphology (52%), band (24%), saber (20%) and gout (4%). The cutaneous biopsy performed in

167 patients confirmed the result in 90.77% of the cases. The blood count indicated that anemia was more common in the systemic sclerosis (70.9%). HIV infection in patients with systemic sclerosis was 13.6% whereas no patient with localized scleroderma was detected positive. The pulmonary radiography revealed a fibrosis (28.8%). In 16 patients who had a TOGD we noted that the digestive lesions predominate in the esophagus (50%). Repolarisation disorders were predominant (60%). The HTAP (52%) and fluid pericarditis (17.3%) were observed. There is also a predominance of distal arterial disease (75%).



**Figure 1:** Confetti skin bleaching.

From a therapeutic point of view, the extracts of unsaponifiable avocado and soya (UAS) were the basis of the treatment of our LS patients. The vasodilators (37.5%) and corticosteroids (20.8%) are the most associated with UAS in the SS. The evolution (Table 1: overall

results) was satisfactory in LS patients and 5.5% in SS. Regardless of the type of scleroderma, improvement was observed. There were 43 patients lost from sight and we deplored 1.4% deaths.

	localised Scleroderma n (%)	Systemic Scleroderma (%)
<b>Clinical aspects</b>		
Confetti skin bleaching (63.6%)	103 (47.5)	33 (15.2)
Skin Sclerosis (20.6%)	29 (13.4)	18 (8.3)
Hyperchromic maculae (14.3%)	22 (10.1)	9 (4.1)
Limited mouth opening (0.9 %)	2 (0.9)	0 (0)
Pulp ulcers (0.5%)	1 (0.5)	0 (0)
Sclerodactyly (0.5%)	1 (0.5)	0 (0)

Raynaud's syndrome (1.4%)	2 (0.9)	1 (0.5)
<b>Treatment</b>		
Extracts of unsaponifiable avocado and soya (69.6%)	144 (66.4)	7 (3.2)
Corticosteroid therapy (20.7%)	2 (0.9)	43 (19.8)
Vasodilators (29%)	0 (0)	63 (29)
<b>Evolution</b>		
Improvement	87 (40)	12 (5.5)
Steady state	42 (19.5)	22 (10.1)
Aggravation	0 (0)	8 (3.7)
Deaths	0 (0)	3 (1.4)
Loss from sight (43)	15 (6.9)	28 (12.9)

**Annex: Table 1:** Clinical aspects, treatment and evolution of the scleroderma.



**Figure 2:** Sclerodactyly.

## Discussion

In Côte d'Ivoire, the dermatology services do not have a monopoly on the management of scleroderma patients in hospitals. Some patients may be admitted directly to internal medicine, cardiology, pneumology, rheumatology or even neurology for severe pulmonary arterial hypertension or neurological manifestations. Our results are not exhaustive, but with 217 patients over a period of 32 years in a study on the activity of dermatology services (Treichville and Bouaké)

in Côte d'Ivoire, they confirm the scarcity of scleroderma in black Africa. Indeed, Keita [8] had recruited 35 cases of SS in 10 years in Mali and Adelowo [9] in Nigeria had reported 14 cases in 5 years. We believe that this very low frequency in black Africa could be explained, on the one hand, by the non-economic and geographical accessibility to health care services and, on the other hand, by a low exposure of the population to inducing toxicants (Silica, Solvents), since this part of the world is little industrialised. We found a clear female

predominance with a sex ratio of 1.66. It was 0.19 in the series of Dia et al. [10], confirming that scleroderma is mostly a prerogative of female [9,11] and of young subjects especially because in our study, the average age of patients was 30,7 years.

The semiological profile of our patients with SS matches the one described in the European population [1,7]. On the other hand, the confetti skin bleaching presents in all our patients is a peculiarity of the genetically pigmented skin. It dominates the clinical picture in the study by Keita et al. [8] who found it in 88.57% of their patients. This confetti skin bleaching could raise discussion in the subject of pigmented skin for other affections like a vitiligo, a sclerodermatomyositis, or even a lupus. But the existence of the cutaneous sclerosis and the absence of associated muscular involvement make it possible to eliminate these affections. The importance of the cutaneous sclerosis observed in Africans may suggest the severity of this disease in black Africa [12,13], but may be explained by the delay in consultation. The Raynaud's syndrome was found in 3 of our patients. Adelowo et al. [9] found two cases of Raynaud's phenomenon in a population of 14 patients. This confirms the rarity of this phenomenon in Africa [10] probably related to the warm tropical climate as it is triggered by the cold.

On the paraclinic level, our patients did not carry out certain examinations (manometry, cardiac ultrasound and immunoassay) for technical and financial reasons, shortage of equipments. However, the paraclinical tests carried out made it possible to confirm and monitor the different types of SS. This problem also arises in relation to the non-availability and/or non-accessibility to specific treatments for the management of certain symptoms such as pulp ulcers, Raynaud's phenomenon.

The extracts of unsaponifiable avocado and soya by general route were used in almost all our patients. In 50% of cases, they were used alone. The therapeutic combinations consisted of the prescription of local or general corticosteroids, vasodilators, antibiotics and other various drugs according to the symptomatology or dominant affection in the SS. In the literature, local or general corticosteroids had been constantly used for different forms of SS [14].

Finally, we had a very high rate of patients lost from sight. Twelve point nine (12.9%) of patients with SS were lost from sight after an average follow-up of 43.5 days and 6.9% of patients with SL were lost from sight after 17 days on average. We believe that this severe loss is related to the difficulty to manage chronic pathologies in general and the absence of a well codified treatment of scleroderma, since the therapeutics used only slow down the course of the disease without guaranteeing a definitive cure.

## Conclusion

Our study confirms the scarcity of scleroderma in black Africa and whose peculiarity of genetically pigmented skin is a confetti skin

bleaching. Moreover, it raises the problem of diagnostic and therapeutic difficulties for technical and economic reasons.

## Conflicts of Interest

The authors declare no conflict of interest.

## Author Contributions

All authors contributed to the writing of the manuscript had read and approved the final version.

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