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The Hand in Autoimmune Systemic Diseases: Epidemiology and Clinical Phenotypes of Pigmented Skin

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Abstract

Introduction: Systemic autoimmune diseases are non-organ-specific disorders with a heterogeneous clinical presentation. The clinical signs of these systemic diseases are varied on the hands. There are specific cutaneous or vascular manifestations of autoimmune diseases, and rheumatological manifestations such as polyarthritis, tendonitis and synovitis. The aim of this study was to evaluate the epidemiological aspects, clinical phenotypes and immunological abnormalities associated with the hand in autoimmune systemic diseases.

Patients and methods: We conducted a cross-sectional, descriptive, multicenter study in the Dermatology departments of Dakar from June 01, 2021 to November 01, 2021.All patients followed for systemic autoimmune disease during the study period were registered. Among them all patients with clinical signs of systemic autoimmune disease on the hands were included in the study. Data entry and analysis were performed using Microsoft Excel 2016 and SPSS version 23.

Results: We collected 66 cases of systemic diseases. Hand involvement was noted in 39 cases: systemic lupus erythematosus in 9 cases, systemic scleroderma in 17 cases, dermatomyositis in 2 cases and mixed connective tissue disease in 11 cases. In systemic lupus erythematosus, cutaneous signs included erythema in 2 cases, discoid plaques in 3 cases and annular plaques in one case. Vascular signs included Raynaud's syndrome in 2 cases, purpura in one case, palmar erythrosis in one case and digital necrosis in one case. Nail manifestations included longitudinal melanononychia in 3 cases. Systemic scleroderma was characterized by diffuse sclerosis of the hands in 13 cases, sclerodactyly in 10 cases, mottled achromia in 8 cases, Raynaud's syndrome in 15 cases and stellate pulpal scars in 8 cases. In 2 cases of dermatomyositis, there was transverse band erythema, Gottron's papule, nail set erythema with cuticle hypertrophy and Raynaud's syndrome. In mixed connective tissue disease, the following associations were observed: scleroderma with dermatomyositis in 4 cases, lupus with dermatomyositis in 3 cases, lupus with scleroderma in 3 cases, and Sharp's syndrome with a case of antiphospholipid antibody syndrome.

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Conclusion: The clinical manifestations of the hands are diverse in the course of systemic autoimmune diseases. The semiological particularities of skin lesions on the hands could contribute to the diagnosis and early management of these systemic diseases in order to improve the prognosis of these chronic diseases in our african countries.

Keywords: ***Autoimmunity; Hands; Systemic diseases; Dakar***

1. Introduction

Systemic diseases include all non-organ-specific autoimmune and/or inflammatory conditions [1]. Their prevalence is estimated between 5% to 10% of the world's population [2,3]. In sub-Saharan Africa, prevalence is also variable, and is rising sharply due to the improvement of diagnostic facilities and the availability of specialized doctors who can make the diagnosis [3,4]. Systemic lupus erythematosus remains the main systemic autoimmune disease, with a series of 340 cases reported in dermatology departments in Senegal [4]. The hand is a grasping organ, with anatomical osteoarticular, tendinous and synovial relationships, as well as a highly developed vascular system [5]. This explains the frequency and lesion polymorphism of clinical manifestations of the hand in systemic diseases. These include specific cutaneous manifestations, vascular signs and rheumatological manifestations such as polyarthritis, tendonitis, and synovitis [4]. Cutaneous signs have diagnostic and prognostic value. Systemic diseases are often discovered by severe visceral damage, hence the importance of early diagnosis [4]. Few studies have been reported in Africa on the clinical features of cutaneous manifestations of the hand in systemic diseases. The aim of this study was to evaluate the epidemiology, clinical phenotypes and immunological abnormalities associated with the hand in autoimmune systemic diseases.

2. Patients and Methods

This was a descriptive, cross-sectional, multicenter study with prospective recruitment of patients from June 1, 2021, to November 1, 2021 in the dermatology departments of Aristide Le Dantec hospital and the Hygiena Social Institute in Dakar. All patients followed for systemic autoimmune disease during the study period were registered. Among them all patients with clinical signs of systemic autoimmune disease on the hands were included in the study. The diagnostic criteria used were those of the 2019 ACR/EULAR classification for systemic lupus erythematosus (SLE) [6], the 2013 ACR/EULAR for systemic scleroderma [7], the 2004 ENMC for dermatomyositis [8] and Sydney 2006 for anti-phospholipid antibody syndrome (APLS) [9]. Data were collected by a dermatologist using a questionnaire that listed epidemiological, clinical, biological and immunological variables. Data entry and analysis were performed using Microsoft Excel 2016 and SPSS version 23. The χ^2 (Chi-square) test was used for bi-variate analysis and was considered significant for a p value <0.05.

3. Results

We collected 66 cases of systemic disease. Of these, 39 cases (59.1%) presented with clinical manifestations on the hands. These were systemic scleroderma in 17 cases (43.6%), mixed connective tissue disease in 11 cases (28.2%), systemic lupus erythematosus in 9 cases (23.1%) and dermatomyositis in 2 cases (5.1%). FIG. 1 shows the distribution of patients according

to the presence of a cutaneous manifestation on the hands in systemic diseases. Patients were followed on an outpatient basis in 31 cases, and as inpatients in 8 cases.

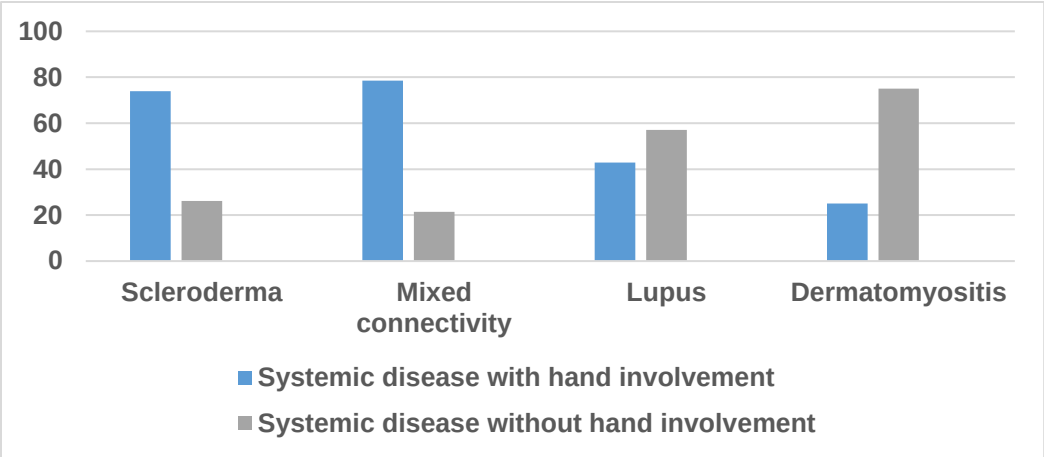

FIG. 1. **Distribution of patients according to the existence of hand involvement in systemic autoimmune diseases.**

Nine patients had lupus involvement of the hand. It involved 5 women and 4 men, i.e., a sex ratio of 0,8. The mean age was 38.7 years, range: 27-59 years. Skin lesions were chronic lupus erythematosus in 4 cases, acute lupus erythematosus in 4 cases and subacute lupus erythematosus in one case.

Acute lupus erythematosus was characterized by inter-articular erythema (FIG. 2A) in one case, inter-articular papules in one case and vascular signs such as Raynaud's syndrome in 2 cases, diffuse purpura in one case (FIG. 2B), palmar erythema in one case and digital necrosis in one case. In subacute lupus erythematosus, the plaques were annular in one case (FIG. 3). In chronic lupus erythematosus the lesions were discoid (FIG. 4) in 3 cases and vitiligoid in one case. The longitudinal melanonychia was showed in 3 cases on chronic lupus. Extracutaneous clinical signs associated with lupus were noted in 8 cases (88.9%). TABLE 1 shows the distribution of patients according to extracutaneous manifestations.

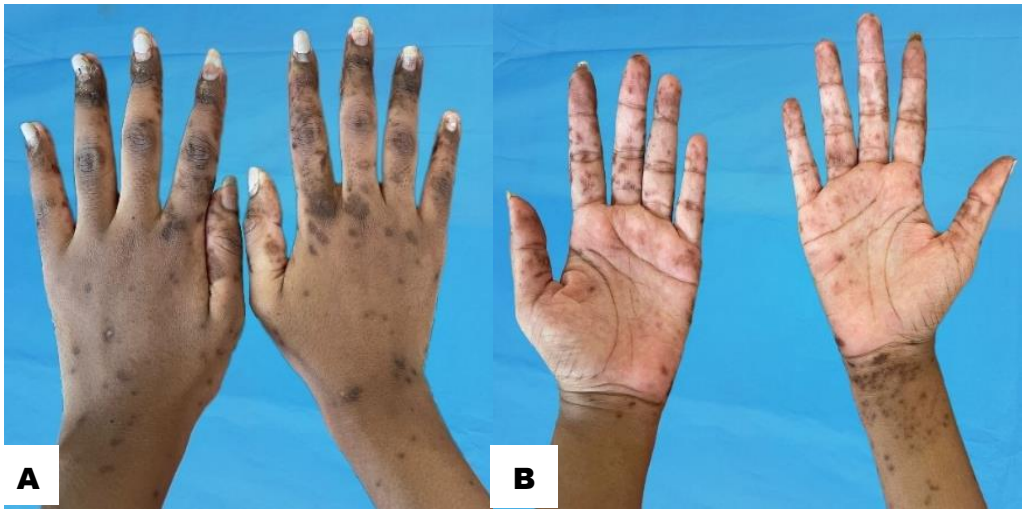

FIG. 2. **Acute lupus: Erythema interarticularis (A), palmar purpura (B).**

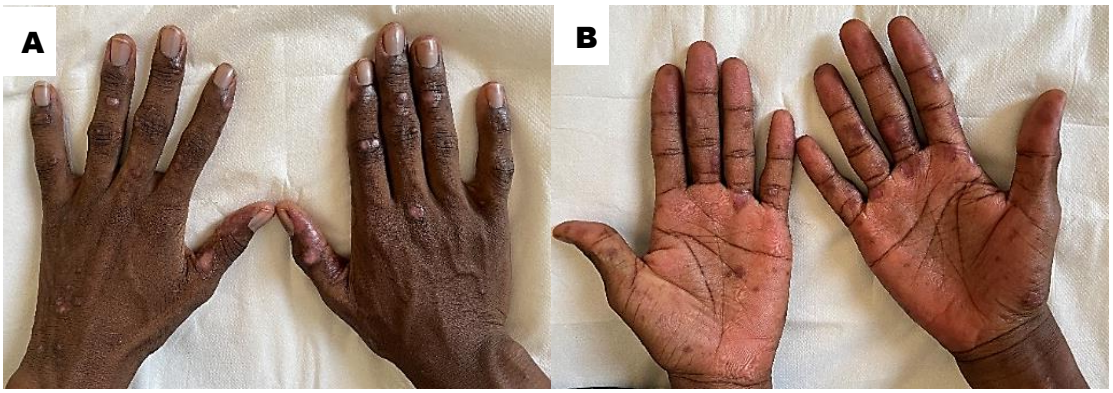

FIG. 3. **Subacute Lupus: Dorsal (A) and palmar (B) annular plates.**

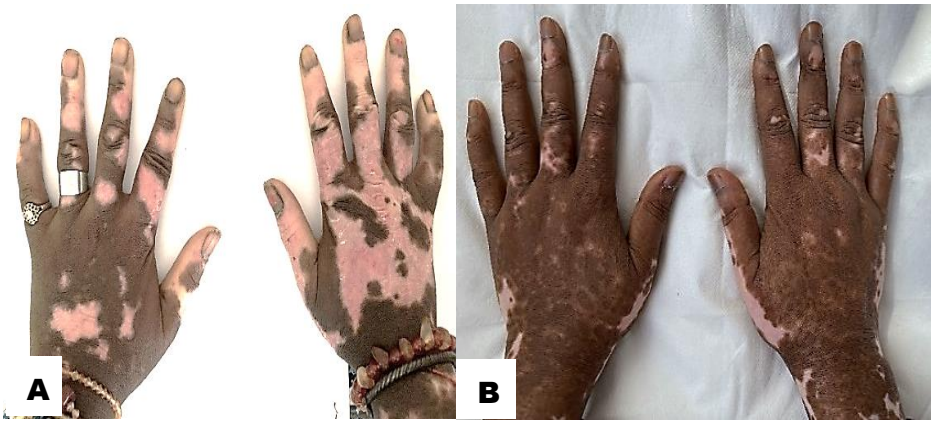

FIG. 4. **Chronic lupus: Discoid lupus (A) and vitiligoid lupus (B) of the hands.**

TABLE 1. **Distribution of patients according to extra-cutaneous manifestations of systemic lupus.**

Extra cutaneous signs	Number	Percentage %
Articular		
inflammatory polyarthritis	7	77,8
Pulmonary		
Pleurisy	1	11,1
Diffuse interstitial lung disease	3	33,4
Renal		
Lupus glomerulonephritis	4	44,4
Haematological		
Anemia	2	22,2
Leukopenia	1	11,1
Immunological		
Antinuclear antibody	1	11,1
Anti sm antibodies	1	11,1
Native anti-DNA antibody	1	11,1

In the case of systemic scleroderma, there were clinical signs on the hands in 17 cases. It involved 14 women and 3 men, i.e., a sex ratio of 0.2. The mean age was 39.2 years, range:15-71 years. Clinical signs were diffuse sclerosis in 13 cases (76.5%), sclerodactyly in 10 cases (58.8%) (FIG. 5)**,** mottled achromia in 8 cases (47.1%) (FIG. 6)**,** skin atrophy in 3 cases (17.6%) and calcinosis in 1 case (5.9%). Vascular manifestations were noted in 16 cases (94.1%). These were Raynaud's syndrome in 15 cases (88.2%) (FIG. 7), stellate pulpal scars in 8 cases (47.1%), digital ulcers in 4 cases (23.5%) and digital necrosis in 2 cases (11.8%) (FIG. 8). Nail manifestations included cuticle hypertrophy in 5 cases (29.4%). TABLE 2 shows the distribution of patients according to extracutaneous signs of systemic scleroderma.

FIG. 5. **Sclerodactyly with deforming arthritis.**

FIG. 6. **Speckled achromia in scleroderma.**

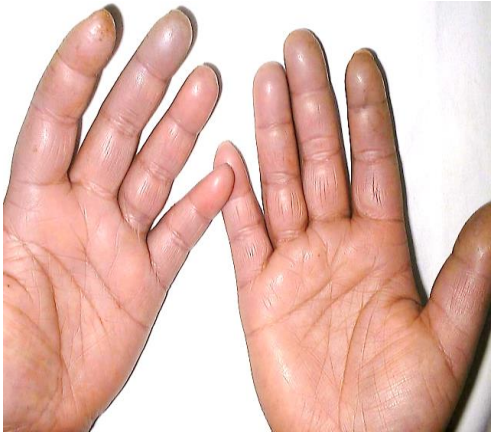

FIG. 7. **Raynaud's syndrome of the hands.**

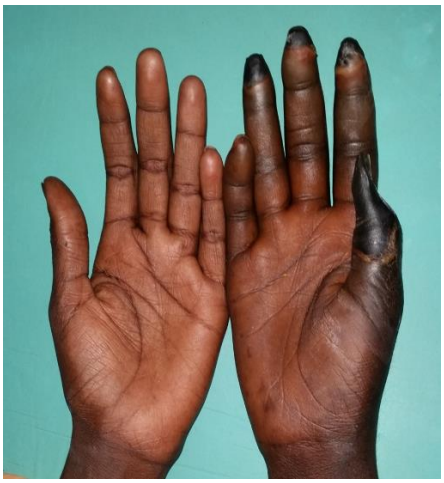

FIG. 8. **Digital necrosis of the hand.**

TABLE 2. **Distribution of patients according to extra cutaneous signs of systemic sclerosis.**

Extra cutaneous signs	Number	Percentage %
Digestive		
Gastroesophageal reflux	15	88,2
Dysphagia	6	35,3
Diarrhea	5	29,4
Constipation	4	23,5
Pulmonary		
Dyspnea	14	82,4
Chronic cough	6	35,3
Diffuse interstitial lung disease	9	52,9
Cardiac		
Arrhythmia	4	23,5
Pulmonary arterial hypertension	3	17,6
Heart failure	1	5,9
Articular		
Inflammatory arthritis	16	94,1
Immunological		
Antinuclear antibody	4	23,5
Anti-SCL70 antibody	5	29,4
Anti-SSA antibodies	3	17,6

Dermatomyositis concerned two women aged 27 and 33. One of the women presented with erythema in bands over the proximal metacarpophalangeal and proximal interphalangeal joints, papules of Gottron (FIG. 9) over the proximal interphalangeal joints. the other woman had nail crimp erythema and cuticle hypertrophy and Raynaud's syndrome.

FIG. 9. **Gottron's papules of the hand in dermatomyositis.**

The patients were presented with a myogenic syndrome and non-deforming peripheral polyarthritis in both cases. Diffuse interstitial lung disease with positive anti-MDA5 (i.e., melanoma differentiation-associated gene 5) antibodies was noted in only one patient. In the case of mixed connective tissue disease, an association between several systemic diseases was noted in 11 cases. Scleroderma was associated with dermatomyositis in 4 cases, systemic lupus erythematosus with dermatomyositis (FIG. 10) in 3 cases, systemic lupus erythematosus with scleroderma in 3 cases, and Sharp's syndrome with phospholipid antibody syndrome in one case. Nine women and two men were involved, i.e., a sex ratio of 0.2. The mean age was 37.4 years, range: 12- 65 years. TABLE 3 illustrates the various clinical signs of mixed connective tissue diseases. Deforming and reducible polyarthritis (Jaccoud's hand) was noted in 2 cases. Inflammatory anemia was noted in 2 cases (18,1%), and anti-nuclear antibodies were positive in 2 cases, anti-U1RNP (i.e., small nuclear ribonucleoprotein) antibodies in 3 cases, anti-Sm (i.e., Smith) antibodies in one case and anti-phospholipid antibodies in one case. In bivariate analysis, there was a significant association between pulpal scarring and cardiac rhythm disorders (p=0.043).

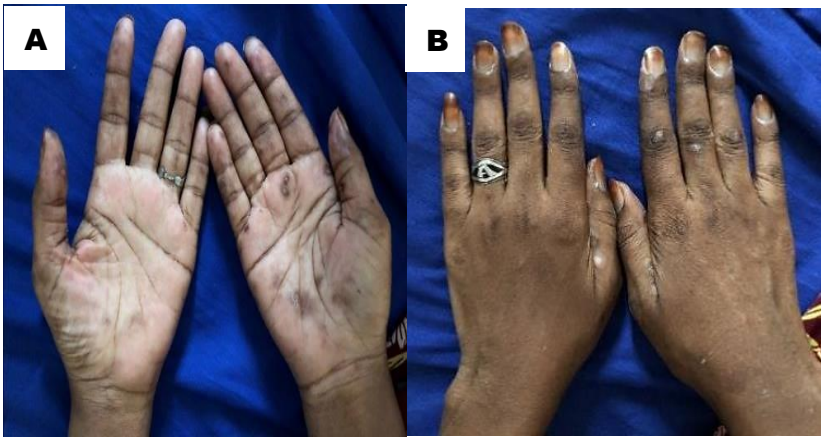

FIG. 10. **Mixed connective tissue disease associating lupus (A) and dermatomyositis (B).**

TABLE 3. **The different clinical signs of mixed connectivitis.**

Clinical Signs	Number	Percentage %
Cutaneous signs		
Cutaneous sclerosis	3	27,3
Sclerodactyly	1	9,1
Speckled Achromia	1	9,1
Gottron Papules	3	27,3
Erythema in transverse bands	1	9,1
Poikiloderma	1	9,1
Vascular signs		
Raynaud's Syndrome	3	27,3
Stellate pulp scars	2	18,2
Digital Ulcers	2	18,2
Nail signs		
Nail cuticle hypertrophy	2	18,2
Nail crimp erythema	3	27,3
Osteoarticular signs		
Finger edema	2	18,2
Muscle signs		
Muscle deficit	4	36,3
Respiratory signs		
Diffuse interstitial lung disease	2	18,2

4. Discussion

We report the first study in Senegal of cutaneous manifestations of autoimmune systemic diseases on the hands in Dermatology departments. The particularities of our study are the frequency of cutaneous symptoms of the hand in young women, the clinical polymorphism of cutaneous lesions with a predominance of vasculitis, and the association of cutaneous signs with severe visceral damage.

In systemic lupus erythematosus, cutaneous manifestations of the hands were polymorphous, with a predominance of acute and subacute lupus lesions. These lesions were associated with visceral manifestations of systemic lupus erythematosus in 8 cases. We noted only one patient with isolated discoid lupus lesions on the hands, with no extracutaneous damage. This could be explained by the diagnostic delay and the difficulties of access to specialized care in our countries. However, the literature notes a higher frequency of 78.2% of cutaneous lesions in chronic lupus [10]. Cutaneous vasculitis was predominant, often associated with visceral lupus damage. Yell [11] and Heimovski [12] reported a 50 to 60% frequency of Raynaud's syndrome of hand involvement in systemic diseases. Digital necrosis was frequently observed and was associated with anti-phospholipid antibody syndrome in our lupus patients. The prevalence of digital necrosis in lupus was estimated at 0.67% in a Chinese study [13]. It is a major prognostic factor in lupus, as it may be primary or secondary to antiphospholipid antibody syndrome. In

addition, digital necrotic lesions are prone to infectious by gram-negative bacilli or anaerobes [14]. We have noted specific signs of lupus on the hands, in the form of acute, subacute or chronic lesions. These lesions were sometimes isolated or associated in the same patient. Their semeiologic characteristics, in particular palmar erythema, interarticular lesions and discoid or vitiligoid plaques, suggest the diagnosis of lupus [10]. Lupus lesions on the hands were associated with lupus glomerulonephritis in 4 cases. Hence the need to screen lupus patients for lupus nephropathy.

Systemic scleroderma had the most clinical manifestations in the hands. Clinical symptoms were early-onset, disabling and often the reason for the discovery of scleroderma [15]. Edematous infiltration of the hands, Raynaud's syndrome and sclerodactyly were the clinical signs most frequently reported in the literature [15,16]. Vascular signs and symptoms were predominant in our study. Raynaud's syndrome was predominant in 88.2% of cases. Bouissar reported it in 98% and Meier in 96.3% [16,17]. Sclerosis of the hands was noted in 76.5%, associated with inflammatory polyarthritis deformans. Sclerodactyly is also one of the early clinical manifestations of scleroderma, with 86% of cases reported in the literature [15,16]. Stellate pulpal scars were present in 47.1%. In his study, Hughes reported the presence of digital scars in 50.9% of a cohort of 9671 scleroderma cases [18]. He also noted a statistically significant association between stellate pulpal scars and the occurrence of interstitial lung disease, rhythm disorders and dermal calcinosis ($p \leq 0.001$) [18].

We noted a frequency of 23.5% of digital ulcers during the course of scleroderma, as opposed to the literature, where a frequency of 16.9% was noted [19]. Ungual manifestations associated with scleroderma were present in 29.4% of cases, with hypertrophy of the nail cuticle. However, Marie noted a higher frequency in 80.6% of patients [20]. Two patients presented with clinical signs on the hands during the course of dermatomyositis. Raynaud's syndrome was more frequent on the hands, and Gottron's papules in our study were similar to those reported in the literature [21].

Nail involvement associated with dermatomyositis is rare. Authors often report periungual erythema in 60%, a manicure sign in 70% and dystrophic cuticles in 80% [22]. In mixed connective tissue disease scleroderma associated with dermatomyositis was more frequent in 36.3%. Deforming and reducible polyarthritis (Jaccoud's hand) was noted in 2 cases (18.2%). These joint symptoms are more common in lupus [23].

5. Conclusion

Clinical manifestations of the hands are various in the course of systemic autoimmune diseases. The semiological particularities of skin lesions on the hands could contribute to the diagnosis and early management of these systemic diseases in order to improve the prognosis of these chronic diseases in our african countries.

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