Scientific Research Publishing

Open Journal of Rheumatology and Autoimmune Diseases, 2025, 15(3), 118-125
https://www.scirp.org/journal/ojra
ISSN Online: 2164-005X
ISSN Print: 2163-9914

Severe Visceral Damage in Systemic Scleroderma in Abidjan (Ivory Coast)

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How to cite this paper: Sompougdou, C., Djaha Kouassi, J.M., Kaboré, F., Kenagnon, D.A.S., Rouamba/Toé, N., Diomandé, M. and Eti, E. (2025) Severe Visceral Damage in Systemic Scleroderma in Abidjan (Ivory Coast). *Open Journal of Rheumatology and Autoimmune Diseases*, **15**, 118-125.
https://doi.org/10.4236/ojra.2025.153014

Received: March 20, 2025
Accepted: August 1, 2025
Published: August 4, 2025

Abstract

Objective: Study of severe visceral damage in systemic scleroderma in the rheumatology Department of CHU de Cocody. **Patients and Methods:** We conducted a retrospective descriptive study of the records of patients hospitalised for systemic scleroderma according to ACR/EULAR 2013 criteria in the rheumatology Department of CHU de Cocody over an 11-year period from January 1, 2005 to December 31, 2016. **Results:** A total of 18 patients had systemic scleroderma, 10 of whom had severe visceral damage with a frequency of 55.5%. All patients were female. The mean age was 34.5 ± 15.4 years. The mean duration of the disease was 3.5 ± 4.1 years. Clinically, 7 patients (70%) had a dry cough, 5 (50%) had stage dyspnoea and 2 (20%) had palpitations. Serious damage was pulmonary, cardiac and digestive in 8 patients (80%), 4 patients (40%) and 3 patients (30%) respectively. Pulmonary damage was mainly a combination of pulmonary arterial hypertension and interstitial lung disease in 5 patients (62.5%). Cardiac damage was represented by pericarditis in 3 patients (75%). Gastro-oesophageal reflux disease occurred in 2 patients (66.7%). Biologically, anti-nuclear antibodies were positive in all patients, Anti-Scl-70 in 44.4% and anti-centromeres in 22.2%. The course was marked by 3 deaths due to cardiovascular failure. **Conclusion:** Severe visceral damage is common in systemic scleroderma, dominated by cardiac and pulmonary attacks.

Keywords

Severe Visceral Damage, Systemic Scleroderma, Abidjan, Sub-Saharan Africa

1. Introduction

Systemic sclerosis (SSc) is a generalized connective tissue disorder affecting arterioles and microvessels, characterized by the occurrence of tissue fibrosis and vascular obliteration [1]. Its exact prevalence remains poorly understood, with disparities across regions and countries. In France, the prevalence of SSc is 158 cases per million adults in a study conducted in 2004 in the Seine-Saint-Denis department, 132 cases per million adults in a study carried out in Lorraine and published in 2013, and 228 cases per million adults in a study published in 2016 conducted in Alsace, allowing for an extrapolation estimating the number of adult patients with SSc in France to be between 6000 and 9000 [2]-[4]. The prevalence in the United States is approximately 260 per million inhabitants [5]. In sub-Saharan Africa, a systematic review identified 1884 patients with systemic sclerosis in 17 of the 48 sub-Saharan African countries [6].

SSc is a systemic autoimmune disease that causes premature death due to complications from interstitial lung disease, pulmonary arterial hypertension, gastrointestinal dysmotility, renal crisis, and malnutrition [7]. The prognosis of SSc is highly variable, with mortality remaining significantly higher than that of the general population [8]. Mortality is 9% in sub-Saharan Africa, with cardiorespiratory complications being the primary causes of death [6] [9].

Regardless of the extent of skin involvement, SSc is associated with a significant risk of severe visceral involvement, particularly affecting the digestive, pulmonary, cardiac, or renal systems, which can be life-threatening [1]. Indeed, in a series of nearly 1000 patients seen between 1972 and 1995, severe renal, cardiac, pulmonary, and digestive involvement concerned 19%, 15%, 16%, and 8% of patients, respectively [10].

In sub-Saharan Africa, a systematic review on systemic sclerosis reported interstitial lung involvement in 50%, pulmonary hypertension in 30%, and heart involvement in 28% of patients, esophageal reflux was observed in 70% and dysphagia in 37% of patients [6]. In Kenya, Ilovi *et al.* noted severe pulmonary, cardiac, and renal involvement in 56%, 22%, and 4% of patients, respectively [11]. In South Africa, a recent study found interstitial lung disease and pulmonary arterial hypertension in 55% and 38.3% of patients, respectively; scleroderma renal crisis in 3.1%; and renal insufficiency in 16.7% [9].

In Ivory Coast, few studies have focused on severe visceral involvement in systemic sclerosis, hence the relevance of our study. The aim of our work was to identify severe visceral damage in systemic scleroderma in order to contribute to a better understanding of severe visceral damage in systemic scleroderma in the Ivory Coast.

2. Patients and Method

This was a retrospective descriptive study of data from January 1, 2005, to December 31, 2016, in the rheumatology department of the Cocody University Hospital

Center in Abidjan, Ivory Coast.

Patient records were reviewed and those fulfilling the American College of Rheumatology/European League against Rheumatism collaborative initiative 2013 criteria for systemic scleroderma were recruited into the study [12]. Patients with incomplete data (less than 75% completeness) were not included. Data were collected on a form including socio-demographic variables (age, sex), disease duration, clinical signs (cardiac, digestive, respiratory, renal, cutaneous, and rheumatological), and paraclinical findings (ECG, echocardiography, chest X-ray, chest CT scan, Antinuclear Antibodies, Anti-Scl-70, Anti-centromere, native DNA, Anti-U3RNP, Anti-PM Scl, Rheumatoid Factors, Anti-mitochondrial antibodies, Anti-RNA polymerase III), and treatment.

SSc was classified as limited or diffuse based on the extent of skin involvement according to the method described by LeRoy *et al.* [13].

Severe pulmonary involvement was defined by the presence of interstitial lung disease or pulmonary arterial hypertension, diagnosed by clinical findings and paraclinical examinations (pulmonary computed tomography, cardiac Doppler echocardiography).

Severe cardiac involvement was defined by the presence of pericarditis, rhythm and conduction disorders, diagnosed based on clinical signs, electrocardiogram, and Doppler echocardiography.

Severe digestive involvement was defined by the presence of gastroesophageal reflux with a weight loss of at least 10% of baseline weight, or primary biliary cirrhosis [14]. The diagnosis of primary biliary cirrhosis met the criteria of the American Association for the Study of Liver Diseases [15].

The confidentiality of the information was ensured by the anonymization of collection sheets. The study was conducted in accordance with the ethical recommendations of the Helsinki Declaration. The data were analyzed by the software "Epi Info" version 7.2.4.0.

3. Results

During the study period, 18 patients were collected, of whom 10 patients presented with severe visceral involvement, representing a frequency of 55.5%. All patients were female. The average age of the patients was 34.5 ± 15.4 years with extremes of 24 years to 69 years. The average disease duration was 3.5 ± 4.1 years. All patients had skin sclerosis, and cough was reported in seven patients. **Table 1** shows the distribution of patients according to functional signs. Diffuse cutaneous SSc was noted in seven patients, and limited cutaneous SSc in three patients. Pulmonary, cardiac, and digestive involvement were found in eight, four, and three patients, respectively. **Figure 1** shows the distribution of patients based on severe organ involvement. Pulmonary involvement was predominantly characterized by the combination of pulmonary arterial hypertension and interstitial lung disease in six cases. **Table 2** illustrates the distribution of patients according to severe visceral involvement.

Table 1. Distribution of patients according to functional signs.

Functional Signs	Number (N = 10)	Percentage
Skin sclerosis	10	100
Dry cough	7	70
Arthralgia/Arthritis	5	50
Dyspnoea	5	50
Dysphagia	3	30
Palpitations	2	20
Early satiety	2	20
Calcinosis	1	10
Regurgitation	1	10

*A patient may present multiple functional symptoms.

Table 2. Distribution of patients according to severe visceral damage.

Severe Visceral Damage	Total	Percentage
Lung involvement	N = 8	
Association PAH + ILD*	6	75
Pulmonary Arterial Hypertension	1	12.5
Interstitial lung disease	1	12.5
Heart involvement	N = 4	
Pericarditis + RD**	2	50
Atrioventricular block, 1st degree	1	25
Pericarditis + CCD***	1	25
Digestive involvement	N = 3	
Gastro oesophageal reflux disease	2	66.7
Primary biliary cirrhosis	1	33.3

*Pulmonary Arterial Hypertension associated with Interstitial lung disease; **Rhythm disorder (sinus tachycardia and supra ventricular tachycardia); ***Cardiac conduction disorder (complete right bundle branch block).

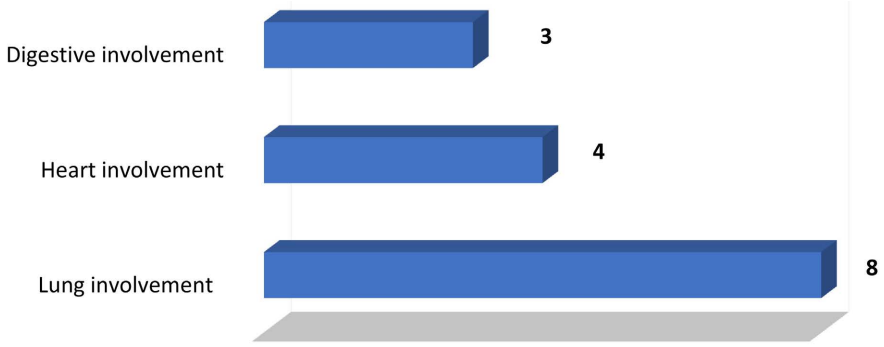

Figure 1. Distribution of patients according to organs affected.

Immunologically, antinuclear antibodies (ANA) were positive in all patients tested (9/9), and anti-topoisomerase I autoantibodies were present in 44.4% of patients (4/9). Anti-centromere and anti-mitochondrial autoantibodies were found in 22.2% (2/9) and 11.1% (1/9) of cases, respectively. Anti-fibrillarin autoantibodies were not detected in our study.

The administered treatment included corticosteroids in all patients, D-penicillamine in six patients, hydroxychloroquine in two patients, and cyclophosphamide in two patients. The clinical course was marked by the death of three patients due to cardiovascular failure.

4. Discussion

Over a 12-year period, we reported 18 cases of systemic sclerosis, of which 10 cases presented with severe visceral involvement. Consistent with our findings, SSc appears to be rare in African series [16]-[19]. Indeed, reported hospital-based series included 21 cases over 20 years in Benin, 14 cases over 5 years in Nigeria, 7 cases over 13.83 years in Burkina Faso, and 26 cases over 10 years in Senegal [16]-[19]. Our results are lower than those reported in Western studies [20]. These differences could be attributed to a scarcity of population-based studies in Africa, limited access to healthcare services for patients, diagnostic challenges, and reliance on traditional medicine.

A female predominance (100%) and a mean age of 34.5 years were observed. This result aligns with findings reported in both African and Western studies [6] [20].

The peak incidence is typically around the fifth decade of life; however, most studies have reported a younger age at diagnosis in Black patients compared to Caucasians [21] [22].

Diffuse cutaneous SSc was noted in seven patients, and limited cutaneous SSc in three patients during our study. Our data are corroborated by African and American studies [6] [22]. Indeed, diffuse cutaneous SSc is the predominant subset in sub-Saharan Africa, North Africans, and African Americans, whereas limited cutaneous SSc is more frequent in White populations [6] [22]-[24].

Severe visceral involvement was found in 10 patients, representing 55.5% of the cases. Our results were similar to those of African and American series [6] [9] [25]. Severe visceral involvement is more frequent in Black individuals than in Caucasians [6] [22]. Indeed, Steen *et al.* demonstrated that African Americans with SSc experience more severe complications than Caucasians with SSc, and this is related to both the type of autoantibody present and the severity of interstitial lung disease [22]. Furthermore, diffuse cutaneous SSc is more common in African patients and generally has a poorer prognosis than limited SSc, which is the dominant subset in Caucasians and has a more indolent course [6] [25]. This high prevalence of severe visceral involvement in the black population may in part be explained by genetic factors, economic factors and difficulties in accessing healthcare. Several studies have identified differences in genetic associations between Cauca-

sians and African Americans; HLA-DRB1*08 alleles are more frequent in African American patients with SSc than in healthy African Americans and Caucasian patients with SSc [22]. In Africa, only a handful of studies have been performed on genetic risk factors for SSc in Africans, and in the only study to date on HLA associations with SSc, HLA-DRB1*15:01 alleles were associated with SSc overall in South Africans, HLA-DQB1*03:01 alleles were associated with diffuse cutaneous SSc and HLA-DRB*11:01 alleles were associated with anti-U3RNP antibodies [26].

Severe visceral involvement was predominantly characterized by pulmonary involvement in 8 cases during our study. Pulmonary involvement included pulmonary arterial hypertension in seven cases (87.5%) and interstitial lung disease also in seven cases (87.5%). These same findings have been reported in African series [6] [9].

The immunological profile of severe visceral involvement in SSc observed in our study is consistent with those of South Africans and African Americans [9] [22]. However, our results show a higher prevalence of anti-topoisomerase I and anti-centromere antibodies compared to their data, where the prevalence in the South African study was 28.3% and 10.2%, respectively [9]. Anti-topoisomerase I antibodies were found in 26% and anti-centromere antibodies in 7% of African American patients [22]. This difference could be explained by our smaller sample size and the monocentric nature of our study.

5. Conclusion

Severe visceral involvement in systemic sclerosis is frequent in Abidjan. Pulmonary and cardiac involvement are the most common severe manifestations, which can be life-threatening, highlighting the necessity for their systematic investigation in systemic sclerosis. Population-based studies are needed to better assess the frequency of severe visceral involvement in systemic sclerosis in Ivory Coast.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Mouthon, L., Allanore, Y., Cabane, J. and Hachulla, E. (2008) Sclérodermie systémique. In: Guillevin, L., Meyer, O. and Sibilia, J., Eds., *Maladies et syndromes systémiques*, Flammarion Médecine-Sciences, 410-470.

[2] Le Guern, V. (2004) Prevalence of Systemic Sclerosis in a French Multi-Ethnic County. *Rheumatology*, **43**, 1129-1137. https://doi.org/10.1093/rheumatology/keh253

[3] El Adssi, H., Cirstea, D., Virion, J.M., Guillemin, F. and de Korwin, J.D. (2013) Estimating the Prevalence of Systemic Sclerosis in the Lorraine Region, France, by the Capture-Recapture Method. *Seminars in Arthritis and Rheumatism*, **42**, 530-538. https://doi.org/10.1016/j.semarthrit.2012.10.001

[4] Meyer, A., Chifflot, H., Chatelus, E., Kleinmann, J.F., Ronde-Ousteau, C., Klein, D.,

et al. (2016) Brief Report: Spatial Heterogeneity of Systemic Sclerosis in France: High Prevalence in the Northeast Region. *Arthritis & Rheumatology*, **68**, 1731-1737. https://doi.org/10.1002/art.39613

[5] Arnett, F.C., Cho, M., Chatterjee, S., Aguilar, M.B., Reveille, J.D. and Mayes, M.D. (2001) Familial Occurrence Frequencies and Relative Risks for Systemic Sclerosis (Scleroderma) in Three United States Cohorts. *Arthritis & Rheumatism*, **44**, 1359-1362. https://doi.org/10.1002/1529-0131(200106)44:6<1359::aid-art228>3.0.co;2-s

[6] Erzer, J.N., Jaeger, V.K., Tikly, M. and Walker, U.A. (2020) Systemic Sclerosis in Sub-Saharan Africa: A Systematic Review. *Pan African Medical Journal*, **22**, Article 176.

[7] Denton, C.P. and Khanna, D. (2017) Systemic Sclerosis. *The Lancet*, **390**, 1685-1699. https://doi.org/10.1016/s0140-6736(17)30933-9

[8] Rubio-Rivas, M., Royo, C., Simeón, C.P., Corbella, X. and Fonollosa, V. (2014) Mortality and Survival in Systemic Sclerosis: Systematic Review and Meta-Analysis. *Seminars in Arthritis and Rheumatism*, **44**, 208-219. https://doi.org/10.1016/j.semarthrit.2014.05.010

[9] Dire, Z., Ickinger, C. and Tikly, M. (2023) Causes and Predictors of Mortality in South Africans with Systemic Sclerosis. *Rheumatology & Autoimmunity*, **3**, 108-114. https://doi.org/10.1002/rai2.12080

[10] Steen, V.D. and Medsger Jr., T.A. (2000) Severe Organ Involvement in Systemic Sclerosis with Diffuse Scleroderma. *Arthritis & Rheumatology*, **43**, 2437-2444.

[11] Illovi, C. and Oyoo, G. (2013) Characteristics of Systemic Sclerosis Patients in Nairobi, Kenya: A Retrospective Study. *African Journal of Rheumatology*, **1**, 23-27.

[12] van den Hoogen, F., Khanna, D., Fransen, J., Johnson, S.R., Baron, M., Tyndall, A., *et al.* (2013) 2013 Classification Criteria for Systemic Sclerosis: An American College of Rheumatology/European League against Rheumatism Collaborative Initiative. *Arthritis & Rheumatism*, **65**, 2737-2747. https://doi.org/10.1002/art.38098

[13] LeRoy, E.C. and Medsger Jr., T.A. (2001) Criteria for the Classification of Early Systemic Sclerosis. *The Journal of Rheumatology*, **28**, 1573-1576.

[14] Hachulla, E., Launay, D., de Groote, P., Moranne, O., Tillie-Leblond, I., Seguy, D., *et al.* (2005) Les défaillances viscérales graves de la sclérodermie systémique. *Réanimation*, **14**, 576-586. https://doi.org/10.1016/j.reaurg.2005.09.019

[15] Lindor, K.D., Gershwin, E.M., Poupon, R., Kaplan, M., Bergasa, N.V. and Heathcote, J.E. (2009) Primary Biliary Cirrhosis. *Hepatology*, **50**, 291-308. https://doi.org/10.1002/hep.22906

[16] Teclessou, J.N., Saka, B., Akakpo, S.A., Matakloe, H., Mouhari-Toure, A., Kombate, K., *et al.* (2018) Les connectivites en milieu hospitalier à Lomé: Étude rétrospective de 231 cas. *Pan African Medical Journal*, **30**, Article 176. https://doi.org/10.11604/pamj.2018.30.176.14565

[17] Adelowo, O.O. and Oguntona, S. (2009) Scleroderma (Systemic Sclerosis) among Nigerians. *Clinical Rheumatology*, **28**, 1121-1125. https://doi.org/10.1007/s10067-009-1191-2

[18] Kane, B.S., Ndongo, S., Ndiaye, A.A., Djiba, B., Niasse, M., Diack, N., *et al.* (2016) Maladies systémiques en médecine interne "contexte Africain": Aspects épidémiologiques et classification. *La Revue de Médecine Interne*, **37**, A37. https://doi.org/10.1016/j.revmed.2016.04.237

[19] Tiendrébéogo, J.W.S., Kaboré, F., Sougué, C., Sankara, V., Zongo, E., Savadogo, B., *et al.* (2022) Epidemiology of Rheumatic Diseases: A Cohort of 23,550 Patients in Rheumatology Clinics in Burkina Faso. *Clinical Rheumatology*, **42**, 371-376.

https://doi.org/10.1007/s10067-022-06470-y

[20] Chifflot, H., Fautrel, B., Sordet, C., Chatelus, E. and Sibilia, J. (2008) Incidence and Prevalence of Systemic Sclerosis: A Systematic Literature Review. *Seminars in Arthritis and Rheumatism*, **37**, 223-235. https://doi.org/10.1016/j.semarthrit.2007.05.003

[21] Laing, T.J., Gillespie, B.W., Toth, M.B., Mayes, M.D., Gallavan, R.H., Burns, C.J., *et al.* (1997) Racial Differences in Scleroderma among Women in Michigan. *Arthritis & Rheumatism*, **40**, 734-742. https://doi.org/10.1002/art.1780400421

[22] Steen, V., Domsic, R.T., Lucas, M., Fertig, N. and Medsger, Jr. (2012) A Clinical and Serologic Comparison of African American and Caucasian Patients with Systemic Sclerosis. *Arthritis & Rheumatism*, **64**, 2986-2994. https://doi.org/10.1002/art.34482

[23] Yacoub, Y.I., Amine, B., Laatiris, A., Wafki, F., Znat, F. and Hajjaj-Hassouni, N. (2012) Bone Density in Moroccan Women with Systemic Scleroderma and Its Relationships with Disease-Related Parameters and Vitamin D Status. *Rheumatology International*, **32**, 3143-3148. https://doi.org/10.1007/s00296-011-2150-1

[24] Ben Salah, R., Frikha, F., Hachicha, H., Chabchoub, I., Dammak, C., Snoussi, M., *et al.* (2019) Clinical and Serological Profile of Systemic Sclerosis in Tunisia: A Retrospective Observational Study. *La Presse Médicale*, **48**, e284-e291. https://doi.org/10.1016/j.lpm.2019.07.037

[25] Yen, E.Y., Singh, D.R. and Singh, R.R. (2021) Trends in Systemic Sclerosis Mortality over Forty-Eight Years, 1968-2015: A US Population-Based Study. *Arthritis Care & Research*, **73**, 1502-1510. https://doi.org/10.1002/acr.24411

[26] Tikly, M., Rands, A., McHugh, N., Wordsworth, P. and Welsh, K. (2004) Human Leukocyte Antigen Class II Associations with Systemic Sclerosis in South Africans. *Tissue Antigens*, **63**, 487-490. https://doi.org/10.1111/j.0001-2815.2004.00199.x