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CASE REPORT

Scleroderma (systemic sclerosis) among Nigerians

O. O. Adelowo · S. Oguntona

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Abstract Scleroderma and other connective tissue diseases have rarely been reported among Africans. The objective of this paper is to have a retrospective study of the clinical and investigative characteristics of scleroderma patients seen in a rheumatology clinic. This was done in a private practice rheumatology clinic in Lagos, Nigeria. Patients were identified using the American College of Rheumatology criteria for diagnosis of scleroderma. A total of 14 cases of scleroderma are reported. Most of the patients were females and diffuse scleroderma was more frequently seen. Arthritis and reflux esophagitis were the most common nondermatological presentation while Raynaud's phenomenon and dysphagia were the least seen. Restrictive pattern of lung function tests were seen in most of those tested and pulmonary fibrosis was seen in some cases. Antinuclear antibodies were the commonest serological findings with the speckled staining pattern in most cases. Treatments were with standard medications. Scleroderma among Nigerians is rare as elsewhere and there are certain common characteristics as seen elsewhere as well as certain differences.

Keywords Clinical features · Nigerians · Scleroderma

O. O. Adelowo · S. Oguntona
Rheumatology Unit, Department of Medicine,
Olabisi Onabanjo University Teaching Hospital,
Sagamu, Ogun State, Nigeria

O. O. Adelowo (✉)
P. O. Box 7231, Ikeja,
Lagos, Nigeria
e-mail: femiadelowo2003@yahoo.com

Introduction

Scleroderma (systemic sclerosis) is a rare systemic autoimmune connective tissue disease of unknown etiology. The pathogenetic mechanisms of the disease include immune dysregulation, endothelial dysfunction, and excessive fibrous tissue deposits in the skin and internal organs. The incidence varies among populations with a report of 3.7 million per year in UK and Iceland, whereas, USA has an incidence of 18.7–22.8 million per year [1–3].

Most of the studies have shown some ethnic variation with a higher frequency of diffuse subset among African Americans [3, 4]. There is also evidence that it runs a more aggressive course in this group [5].

The higher incidence of systemic sclerosis among blacks have been attributed to the postulation that in this ethnic group certain connective tissue responses which are involved in protection against infection and repair after injury may also predispose to certain chronic diseases [6].

Scleroderma has rarely been reported among black Africans and these have mostly been single-case reports [7–9]. About the largest case series was from South Africa in a case study of 63 blacks living in a gold mine region [10].

In view of the rarity of reports on scleroderma among African blacks, this study looked at the clinical and investigative characteristics of this condition among Nigerian subjects presenting with scleroderma.

Materials and methods

This study is a case study series of scleroderma cases seen among patients attending a private practice rheumatology clinic. This was a retrospective study of cases of scleroder-

Table 1 Demography of scleroderma patients

Female	12 (85.7%)
Male	2 (14.3%)
Age range	26–69
Mean	40.3 years

ma seen in a rheumatology clinic over a 5-year period from January 2002 to December 2006. The diagnosis was based on the American College of Rheumatology criteria for scleroderma [10]. The cases reported formed part of a retrospective study of all rheumatology cases seen in this clinic within the stated period.

Laboratory, serology, and pulmonary function tests and radiographs were reviewed where available.

Results

A total of 14 cases were seen (F-12; M-2) out of the total of 1,240 rheumatology cases during the period under study, thus constituting 1.1% of all cases seen. The demography is shown in Table 1. Figures 1, 2, 3, 4, and 5 show the clinical features in some of the subjects.

Most of subjects had diffuse skin presentation as shown in Table 2. The commonest presentation was the "salt and pepper" appearance of alternating hypopigmentations and hyperpigmentations. The commonest systemic presentation was in the musculoskeletal system with arthritis while the least common presentations were Raynaud's phenomenon and dysphagia (Table 3).

Pulmonary function tests of force expiratory volume and forced vital capacity were available in six cases with restrictive pattern present in four of those tested (66.7%). Chest radiographic investigations were available in seven

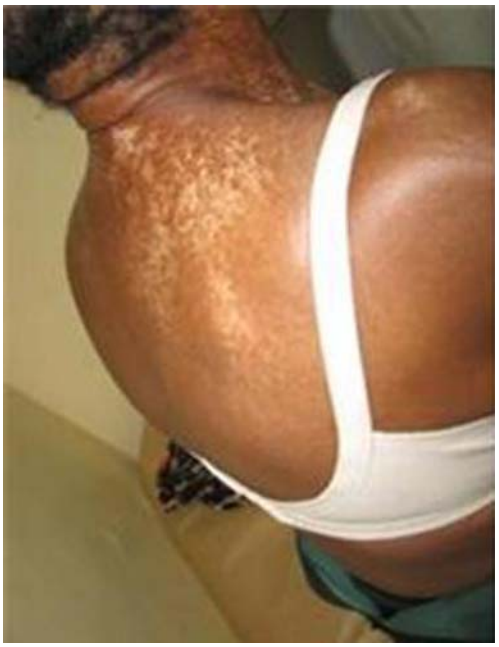

Fig. 2 Salt and pepper appearance over the back

Fig. 1 Diffuse hypopigmentation and hyperpigmentation in the neck and face (salt and pepper appearance)

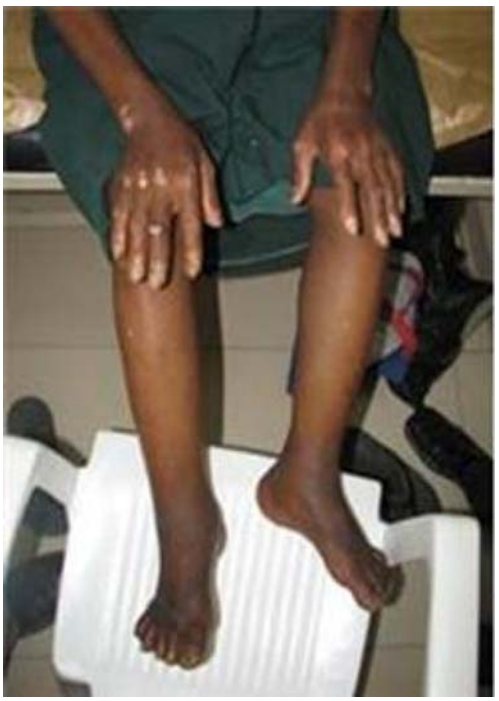

Fig. 3 Hypopigmentation on the hands, shin, and hyperpigmentation over the feet

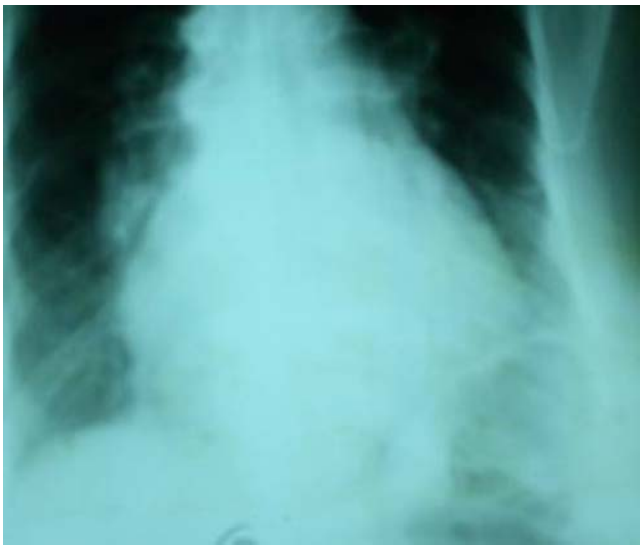

Fig. 4 Chest X-ray showing reticular shadows of pulmonary fibrosis in a 70-year-old patient with sine scleroderma

subjects with three showing basal pulmonary fibrosis (Fig. 4).

There were no hematological or biochemical abnormalities, though erythrocyte sedimentation rate was elevated in 11 subjects with a range of 49–76 mm/h.

Electrocardiography done in six patients showed abnormality in four cases (66.7%) with sinus tachycardia in two cases while one had premature supraventricular complexes, left axis deviation, and left ventricular hypertrophy. One patient had sinus bradycardia.

Serological findings are as shown on Table 4 with most of those tested showing significant antinuclear antibody positivity, mostly speckled staining pattern.

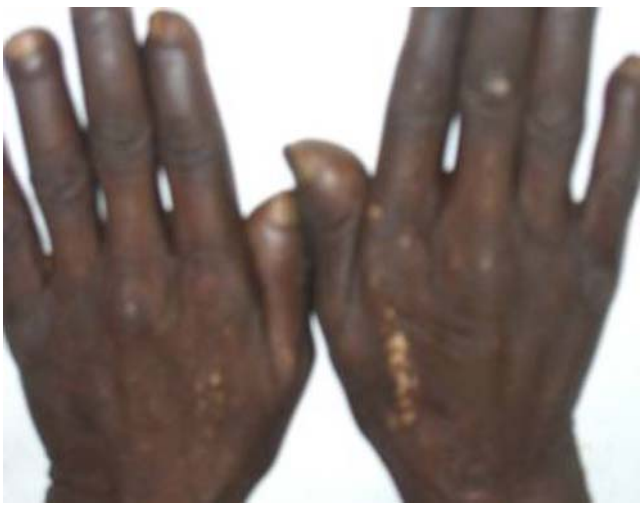

Fig. 5 Scleroderma hands showing absorption of terminal tufts of left index and ring fingers as well as both thumbs

Table 2 Subsets of scleroderma (%)

Diffuse	8 (57.1%)
Limited	3 (21.4)
Undifferentiated	2 (14.3)
Sine scleroderma	1 (7.2)

Treatment

All the patients were placed on D-penicillamine at doses ranging between 250 and 750 mg daily. They all had proton pump inhibitors, omeprazole 20–40 mg daily as well as lisinopril 2.5–5 mg daily. Subjects also had symptomatic treatment with nonsteroidal anti-inflammatory drugs though corticosteroid was not administered in any of the patients.

Outcome

Five patients are known to have died, though causes of death were not known. Three were lost to follow-up but six are still being followed up though on an irregular basis.

Discussion

Scleroderma is a rare disease and this is confirmed in this report, being seen in 1.1% of all the cases seen in a rheumatology clinic. It is possible however that some of these cases are also seen in dermatology clinics but literature search in Nigeria has only highlighted two single-case reports [7, 8]. This possibly confirms the rarity of these cases among Nigerians. According to Medsger [1], if these cases were not so rare, there should be new cases of scleroderma at the rate of 18–20 million per year. The higher frequency of diffuse scleroderma among blacks is again confirmed, being present in eight cases out of the 14 seen. Limited scleroderma was seen in only three cases.

Table 3 Non dermatological presentation among 14 scleroderma patients

	Clinical presentation	Number	Frequency (%)
1	Arthritis	10	71.4
2	Reflux esophagitis	8	57.1
3	Cough/dyspnea	7	50
4	Hypertension	6	42.9
5	Constipation	3	21.4
6	Dysphagia	2	14.3
7	Raynaud's phenomenon	2	14.3

Table 4 Serological findings in nine patients

Serology	Number
Rheumatoid factor	2/9
ANA	Range(1:80–1:1,280)
Speckled	5/9
Nucleolar	3/9
Homogeneous	1/9
Anti Scl-70	1/9
Anti RNP	1/9
RO/SSA	1/9
IgG ACA	1/9

Sine scleroderma is rarely reported though it was seen in one of our cases. It was however not reported in the largest black African series of Tager and Tikly [11]. The only patient seen in our study was a 69-year-old woman who had extensive pulmonary fibrosis and antitopoisomerase antibodies but did not have any skin manifestation. She eventually died from a combination of pulmonary failure and Cor pulmonale.

The female preponderance seen in other reports is once again confirmed as well as the younger age of presentation among blacks. The mean age in our series is 40.3 years as against 36.1 years in the series of Tikly [11] and 51.5 years among Caucasians [4].

Raynaud's phenomenon is rarely reported among Black Africans and was seen in only two cases in our series; one with undifferentiated connective tissue disease and the other with diffuse scleroderma. This is in contrast to reports of identical frequencies between white and black subjects in other population studies [12–14].

Impaired pulmonary function tests were positive in six of the 11 tested. This is in consonance with reports of high frequency among American blacks [15]

The high frequency of ANA positivity is again confirmed, with most of the patients having a speckled staining pattern. This is in consonance with previous reports among scleroderma subjects [16, 17]. Antinucleolar staining pattern was seen in three out of nine tested, which is similar to the frequency of 35% reported among African Americans. Antitopoisomerase has been said to be rare among African Americans and is thought to be mostly associated with pulmonary disease [18].

Anticentromere antibodies are rarely seen among blacks [16] though this was not present in any of our patients.

Scleroderma is a rare disease and it may even be rarer among Black Africans. The relative high frequency reported from South African blacks may be related to their proximity to gold mines and exposure to dust.

There are limitations with this study which include the relative lack of investigation facilities, especially radiographic and serology. Another limitation is the usual difficulty in follow-up of patients in this part of the world. This is particularly so with all chronic diseases for which patients often default because of lack of obvious clinical improvements. It may be difficult in such situation to determine the cause of deaths or even the morbidities among such patients.

It is hoped that further studies can be carried out among Nigerians on this rare disease.

Disclosures None.

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