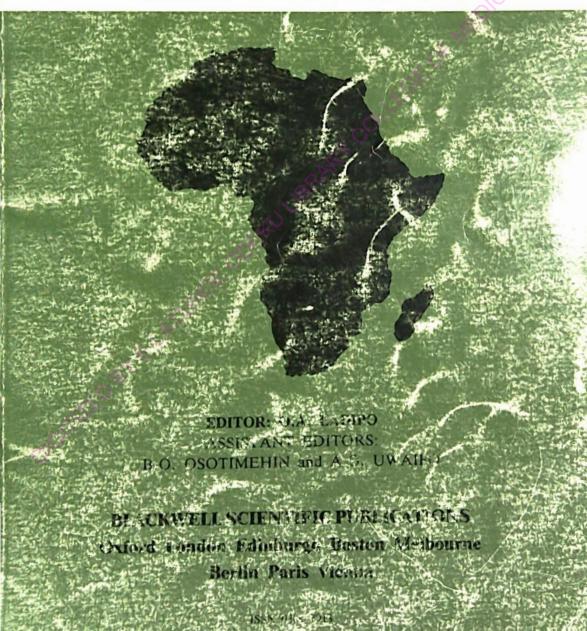
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counts of each patient, as well as the social status.

Detailed medical examination was carried out on all patients who were still attending the clinic at the time of this study. The following investigations were done when necessary: cultures of the wound swabs, X-ray of the affected limb(s), and blood cultures.

The diagnosis of sickle cell disease was by standard methods [10].

Results

There were 872 patients: 630 HbSS and 242 HbSC. The youngest was 14 years old and the eldest 60 years for HbSS and 58 years for HbSC.

A prevalence of 7.5% was obtained for HbSS with a male:female ratio of 2:1. Only 1.7% of HbSC patients had leg ulcers and three of the four were females. The mean age of HbSS patients with leg ulcers was 22 ± 3.9 years. HbSC patients were aged 25.8 ± 6.4 years. The mean haematocrit of 0.24 ± 0.03 I/I and reticulocyte counts of $8 \pm 3.5\%$ were no different from those of HbSS with no lesions. Similar haematocrit levels of 0.30 ± 0.03 I/I and reticulocyte counts of $3.3 \pm 2.3\%$ were obtained for HbSC patients with ulcers and those without.

Table 1 shows the ages at onset of lesions while Table 2 shows the duration of the ulcers in both HbSS and HbSC patients.

The sites of the ulcers were around the ankles in 75% of HbSS and in two of the HbSC patients (Table 3). The ulcers were bilateral in 17% of HbSS patients but none of the HbSC had bilateral lesions. Active ulcers, often discharging pus, varied in size from 1 cm to

 Table 1. Age of onset of leg ulcers in HbSS and HbSC patients

| Age range (years) | HbSS | HbSC | |
|-------------------|------|------|--|
| 10-14 | 14 | _ | |
| 15-19 | 22 | 1 | |
| > 20 | 8 | 2 | |
| Not known | 3 | 1 | |
| Total | 47 | 4 | |

Table 2. Duration of ulcers

| Years | HbSS | HbSC |
|-------|------|------|
| < 1 | 3 | 1 |
| 1-3 | 15 | - |
| 4-6 | 14 | 2 |
| 7-9 | 5 | _ |
| 10-12 | 4 | _ |
| > 12 | 3 | 1 |
| Total | 44 | 3 |

Four patients (three HbSS and one HbSC) could not remember how long they had had their ulcers.

Seven patients (five HbSS and two HbSC) defaulted with active ulcers.

12 cm² and were roundish in shape. Seven HbSS patients had gross superficial infection with microbial organisms, including *Pseudomonas*, *Proteus mirabilis*, *Klebsiella* species, anaerobic *Streptococcus* and *Staphylococcus aureus*. The organisms were grown at different times and multiple agents were cultured from some patients at the same time. Only one HbSS patient had an associated chronic osteomyelitis of the left tibia and left medial malleolus. Three HbSS patients had tetanus infection; they all survived.

Table 4 summarizes the treatment and results. Recurrence was very high irrespective of the therapy employed. Skin graft was not helpful. Complete healing occurred in only two patients (one HbSS and one HbSC), whereas 63% of the HbSS patients had relapsed at the time of writing.

Discussion

The very low incidence of leg ulcers of 1.7% in HbSC patients in this study is consistent with observations from other parts of the world [2– 4]. The higher haematocrit (and hence increased oxygen-carrying capacity of the red cells) and the relatively lower haemolysis in these patients may contribute to the rarity of chronic leg ulcers in them. Therapeutic reduction of chronic haemolysis through a hypertransfusion regimen (i.e. keeping the haematocrit above 0.35 I/1 and reticulocytes

Chronic leg ulcers in sickle cell disease: experience in Ibadan, Nigeria

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Summary

The prevalence of chronic leg ulcers was investigated in 872 adults with sickle cell disease (SCD) (630 HbSS and 242 HbSC) at Ibadan, Nigeria. The incidence was 7.5% in HbSS and 1.7% in HbSC patients. The sex ratio in HbSS was 2:1 in favour of males, and three of the four HbSC were females. Ulcers were sited around the ankles in more than 70% of the patients. The duration of the ulcers varied from less than 1 year to more than 20 years. There was no bias for social class. Response to therapy, including autologous skin graft, was poor.

Résumé

Nous avons étudié, à Ibadan (Nigéria), l'incidence des plaies chroniques sur la jambe chez 872 adultes (630 HbSS et 242 HbSC) atteints de drépanocytose. La fréquence était de 7.5% chez les HbSS et 1.7% pour les HbSC. Chez les malades HbSS, la distribution selon le sexe était de deux hommes pour chaque femme tandis que, chez les HbSC, les trois-quarts étaient des femmes. Les plaies surgissaient au niveau de la cheville chez 70% des malades. La durée de ces plaies variait de moins d'un an à plus de vingt ans. Rien n'a permis de faire une répartition sociale. Nous avons relevé une réaction insuffisante à la thérapie y compris l'autogreffe de la peau.

Introduction

Chronic leg ulceration is one of the most emotionally crippling complications of sickle

*Correspondence: M. A. Durosinmi, Department of Haematology and Immunology, Faculty of Health Sciences, Obafemi Awolowo University, Ile-Ife, Nigeria. cell disease (SCD). The lesions commonly appear between the ages of 10 and 50 years [1]. The ulcers are more commonly seen in adult males with homozygous sickle cell disease (HbSS) and are unusual in patients with sickle cell haemoglobin-C disease (HbSC) as well as those with sickle cell beta⁺-thalassaemia [1–4]. Multiple factors are responsible for sickle ulcers, including sloughing of necrotic tissues from areas affected by local infarcts, chronic haemolysis, stasis, thrombosis and to some extent trauma [1,2]. The lesions are almost always located around the ankles and lower tibia [1–8].

Sickle cell ulcers are very common amongst North American and Jamaican patients in whom prevalences of 73.6% and 75% have been reported respectively [6–8]. However, reports from Africa revealed much lower incidence, varying from only 5.4% to 9.6% [4,5,9].

There has been no previous report of chronic leg ulcers in Nigerians with HbSC disease [5,9]. The present report is based on our experience of chronic leg ulceration in HbSC patients at Ibadan, Nigeria. We also reviewed the situation in HbSS patients and compared the results with the previous reports from Africa [4,5,9].

Materials and methods

The case files of 872 adults with confirmed SCD, attending the outpatient clinic of the Haematology Department, University College Hospital, Ibadan, Nigeria, from January 1958 up till December 1985, were reviewed. The notes were reviewed for Hb genotype, sex, age, age at which ulcers developed, duration of ulcer, and modes and outcome of treatment. Other points included the location of the ulcer, the steady state haematocrit, and reticulocyte

blacks with SCD in Jamaica and North America 'emigrated' from Africa.

The sites of ulcers (Table 3) are largely confined to the ankles as previously documented [5,8]. This study confirmed the higher incidence of leg ulcers in HbSS males compared with the females as shown previously [5,7]. Sex ratio in HbSC patients would not be reliable in this study because of the very small numbers involved.

Leg ulcers in these patients are very resistant to therapy, including skin graft. This is consistent with previous reports [5–8]. Recurrence rate remains very high (Table 4). There is a need to formulate a workable treatment package for this environment, the most practicable being wound toileting with EUSOL. The use of local antibiotics may also be helpful especially where superficial infections are evident. All patients should have tetanus toxoid and antitetanus serum (ATS) as a protection. The use of amnion graft may be explored since autologous skin graft has not prevented recurrence [8].

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Leg ulcers in SCD

| | Haemoglobin genotype | | |
|-----------------------------------|----------------------|-----------------|--|
| Location | SS | S+C | |
| Ankle(s) | 35 | 2 | |
| Left lateral malleolus | 6 | | |
| Left medial malleolus | 7 | 2 | |
| Left lateral and medial malleoli | 1 | _ | |
| Right lateral malleolus | 4 | — | |
| Right medial malleolus | 8 | | |
| Right lateral and medial malleoli | 1 | _ | |
| Both ankles | 8 | — | |
| Legs | 13 | 1 | |
| Left leg | 7 | _ (| |
| Right leg | 4 | 14. | |
| Both legs | 2 | , ev | |
| Dorsum of feet | 1 | 1 | |
| Right foot | -0 |)~ ₁ | |

Table 3. Sites of ulcers

Table 4. Chronic leg ulcer - treatments and results

| Treatment | HbSS | HbS+C | Result |
|--|------|-------|---|
| Eusol | 38 | 4 | Not encouraging |
| Eusol and Solcoseryl | 4 | _ | No appreciable response |
| Solcoseryl jelly and Solcoseryl injection | 1 | _ | Recurred within 2 months |
| Autologous skin graft | 2 | _ | Failed |
| Admitted for bed rest | 2 | _ | Healed then recurred soon after discharge |
| Eusol + $ZnSO_4$ | 2 | — | No appreciable response |
| Sofratule | 2 | - | No appreciable response |

below 3%) has been found helpful in some HbSS patients with persistent leg ulcers [1,2,8]. Although only four HbSC patients were studied in this report, the chronicity of the ulcers was not in doubt (Table 2); only one patient had his ulcer healed within a few months and two were lost to follow-up having carried their ulcers for 4-6 years. The prevalence of leg ulcers in HbSS patients in this study remains low, as reported previously in Africans [4,5,9]. This is in contrast to the higher prevalence of more than 60% seen in North American and Jamaican patients [6–8]. The disparity is most likely due to environmental factors, including possibly nutrition and climate [5,11,12]. It is worth noting that the