Original Article

Hansen's disease: The need for increased healthcare provider awareness and re-training

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ABSTRACT
CONTEXT: Leprosy continues to be a challenge worldwide. In 2009, the World Health Organisation (WHO) reported 244,796 new cases worldwide. Of this, South East Asia, South Americas and Africa have the highest number of new cases.
OBJECTIVE: To highlight misdiagnosis of Hansen's disease as a drawback to eradication of leprosy.
MATERIALS: A retrospective study of cases of Hansen's disease misdiagnosed by clinicians who were seen in the skin clinics of two tertiary institutions in South-East Nigeria over a one year period.
RESULTS: Of the nine misdiagnosed cases, three were females while six were males. Apart from one child of six years old, the rest of the patients were adults. Referring diagnoses included chronic osteomyelitis, acromegaly, dermatitis and depression. One of the patients presented with a reversal reaction (erythema nodosum leprosum) following treatment in a peripheral health facility. Confirmation of the diagnosis was based on slit skin smears for Acid Fast Bacilli carried out in the side laboratory of skin clinic UNTH, Ituku-Ozalla, Enugu state (four patients) and mile four hospital, Abakaliki, Ebonyi State(three patients). All the smears were positive. Patients were commenced on multi drug therapy and all showed marked clinical improvement following drug therapy.
CONCLUSION: Eradication of leprosy may not be feasible if health care providers are ignorant of the different presentations of the disease. A high index of suspicion for leprosy is needed among health care workers in endemic areas. Development of tools for early diagnosis and detection of infection, improvement in existing tools for data collection, provision of guidelines and training materials on vital information for leprosy control will all contribute to the continued success of the National Tuberculosis and Leprosy Control Programme in Nigeria.
KEYWORDS: Hansen's disease, leprosy, early detection, drug therapy

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INTRODUCTION

Leprosy is a chronic infection caused by the acid-fast bacillus, *Mycobacterium leprae*. The organism which was discovered by Armauer Hansen, a Norwegian leprologist, in 1873 [1] is slow growing, showing preference for cooler areas of the body. *M. leprae* cannot be grown *in vitro* but has been cultured in the foot pad of mice and the nine-banded armadillo [2]. Mode of infection is thought to be from inhalation of bacilli-laden nasal droplets and contact with damaged skin. The disease affects the skin, peripheral nerves and mucosa of the upper respiratory tract primarily. The peripheral nerves most often affected are the ulnar (elbow), median (wrist), radial cutaneous (wrist), common peroneal (knee), posterior tibial and sural (ankle) [2]. Others include the great auricular and facial nerves.

Depending on the host's response to the organism, leprosy shows a wide range of clinical presentations from tuberculoid through borderline forms to lepromatous (Fig. 1). Individuals with a strong cellular immune response to *M. leprae* manifest with the tuberculoid (paucibacillary) form of the disease. These individuals have few skin lesions that tend to be dry and hypoaesthetic with asymmetrical neural involvement. Those with poor cellular immune response manifest with the lepromatous form. Disability, as a result of impairment of nerve function is the main outcome of leprosy and is affected by the type of leprosy and delay in diagnosis [3].

Leprosy classification according to the Ridley and Joplin criteria[4] is based on clinical, immunological and pathological grounds. In 1982, the world health organization [WHO] recommended another criteria[5] largely for operational purposes, to simplify disease recognition and to ensure that patients receive multi drug therapy [MDT] appropriately [6]. Since then, the WHO classification has been modified several times. A positive skin smear at any site was adequate for a diagnosis of multibacillary leprosy (MB) [7] but currently, classification of MB includes anyone with 6 or more skin lesions [8].

Diagnosis is usually made based on clinical symptoms and signs, supported by acid-fast bacilli in slit skin smears and/or typical histology on skin biopsy. Measures aimed at eradicating leprosy include availability and accessibility of control activities. These involve diagnosis, treatment with multidrug therapy, patient and family counselling, and community education and prevention of disabilities. Other aspects are rehabilitation and referral for complication[9]. Evolving research interests are in the areas of immunoprophylaxis and chemoprophylaxis[10].

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Chemotherapy of leprosy consisted of dapsone, usually administered as 100mg monotherapy, until the 1980's.Treatment for paucibacillary type was for 5 years while patients with multibacillary leprosy received therapy for life[11]. This soon led to emergence of dapsone resistant strains of the bacteria due to the slow action and weak bactericidal activity of the drug. With the emergence of the multidrug therapy (MDT) comprising rifampicin, clofazimine and dapsone in 1981, a dramatic improvement in

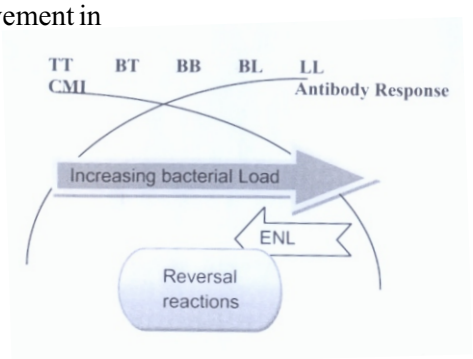

Fig. 1: Clinical-immunological spectrum of leprosy. ENL, erythema nodosum leprosum; CMI, cell mediated immunity; TT, tuberculoid leprosy; BT, borderline tuberculoid leprosy; BB, mid-borderline leprosy; BL, borderline lepromatous leprosy; LL, lepromatous leprosy.

METHODOLOGY

Study background

The study was carried out in the University of Nigeria Teaching Hospital, Ituku Ozalla and the Federal Medical Centre, Abakaliki (now Federal Teaching Hospital, Abakaliki), both tertiary institutions for health care in Enugu State and Ebonyi State respectively.

Study Population

This included all the patients who presented to the skin clinic of the above hospitals for treatment between January 2010 and December 2010. Children were defined as patients between the ages of 0 and 16years.

Study Design

This was a descriptive study that reviewed retrospective data from clinic registers and patient case files.

Methods

Patients' clinic registers were analysed retrospectively. Total number of consecutive new patients presenting in both study sites during the review period were documented from these clinic registers. The patients who were diagnosed with Hansen's disease in the skin clinics by the dermatologists had their folders retrieved. By analysing the case files, the originating referral clinics and diagnoses were determined. Diagnosis of leprosy was based on clinical findings of suspect skin lesions with or without loss of sensation, enlarged peripheral nerves and a positive slit skin smear for acid-fast bacilli.

Results

A total number of 1,836 consecutive new patients presented to the study sites between January and December, 2010. The distribution of the patients seen in the skin clinic of the study sites within the period of review are as shown in Table 1.

Table 1: Distribution of consecutive new patients seen in the study sites between Dec, 2009-May, 2011

UNTH, Ituku Ozalla

Adults		**Children**	
Sex	**Frequency (%)**	**Sex**	**Frequency (%)**
Male	578(43.3)	Male	114(38.6)
Female	758(56.7)	Female	181(61.4)
Total	**1336(100)**	**Total**	**295(100)**

Federal Teaching Hospital, Abakaliki

Adults		**Children**	
Sex	**Frequency (%)**	**Sex**	**Frequency (%)**
Male	74(42.5)	Male	13(41.9)
Female	100(57.5)	Female	18(58.1)
Total	**174(100)**	**Total**	**31(100)**

Of these, 1,631 were seen at the UNTH, Ituku-Ozalla, Enugu while 205 were seen at the Federal Teaching Hospital, Abakaliki. Three hundred and twenty-six (17.8%) were children while 1,510(82.2%) were adults. There were more females than males presenting to the clinics, both in the paediatric and the adult age groups.

Table 2: Sex and age distribution of patients with Hansen's disease in both institutions

Sex	**Frequency (%)**
Male	8(61.5)
Female	5(38.5)
Total	**13(100)**
Age(years)	**Frequency (%)**
<10	1(7.7)
10-20	1(7.7)
21-30	5(38.5)
31-40	1(7.7)
41-50	0(0)
51-60	2(15.4)
>60	3(23)
Total	**13(100)**

Table 2 shows the age and sex distribution of patients with Hansen's disease. Total number of patients with Hansen's disease was thirteen, of these nine were misdiagnosed.

Table 3 shows referral sources and referral diagnosis of the misdiagnosed patients. Figs. 2 to 5 are clinical photographs of some of these patients.

Fig. 2: Right knee of patient with non healing ulcer, note madarosis on patient

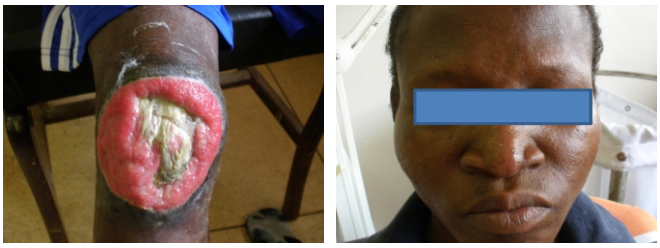

Fig. 3: Female patient with extensive nodules

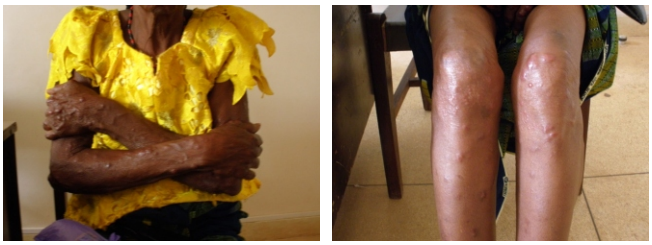

Fig. 4: Young male patient with extensive hypopigmented patches

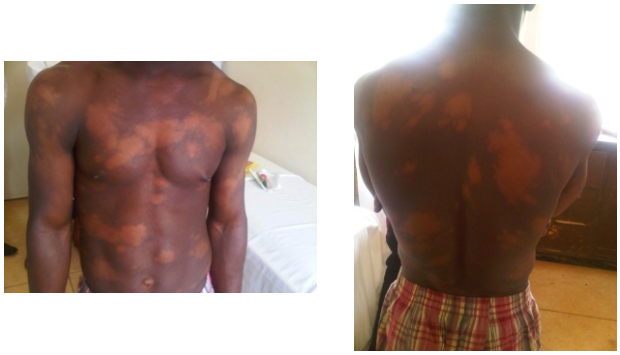

Fig. 5: Young female patient with extensive nodules/non-healing ulcers

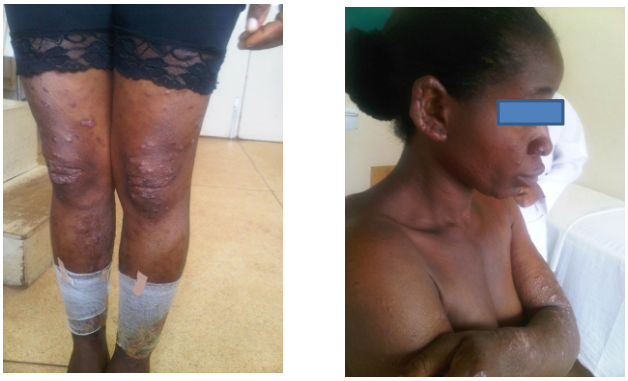

Fig. 6: Male patient with ENL/downgrading

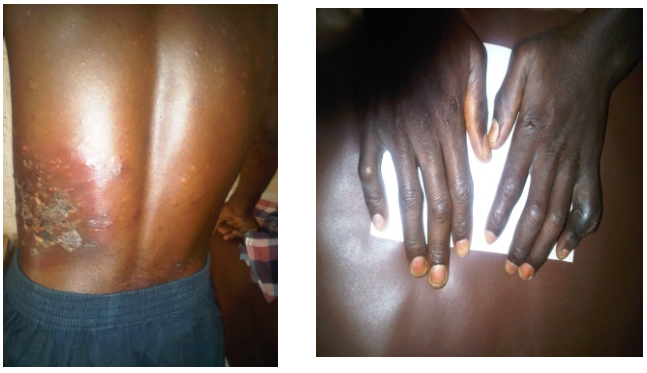

Table 3: Table showing number of misdiagnosed cases (N=9)

Sex	Age(years)	Referral clinic	Referral diagnosis	Skin clinic diagnosis
Female	64	Surgery	Hypopigmentation? Cause	Tuberculoid leprosy(downgrading)
Female	63	GOPD	Hypopigmented patches? Cause	Tuberculoid leprosy
Male	53	Private hospital	Allergic dermatitis? Acromegaly	Lepromatous leprosy
Female	28	Private hospital	Dermatitis	Lepromatous leprosy
Male	62	GOPD	Dermatitis? Cause	Lepromatous leprosy
Male	36	Orthopaedic clinic	Traumatic ulcer	Lepromatous leprosy
Male	6	GOPD	Dermatitis	Lepromatous leprosy
Male	20	Private hospital	Dermatitis	Tuberculoid leprosy(downgraded)
Male	24	GOPD	Hypopigmented patches? Vitiligo	Lepromatous leprosy

DISCUSSION

National leprosy control programmes have been successful in implementing the 2006-2010 global strategy in the World Health Organisation (WHO) regions [12]. Based on early detection of new cases and free provision of multi drug therapy (MDT), this strategy has been said to be effective in reducing the burden of disease in many endemic countries. The WHO has gone further to devise, in collaboration with national leprosy control programmes and other partners, an enhanced strategy which aims to sustain the provision of high-quality patient care, reduce disease burden through early detection of new cases and reduction in disabilities, stigma and discrimination[13]. The strategy will also provide social and economic rehabilitation. In line with these strategies, early detection of this potentially curable disease is of vital importance in order to prevent associated sequelae.

One of the elements of the 'final push strategy' of the WHO is to promote community awareness of leprosy so that individuals with suspicious lesions will report

voluntarily for diagnosis and treatment. [14] The WHO also recommends that national programmes should ensure that new cases of Hansen's disease are detected early to maintain the current declining trend in the burden of leprosy in endemic countries[15]. Since Nigeria is one of the endemic countries, it is pertinent that health care providers, even more than communities, respond to this call. There is, therefore, a vital need to increase awareness of the various unusual facades of this disease in order to arrive at prompt diagnosis.

Overdiagnosis of leprosy may occur where the health workers are poorly trained and so the clinical diagnosis of leprosy may not be reliable in such settings. For instance, Faye et al[16] in their study of 1729 children seen in two villages in Mali identified 71 patients with hypochromic patches. None of these children, though, had Hansen's disease and the authors made a case for general health workers first eliminating the most obvious differential diagnoses of leprosy before referring remaining cases to a higher level where a more specialized leprosy worker would make the diagnosis. The most important potential source of error is the reliability of the examination of an individual patient, by uninitiated health workers [17]

Misdiagnosis of Hansen's disease, on the other hand, has grave consequences not only for the patient and family members but for the society at large. Delays in the diagnosis of leprosy are not uncommon and misdiagnosis is more common in non-endemic countries where the disease is rare due to long incubation period of the organism, its variable clinical presentation and waning expertise of the health workers in making a diagnosis[17]. Neural involvement of Hansen's disease is also another common diagnostic dilemma[18, 19]. Poor knowledge and awareness of leprosy among health care workers is big drawback to accurate and timely diagnosis of Hansen's disease[20,21]

Delays and the associated factors in diagnosis of leprosy differ from study to study and may not be compared directly without bias due to the differences in study population, and the methodology used in data collection and analysis in a context of different cultural and socio-economic settings[22]. In the study by Zhang et al[22], out of the 88 patients diagnosed with leprosy, 23 (26·1%) were diagnosed at the time of their first visit to healthcare services, 40 (45·5%) were diagnosed at their second to fifth visits, eight (9·1%) were diagnosed at their sixth to tenth visit, and 17 (19·3%) at their eleventh or later visits.

Our study showed ignorance in diagnosis of Hansen's disease among clinicians (both those in the tertiary and private hospitals) especially in cases presenting atypically. Zhang et al[22] found that 44%of the patients in their first three health seeking actions are misdiagnosed in dermatological services. Out of the 13 cases of Hansen's disease that were referred to our clinics during the study period, 69% were misdiagnosed. This is alarming and calls for urgent need for improvement in the diagnostic skills of health workers in identifying leprosy patients, development of better laboratory tools, not only for early diagnosis of disease but also for evaluating response to treatment. There was a disproportionate affectation of males presenting with the disease and this is different from a previous study by Nnoruka[11] but compares favourably with other studies[22].This preponderance in males is real and not related to underdiagnosis in women as Britton et al[3] found a male predominance in Hansen's disease patients after puberty; with a male to female ratio of 1.5-2 to 1..

CONCLUSION

At the early stage of the disease, Hansen's disease may manifest in ways that make diagnosis difficult. Delays in diagnosis results in serious consequences for patients and these include nerve damage and disability. Hansen's disease should be considered as a cause of peripheral neuropathy or persistent skin lesions in patients residing in leprosy-endemic countries. Healthcare providers need to maintain a high index of suspicion and improve their diagnostic skills in order to reduce the time to diagnosis of the disease and the attendant sequelae.

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