Case Report

Hansen's Disease: A Neglected Tropical Infectious Condition

Malumani Malan[1], Chiyenu O.R Kaseya[2], Song Ji Quan[*3]

[1](a) Department of Medicine-Dermatology and Venereology Unit, Livingstone Central Hospital, Southern Zambia. (b) Department of Dermatology and Venereology at Zhongnan Hospital of Wuhan University, Wuhan City, Hubei Province, China. Email: drmalumani@yahoo.com

[2]Department of Medicine, Livingstone Central Hospital, Southern Zambia. Email: kaseyachiyenu@gmail.com

[3]Department of Dermatology and Venereology at Zhongnan Hospital of Wuhan University, Wuhan City, Hubei Province, China. Email: songjiq@126.com.

Abstract: This case of Hansen's disease reminds clinicians of all ages and era that leprosy is not a disease of the past as such a systematized multidisplinary approach and timely referral to specialists should be followed when evaluating any patient with chronic dermatoses of unknown cause. Early diagnosis of leprosy coupled with prompt treatment with Multi Drug Therapy (MDT) are the most important steps in preventing permanent debilitating deformity and disability.

INTRODUCTION

Hansen's disease which is Leprosy is defined as a chronic granulomatous disease caused by the acid fastidious bacillus called Mycobacterium leprae that primarily affects the peripheral nervous system and the skin.[1] Although it does affect other body parts as well like the reticulo-endothelial system (spleen, lymph nodes), musculoskeletal system, mucous membranes, eyes, testes, adrenals and other systems.[2]

In 1998, WHO categorically defined leprosy case, "as any person having one or more of the following features, and who has still to complete a full course of multiple drug treatment: hypopigmented or reddish skin lesion(s) with definite loss of sensation in those areas; involvement of the peripheral nerves, as evidenced by definite thickening with loss of sensation (varying degrees); and skin-smear positive for acid-fast bacilli".[3]

The current case notification of Leprosy in Zambia is at about 0.43 per 10,000 populations as recorded by Kapata et al,.[4] However, as of year 2000, the prevalence rates had decreased to 0.67 / 10 000 population. The author also notes that the Zambian scenario has a lot of inadequate data recording, poor surveillance systems, record keeping, and insufficient information on disaggregation by geographical origin or gender.[4]

Historically, in 1873, a Norwegian doctor called Gerhard Armauer Hansen first discovered Mycobacterium leprae while working in a leprosarium in Bergen, Norway. At that time in Europe, this region had the highest prevalence of leprosy. However, the scourge of leprosy appeared to have peaked in the Middle Ages as the disease spread through Europe as scribed by the ancient Greek, Roman and Egyptian writings. Hansen's disease was generally thought to be a heritable disease before the era of modern and current microbiology was developed.[5]

Clinically this neglected tropical infectious condition has a variable clinical presentation in reference to the patient's immune status. The Ridley and Jopling /WHO classification criteria, is based totally on; scientifically clinical symptoms, pathological, bacilloscopic, and immunological

Correspondence:
Professor Song Ji Quan,
Head of Department of Dermatology and Venereology at Zhongnan Hospital of Wuhan University, Wuhan City, Hubei Province, China.songjiq@126.com. phone: +86 13797094252

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components of the disorder, thus, allowing for a radical characterization or instead categorization of six leprosy class: the polar tuberculoid leprosy (TT), borderline tuberculoid (BT), the intermediate borderline-borderline (BB), borderline lepromatous (BL) and lepromatous leprosy (LL) form.[6] A sixth form is the indeterminate leprosy (IL), which is also commonly used to refer to leprosy which may resolve or progress further to anyone of the five forms of leprosy within the Ridley-Jopling system.

Another simplified classification of leprosy based on the bacterial index (BI) classifies leprosy into two categories: paucibacillary (PB) and multibacillary (MB) which is helpful in operational and therapeutic purposes. In PB, the BI is lower than 2+ and in MB the index higher than or equal to 2+.[7]

The complications of leprosy can be both socially and emotionally debilitating both to the patient and the next of kin. In this case report we discuss and review a case of a 35-year-old female with physically debilitating Lepromatous leprosy.

Case Presentation: A 35-years-old single woman had presented to the out-patient department with a with a long-standing history of worsening macular-papular rash with associated development of hyper and hypopigmented skin lesions for over two years. Since the first presentation, she was managed as a case of chronic dermatoses with infected skin ulcers under investigation and was covered on systemic short course of antibiotics and advised on daily wound care.

When we first saw the patient, she reported worsening skin lesions, coalescing of the macular papular rashes, hardening of the skin, facial changes disfigurement, worsening of skin ulcers especially on legs which where discharging chalky-watery fluid, malaise and occasional fevers since the past three years of being evaluated as an out-patient.

The review of other systems was unremarkable. The client was found to be HIV/AIDS sero-negative and had no significant medical nor surgical history.

Her local skin exam revealed granulomatous, shiny coalesced nodular-papular lesions around the face and ears with associated skin ulceration. She had a saddle nose, loss of eye brows. The limbs had a mixture of both hyper and hypopigmented lesions with multiple ulcers on the upper and lower limbs which had clear margins, granulating clean surfaces with sero-purulent discharge on some lesions. There were areas of skin induration and woody hard on palpation as seen in figure 1 (A-E). She had normal morphology of fingers and toes. However, neurological exam could not be accessed due to woody hard nature of the limbs. Also peripheral nerve enlargement was not appreciated.

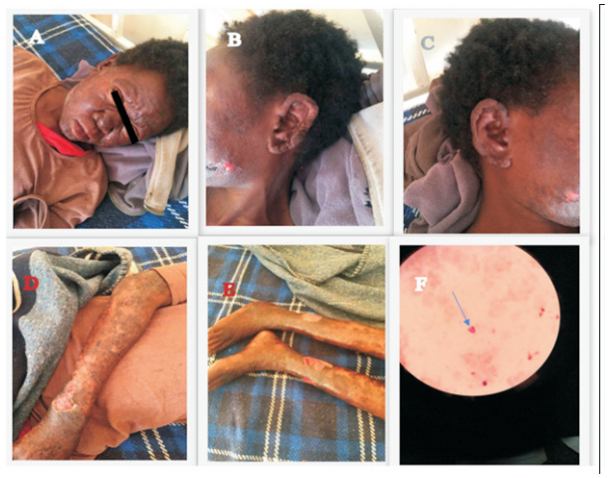

Figure: (A-C) Lepromatous leprosy: dysfiguration, infiltrated face with nodular-papular lesions, saddle nose and madarosis, granulomatous lesions on ear lobes with superficial ulcers on the cheeks.**(D-E)** complications of nerve damage evidenced as excoriations, multiple skin ulcers with hyper and hypopigmented lesions on limbs, however she had normal fingers and toes **(F)** Indicates positive (+++) mycobacterium leprae of over ten per High Power Field(>10AFB/1HPF) from the nose, both ears, hands and legs under Ziehl-Neelsen

Skin biopsy/ scrapping 'revealed' positive (+++) mycobacterium leprae of over 10 per High Power Field (>10AFB/1HPF) from the nose, both ears, hands and legs under Ziehl-Neelsen as shown in **figure 1F**. Other investigations were within the normal range as indicated in **table 1**.

The patient was managed as Lepromatous Leprosy based on the clinical picture and positive laboratory findings and was initiated on Multi Drug Therapy (MDT), as per protocol on: Clofazimine 300mg

once a day, Rifampicin 600mg once a day, and Dapsone 100mg once a day for a year with scheduled reviews. Also, the client was offered daily wound care, pyridoxine, multivitamins and vitamin B complex supplement.

Table 1: investigations done during period of admission

Test	Value	Normal range
Complete Blood count		
WBC	4.10x 10^9/L	4.00 - 10.00
Neutrophils	2.21x10^9/L	0.96- 6.40
Lymphocytes	1.37x10^9/L	0.84 – 3.26
Monocytes	0.52 x10^9/L	0.08 – 0.61
RBC	3.99x 10^{12}/L	4.13 – 5.67
Hemoglobin	10.2g/dL	12.1 – 16.3
HCT	34.7%	35.0 – 47.0
MCV	87.0fl	79.1 - 98.9
MCHC	29.4g/dL	32.0 – 36.0
Red cell distribution width	19.3%	11.6 -14.0
Platelets	443x 10^9/L	150 - 400
ESR	Nil	
Biochemistry		
S-Urea	3.23mmol/L	2.80- 7.10
Creatinine	80.1µmol/L	45.0 – 84.0
Alanine Transaminase	20.6U/L	0.0 – 34.0
Aspartate Transaminase	29.6 IU/L	0.0 – 31.0
Others		
Syphilis RPR	Non-Reactive	
Malaria	Negative Slide	

After eight weeks of the above constituted management, the patient had shown tremendous improvement and the ulcers had healed. However, for the woody hard and scared skin lesions, nothing more could be done. She was advised that she would have to bear with the 'mummified' kind of skin with the hope that it may soften with time. A social worker and psychologist where incorporated to address her psychosocial needs.

DISCUSSION

The mycobacterium leprae organism was declared less virulent and almost eradicated in most parts of the world with most sanatoriums converted to either health centers or hospitals for all kinds of conditions with the best example in Zambia being Liteta General Hospital in Chibombo district, Central province. Yet the organism has found means and ways to still cause devastating conditions to our client in this era in a sporadic pattern. Our client battled with symptoms of leprosy but they were dismissed due to the fact that the condition was considered controlled as per the World Health Organization (WHO) target of yearly overall prevalence rate of less than one case for every 10,000 populations.[8]

Most clinicians have never seen an active case of leprosy in this era, thus the patient was treated as unknown chronic dermatoses in the early days. This case-review reminds us that these infectious and ancient ailments should always be thought of despite the disease being considered as an eradicated disease especially in our part of the world. The earlier manifestations of leprosy in our client was completely missed due to either due to lack of specialist availability in the area of her origin or and largely due to knowledge gap amongst clinicians. There has not being any cases of leprosy in the region of her origin. Also, the health facilities that our client initially sort medical attention had no capacity to further investigate the clinical presentation of the patient thus, she was managed as a case of infected chronic dermatoses of unknown cause.

Clinically, Leprosy affects mostly the peripheral nervous system, the mucocutaneous system; the mucosa membranes of the upper respiratory pathways are never spared.[9] It is essentially a disease of the poor with its transmission thought to be through nasal droplets as is the suspicion with our patient. Although she denied contact with any person with similar presentation. The point of contact with the organism in our patient was illusive and thus, earliest thoughts' of Hansen's disease was completely never entertained. Zoonotic transmission has been recorded especially when in contact with armadillos as documented by Storrs E.E et al.,1974. Prolonged skin contact with an untreated patient and direct inoculation are some other mode of transmission.[10] However, the precise mode of transmission is still unclear but frequent, repeated, close contact with an untreated person for a

longer period of time can lead to contracting leprosy. The disease isn't highly contagious as compared to other mycobacterium species.

In an attempt to explain the clinical presentation of our client, the development of symptoms is due to the immune status and cellular immune response of the client or host to M. leprae organism than on the bacillary penetration virulence and multiplication ability. Clinical presentations are almost always followed by a long incubation period, ranging between several months and twenty years with an average period of two to four years. Hence, if clinician has a low index of suspicion, may miss or misdiagnose the disease.[11]

The pathophysiological progress of Hansen's disease is thought to be a model of graded cell-mediated immunity, with the trigger in this case to the causative organism, Mycobacterium leprae. The clinical presentations are as a result of constellation of serval factors: a) due to bacterial progression, b) immunologic responses of the host, c) peripheral nerve damage as a result of either or both bacterial progression and immunologic responses of the host, and d) Ultimately the preventable secondary deformities because of nerve damage, which basically account for most of the psychosocial stigma of the disorder in leprosy patients.[9]

The evolving clinical picture starts as innocent as small number of hypochromic spots or macules (light skin patches), with associated slight decrease in sensitivity to pain and light touch, without noticeable increase in nerve thickness to single skin lesions or a small number of asymmetric skin lesions. They are defined by erythematous plaques and nodules in some cases. Often the ulcers are with elevated external borders and hypochromic centers, also presenting with significant change in sensitivity of pain, light touch and pressure. Alopecia and anhidrosis are other features of leprosy as a result of denervation of the skin appendages, and thickening of the nearby nerve sheath, and hyperkeratosis and or ulceration in the compression areas. These aforementioned features were present in our client at the time diagnosis. The peripheral nerve damage in leprosy often results in severe sensory and motor dysfunctions that often lead to permanent deformities and or disabilities.[12] The sensitive change within the nerve route, with or without any substantive nerve thickening, has been the only manifestation in some instances, characterizing the primary neural form of the disease.[11]

MDT treatment options are readily available to control or minimize the effects of bacterial progression, undesirable immunologic responses of the host, also peripheral nerve damage, and secondary deformities. Preventive measures have also been put in place from vaccination of neonates with Bacillus Calmette–Guérin (BCG) vaccines to treatment of contacts of leprosy cases with protracted course of rifampicin. However, isolation of leprosy cases is not required since the condition is not highly infectious.[13]

Untreated patients or late diagnosis almost always leads to irreversible or rather permanent disabilities and depressing disfiguring complications, like the leonine facies. These physical complications are associated with socio-cultural construction of leprosy are responsible for stigma and psychosocial exclusion of leprosy victims.[14,15]

CONCLUSION

This case of leprosy reminds general practitioners that leprosy and other neglected tropical diseases are not ailments of the past and thus, a systemic and multidisplinary approach have to constantly be followed when in limbo. Early diagnosis of leprosy is a prerequisite for effective therapy and rehabilitation. Early detection and treatment by MDT are the most important steps in preventing and decreasing chances of severe deformity and disability.

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Ethical Consideration: Informed consent was obtained from the patient to use her images.

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