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## **P R I S M   S E C T I O N**

**(Patients' Revelations as Insightful Studies of their Management)**

- A Patient With Coarse Facies And Skin Changes — A Case Report Of Undiagnosed Hansen's Disease Presenting To Primary Care. When Should We Suspect It, And What Should We Do?
- Unsteadiness And Clumsiness — Could There Be Something More Than A Minor Stroke? A Case Study To Remember.

## A PATIENT WITH COARSE FACIES AND SKIN CHANGES — A CASE REPORT OF UNDIAGNOSED HANSEN'S DISEASE PRESENTING TO PRIMARY CARE. WHEN SHOULD WE SUSPECT IT, AND WHAT SHOULD WE DO?

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### ABSTRACT

**A 44-year-old Malay gentleman was seen at the polyclinic on the advice of a medical social worker. He was noted to have coarse facies, extensive skin peeling, and nail deformities in all 4 limbs. He was suspected to have leprosy and referred to the National Skin Centre (NSC) which confirmed the diagnosis of Lepromatous Leprosy by histology. He has shown good progress since starting on a course of treatment. A brief review is presented here as a reminder of the challenges faced in the care of a condition largely forgotten in our highly urbanised setting: Hansen's disease.**

**Keywords: Leprosy, Hansen's, Lepra Reaction, Coarse Facies**

SFP2016; 42(1): 49-53

### PATIENT'S REVELATION: WHAT HAPPENED?

H, a 44-year-old Malay gentleman was accompanied by his brother, S, when he walked into the polyclinic consultation room with a white cane in hand. The medical social worker at the local hospital who was alarmed by the noticeable skin changes the day before had encouraged him to seek further medical advice.

### History of Present Illness

H shared that he had been having rough skin, peeling, pain and bleeding in all four limbs for some time now. He was unable to quantify how long it had been, or to describe how the skin changes had evolved.

H was visually impaired. His right eye was blind from retinal detachment, and his left eye had suffered severe visual impairment for reasons he was unclear about. He has been registered with the Singapore Association of the Visually Handicapped (SAVH) since 1997.

### Occupational History

H worked previously as a masseur. He has since stopped this job for a few years due to his skin condition.

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### Social History

Both his parents passed away in 2014. Although his seven siblings are mainly in Malaysia, H has stayed mostly in Singapore. He stayed with his 1st brother until the brother was sent to prison. He now stays with S, his 2nd brother, who is married with 2 children. S has recently found a job with the Maritime and Port Authority of Singapore (MPA). H currently receives aid from the social welfare department in view of his financial circumstances.

### Family History

H has no significant family history of note. Notably, no one else in his family has a similar condition.

### Physical Findings

H was noted to have coarse facies. See Figure 1.

**Figure 1: Face — before treatment**



He was totally blind in his right eye while the vision in his left eye was at best that of light perception.

There was extensive peeling, desquamation and excoriations of the skin involving all digits of all four limbs. In the upper limbs, bilateral erythematous patches that were very mildly tender, digital resorption with nail preservation and severe onycholysis was noticed. A nodule was noted over the left proximal forearm while ulcers were noted over bilateral soles. See Figures 2-4.

Neurologically, impaired sensation to pinprick, soft touch and temperature was detected, in a glove-and-stockings fashion in all four limbs. Bilateral ulnar nerves were notably thickened.

### Diagnosis and Differential Diagnosis

I was intrigued by H's coarse facies when he first walked into the consultation room. My initial differentials included acromegaly, hypothyroidism, and congenital inborn errors of metabolism. After seeing his skin changes, I later added psoriasis to the list.

**Figure 2: Hands — Pretreatment dorsum**



**Figure 3: Hands — Pretreatment palmar**



**Figure 4: Feet — Pretreatment plantar**



After further reflection on the combination of signs and symptoms, it later dawned on me that H might actually have Leprosy. In view of the growing suspicion, an online notification to the Ministry of Health was submitted, and an appointment was arranged for H to be seen by a dermatologist.

### **GAINING INSIGHT: WHAT ARE THE ISSUES?**

This case highlighted 2 main issues:

- i) Why did H not seek treatment all these years?
- ii) What clinical features — history and clinical findings — help in the diagnosis of lepromatous leprosy?

#### **Reasons for Delay in Seeking Treatment**

##### **i) Financial constraint**

H's meager financial circumstances understandably

contributed to his placing health matters below the concerns of day-to-day living. We found it curious nonetheless that he did not seek medical advice when his vision was failing.

On learning about the possibility of reduced medical costs, he sought medical treatment with the accompaniment of his brother, S. Unlike treatment for Tuberculosis, Leprosy treatment is not free in Singapore. However, Mr H was reassured that he will receive financial assistance for the costs of leprosy treatment under the Medical Fee Assistance Card. This likely removed a major hurdle to treatment adherence.

At present, he continues to be assisted by the medical social worker at NSC.

##### **ii) Poor knowledge/health literacy regarding access to healthcare**

Without knowledge of subsidised care or help avenues available, H literally turned a blind eye to his skin condition even when it was deteriorating. He had, over time, adopted a passive attitude towards his health.

Perhaps the demise of both of H's parents in 2014 caused him to consider his health more seriously.

#### **Clinical Features that Help in Diagnosis of Lepromatous Leprosy**

Leprosy (Hansen's Disease), is an infectious disease caused by *Mycobacterium leprae*. Infection is usually acquired at a young age. In endemic countries, cases are diagnosed at a young age. But in non-endemic Singapore, they are usually diagnosed in their older years and diagnosis is delayed as leprosy may not be considered.

Its incubation period averages 5 years but symptoms may take up to 20 years to manifest.

The route of transmission is via droplets from the mouth and nose through close contact, but infectivity is low.

Leprosy can be classified into 2 types — Multibacillary and Paucibacillary. The former is infectious whereas the latter is not.

It can affect many organs, but mainly the skin, eyes, nerves, and limbs.

Our patient had the following features (See Figures 1-4)

- digital resorption with nail preservation
- thickened nerves
- peripheral neuropathy
- madarosis
- leonine facies

Refer to Table 1 for the other features you may find in a patient with Hansen's Disease.

Table 1: Features of Hansen's Disease
Skin
-shiny, non pruritic, erythematous nodules and plaques
- hypoaesthetic hypopigmented macules
-leonine facies
-madarosis
Eyes
-loss of vision
Nerves
-thickened nerves
-peripheral neuropathy
Limbs
-digital resorption

visibly less coarse compared to the initial polyclinic consultation 9 months earlier (See Figure 5). His skin condition had improved, with decreased desquamation, absence of nodules, and the resolution of ulcers on his extremities (See Figures 6-8). Skin sensation, however, showed no improvement compared to 9 months earlier. A repeat slit skin smear done 9 months into treatment showed a persistent bacterial index of 6+, but with no viable organism.

Overall, H appeared more cheerful, readily cracking jokes with us during the consultation. Although unable to see the physical improvements in his skin, he clearly felt better from having been on treatment.

H is currently staying with S and is being taken care of by S's family. As family physicians, besides medical management, we can value-add by educating patients on the condition, clearing any misconceptions and stigma about the disease, and directing them to the various social services available.

## DISCUSSION

### Lessons from H

H's case reminds us that leprosy can still present to us in the primary care setting. We need to be vigilant in order to suspect it. Leprosy can lead to potentially severe neurological sequelae if left untreated.

Patients often do not present with the typical features described above. In fact, it is very rare for patients to present with the leonine facies that H had.

Hansen's may be suspected on clinical grounds, but a

## STUDY THE MANAGEMENT: HOW DO WE APPLY THESE INSIGHTS IN OUR CLINICAL PRACTICE?

### Management in NSC

H was seen by NSC, where a forearm biopsy confirmed the diagnosis of Lepromatous Leprosy. A slit skin smear taken from his ear lobe showed a bacterial index of 6+ with 1 percent activity. He was initiated on the World Health Organisation multidrug regimen (Dapsone, Rifampicin, Clofazimine). A minimum of 24 months of multidrug therapy was planned for him. At the subsequent visit 2 months later, he was noted to be having swelling over several skin lesions and neuropathic pain due to neuritis. He was diagnosed as having a Type 1 Lepra reaction (Refer to Table 2) and was started on Prednisolone.

### Progress After 9 Months of Treatment

We reviewed him together with his NSC treating physician during his consultation in January 2015, 9 months after treatment initiation. H came with his brother, S. His face was

Table 2: Types of Lepra Reactions

Type 1 (reversal)
: due to increased cell-mediated response against leprosy bacilli or remnants of dead bacilli, typically in first few months after treatment initiation
: skin lesions become larger and more erythematous, may ulcerate
: may have new or worsening symptoms of neuritis, both motor and sensory
: systemic symptoms uncommon
Type 2 (erythema nodosum leprosum)
: due to a humoral (antibody-antigen) reaction to immune complexes, typically occurs a few years after treatment
: fever, malaise
: rapid onset of new subcutaneous painful and erythematous nodules
: painful neuritis
Erythema necroticans (Lucio's phenomenon)
: refers to a cutaneous vasculitis in patients with lepromatous leprosy, usually affecting patients who are not compliant to their medicine regimen
: rare, but potentially life threatening
: violaceous and haemorrhagic plaques
: necrotic ulcerations

**Figure 5: Face — After 9 months' treatment**



**Figure 6: Hands — After 9 months' treatment, dorsum**



**Figure 7: Hands — After 9 months' treatment, palmar**



**Figure 8: Feet — After 9 months' treatment, plantar**



histological or split skin smear confirmation is strongly preferred before subjecting patients to the prolonged regimen of leprosy treatment.

Diagnosis is usually clinical, as discussed earlier and elaborated in Table 1. Confirmation is via histology, obtained via a skin biopsy or a split skin smear.

Treatment would involve multidrug therapy with Dapsone/ Rifampicin/ Clofazimine for six months for paucibacillary leprosy, and 24 months for multibacillary leprosy.

Leprosy is spread via droplets among close contacts over prolonged exposure, but its infectivity is low. There is no vaccine to prevent Leprosy. A few studies have advocated the use of single-dose Rifampicin as chemoprophylaxis in close household contacts of patients newly diagnosed with Leprosy, but a few other studies have shown this not to be effective. As of today, there is no consensus on the use of chemoprophylaxis among household contacts. Currently, the best way of preventing its spread is via early detection and treatment of infected patients.

From our encounter with H, we have gleaned the following important points:

- 1) Although the disease is rare in Singapore, the general public and health professionals in particular need to be mindful of the possibility of Leprosy in our patient encounters.
- 2) Leprosy still exists in the community. Imported cases can also occur. We need to be vigilant and have a high index of suspicion, especially when patients do not present with the typical signs and symptoms.
- 3) A Lepra reaction (Refer to Table 2), if present, is a medical emergency as it can cause irreversible nerve damage and permanent disabilities. It also needs treatment with oral steroids.
- 4) Many leprosy patients come from poor socioeconomic backgrounds. Our management needs to include offering avenues for financial help for them to be compliant with treatment. Only with compliance will we be working towards eradication.

### **Leprosy in Singapore**

Leprosy remains a notifiable communicable disease in Singapore. Cases encountered in the community should be referred early for specialist diagnosis and initiation of treatment.

NSC estimates the current prevalence of Leprosy patients in Singapore to be about 300, with an incidence of about 10-15 cases yearly, most of whom are foreigners. Three-quarters of the cases seen in Singapore in the past 10 years were "imported" cases from endemic countries, which are countries of origin of migrant workers. The Ministry of Health estimates the incidence to have fallen from 21.3/100,000 in 1960 to 0.1/100,000 in 2013. In 2013, 3 non-residents were diagnosed with leprosy, accounting for 43 percent of the total incident cases that year.

Leprosy used to be a condition that drove fear into people around those infected. Sufferers of the disease were shunned. Fortunately, this has since improved. However, there are still leprosy sufferers out there who are alone and without friends and family. SILRA, Singapore Leprosy Relief Association, is a home located along Buangkok View, set up in 1971 to provide care for ex-leprosy patients who are destitute. It provides the basic necessities for daily living and healthcare, as well as teaches them handicraft skills and involves them in rehabilitative work.

### Public Health Threat Worldwide

Leprosy is still considered a public health threat in the developing Third World countries, with 9 countries in Africa, Asia and Latin America accounting for 75 percent of the global disease burden. Intensive efforts are still needed to reach Leprosy elimination targets in 5 countries, namely Brazil, India, Madagascar, Mozambique, Nepal. Therefore we need to be alert to imported cases.

### CONCLUSION

Leprosy keeps the medical profession humble. Even with current advances, we have been unable to eradicate this age-old disease. It still lurks in the midst of our community. The onus is on us to pick it up and institute appropriate management in a timely fashion.

### DECLARATION OF INTEREST

The authors declare that they have no conflict of interest in relation to this article.

### Contributorship

Dr Victor Loh contributed to the initial diagnosis and discussion of the patient's diagnosis when he first presented, and made important suggestions in the drafting of the manuscript. He was involved in the final approval of the manuscript to be published and agrees to be accountable for all aspects of the work. Dr Seow Chew Swee contributed to the analysis and discussion of the case, and made important suggestions in the drafting of the manuscript. He was involved in the final approval of the manuscript to be published and agrees to be accountable for all aspects of the work.

### Acknowledgement

Besides Dr Victor Loh and Dr Seow Chew Swee, I would like to thank Prof Goh Lee Gan for offering me utmost guidance in writing this article, for reviewing the patient with me, and for editing my drafts.

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