Malaysian Family Physician 2009; Volume 4, Number 2&3
ISSN: 1985-207X (print), 1985-2274 (electronic)

Online version: http://www.e-mfp.org

Test Your Knowledge

A RIGHT "SPIDER" HAND AND A LEFT "EAGLE" HAND

SB Khoo *FRACGP*, Penang Medical College.

Address for correspondence: Dr Khoo Siew Beng, Senior Lecture and Family Physician, Penang Medical College, No 4, Sepoy Lines Road, Penang 10450, Malaysia. Tel: 604-2263459, Email: siewbeng@gmail.com

Khoo SB. Test Your Knowledge: A right "spider" hand and a left "eagle" hand. Malaysian Family Physician. 2009;4(2&3):100-2

CASE HISTORY

Jim (not his real name) is a 68 year-old friendly and pleasant Eurasian man who has been very pleased to participate in our clinical teaching sessions for many years. To the medical students, Jim is fond of showing his hands and to test them by providing the following information:

"My left hand resembles the claws of an eagle and my right hand those of a spider."

"I was born with normal hands. More than 30 years ago, I was diagnosed with a disease for which I was treated with a particular drug that caused my skin to become very dark especially on sun exposed areas on my face and arms. Other people mistook me to be an Indian and spoke Tamil to me. The colour of my skin returned to normal several months after I completed a 2 year course of this medication"

"What do you think is wrong with me?"

QUESTIONS:

1. Describe what you see on Jim's hands.
2. What are possible causes for this type of hand deformity?
3. What was the confirmed diagnosis? What was the drug that Jim was referring to?

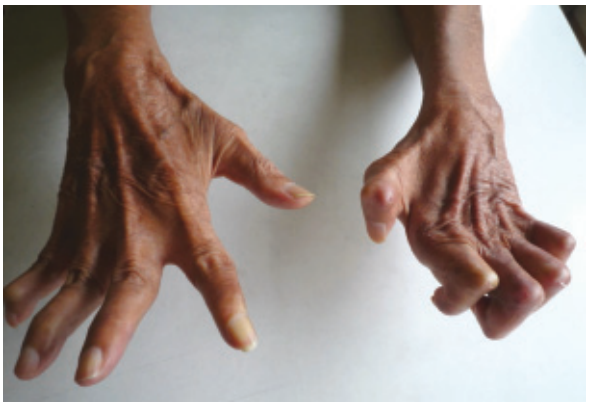

Figure 1. Dorsal view of both hands.

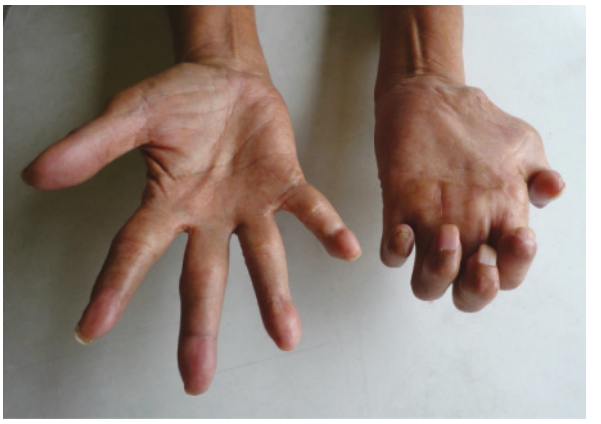

Figure 2. Palmar view of both hands.

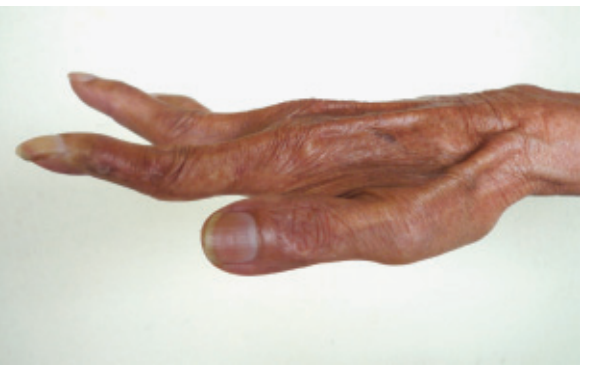

Figure 3. Lateral view of the right hand.

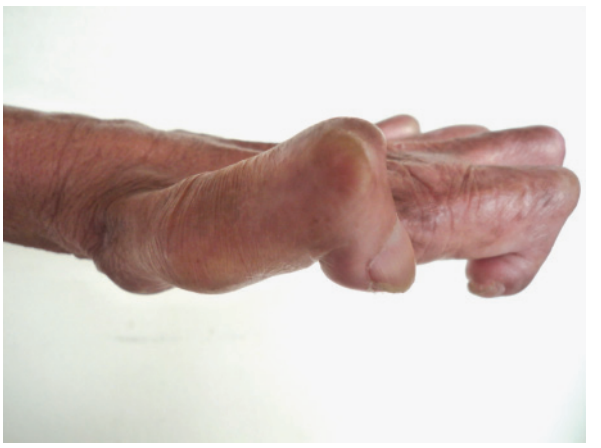

Figure 4. Lateral view of the left hand.

Malaysian Family Physician 2009; Volume 4, Number 2&3
ISSN: 1985-207X (print), 1985-2274 (electronic)

Online version: http://www.e-mfp.org

4. List down other associated physical findings likely to be found in this clinical condition.
5. What is the treatment guideline for this chronic disease?
6. What is the prognosis?

ANSWERS:

1. Right hand shows mild hyperextension at the metacarpophalangeal joints and flexion at the distal interphalangeal joints. Fingers are splayed out like those of a spider. Muscle wasting is minimal. Left hand shows severe hyperextension at the metacarpophalangeal joints and complete flexion at the proximal and distal interphalangeal joints likened to the claws of an eagle. There is marked wasting of interosseous muscles, thenar and hypothenar eminence of left hand. Right hand: Partial or ulnar-like claw hand. Left hand: Complete or true claw hand. Alternative term is "main en griffe".

2. Claw hand deformity is produced by imbalance of intrinsic and extrinsic muscles due to low ulnar and median nerve palsies. Intrinsic muscles are markedly weakened or paralysed. Long extensors hyperextend the MCP joints and long flexors flex the PIP and DIP joints. True claw hand may be found in several conditions:[1]
    - Lesion of both median and ulnar nerves (e.g. Hansen's disease)
    - Lesion of inner cord of brachial plexus
    - Klumpke's paralysis
    - Volkmann's ischaemic contractures
    - End result of suppurative tenosynovitis
    - Anterior poliomyelitis
    - Advanced rheumatoid arthritis
    - Uncommon neurological conditions such as syringomyelia, progressive muscular atrophy, polyneuritis, amyotrophic lateral sclerosis

3. In view of bilateral ulnar and median nerve palsies due to a chronic disease that had to be treated for 2 years, and a disease that was prevalent in Malaysia in the 60's to 70's, the likely drug that Jim was referring to was clofazimine and the chronic disease was Hansen's disease. Drugs with side effect of hyperpigmentation include clofazimine, tetracycline, azidothymidine, amiodarone, anti-malarial drugs, psoralens and chemotherapeutic agents such as 5 fluorouracil, busulphan and bleomycin.[2] Clofazimine is a phenazine dye used to treat rhinoscleroma, discoid lupus, leprosy, and other mycobacterial infections; it regularly induces a diffuse, reddish cutaneous and conjunctival discoloration within the first few weeks of use. With prolonged ingestion, affected patients typically develop violet-brown or bluish cutaneous pigmentation most apparent in lesional skin.[2]

4. Hansen's disease can be classified as tuberculoid, lepromatous, or borderline leprosy according to the severity and clinical presentation of signs and symptoms. People with tuberculoid leprosy typically have few skin areas affected (paucibacillary), and the disease is milder, less common, and less contagious. People with lepromatous and borderline typically have more skin areas affected (multibacillary), and the disease is more severe, common, and contagious:[3]
    - **Tuberculoid leprosy:** A rash appears, consisting of one or a few hypopigmented macules or plaques. Sensory impairment to pin prick and temperature may be present in areas affected by this rash because the bacteria damage the underlying nerves.
    - **Lepromatous leprosy:** General rashes of variable size and shape appear on the skin. It is a systemic disease affecting many areas of the body including the eyes (keratitis, glaucoma), face (leonine facie), reticuloendothelial system (hepatosplenomegaly, lymphadenopathy), peripheral nerves (mononeuritis multiplex, peripheral neuropathy, polyneuropathy), nose (epistaxis, collapse of nasal cartilage), kidneys (glomerulonephritis, renal failure), and testes (erectile dysfunction, infertility).
    - **Borderline leprosy:** Features of both tuberculoid and lepromatous leprosy are present. Without treatment, borderline leprosy may become less severe and more like the tuberculoid form, or it may worsen and become more like the lepromatous form.
    - The most severe symptoms result from infection of the peripheral nerves, which causes deterioration of the sense of touch and a corresponding inability to feel pain and temperature. Repeated damage may eventually lead to loss of fingers and toes. Weakness of muscles of fingers causes claw hands, weakness of muscles of foot causes foot drop. Infected nerves may be thickened and palpable on clinical examination.[3]

5. A World Health Organization (WHO) Study Group recommended multidrug therapy (MDT) in 1981. MDT consists of three drugs: dapsone, rifampicin and clofazimine. WHO estimates that early detection and

Malaysian Family Physician 2009; Volume 4, Number 2&3
ISSN: 1985-207X (print), 1985-2274 (electronic)

Online version: http://www.e-mfp.org

treatment with MDT has prevented about four million people from being disabled. This suggests great cost-effectiveness of MDT as a health intervention, considering the economic and social loss averted.[4]

- Paucibacillary leprosy should be treated for 6-12 months with dapsone 100mg/day unsupervised plus rifampicin 600mg/month supervised. This regimen should be followed by treatment with dapsone as monotherapy for 3 years in patients with tuberculoid leprosy or 5 years in patients with borderline lepromatous leprosy.[3,4]
- Multibacillary leprosy should be treated for 24 months with dapsone 100mg/day unsupervised, clofazimine 50 mg/day unsupervised, and rifampin 600mg plus clofazimine 300mg/month supervised.[3,4]
- Prednisolone is believed to minimize pain and acute inflammation. The recommended initial dose is prednisolone 40mg daily.[5]
- In US, lepromatous leprosy is treated with rifampicin 600mg daily for 2-3 years and dapsone 100mg daily for life. Tuberculoid leprosy is treated with dapsone 100mg daily for 5 years.[3,4]
- Once treatment is completed, the patient should be monitored for the next 5-10 years to evaluate for signs of relapse. Full blood count, serum creatinine and liver function test should be monitored at follow up visits.[3,5]

6. Hansen's disease is rarely fatal, and the primary consequence of infection is nerve impairment and debilitating sequelae. Nerve involvement results in loss of sensory and motor function, which may lead to frequent trauma and amputation. The ulnar nerve is most commonly involved.[5] Damage in the following nerves is associated with characteristic impairments in Hansen's disease:[5] Worldwide, Hansen's disease is considered the most common cause of crippling of the hand that is caused by ulnar nerve involvement.[5]
   - Ulnar and median nerves – clawed hand
   - Posterior tibial nerve – plantar insensitivity and clawed toes
   - Common peroneal nerve – foot drop
   - Radial cutaneous, facial, and greater auricular nerves may also be involved

ACKNOWLEDGEMENT

Sincere thanks to Jim for sharing his case history with us and giving permission for photography of his hands.

REFERENCES

1. Hamilton Bailey. Physical signs in clinical surgery. 3rd ed. Bristol: John Wright & Sons Ltd
2. Butler DF, Henderson DZ. Drug induced pigmentation. eMedicine Dermatology. Available at: http://emedicine.medscape.com/article/1069686-overview
3. Leprosy: Infections: Mycobacteria. Merck Manual Professional. Available at: http://www.merck.com/mmpe/sec14/ch179/ch179d.html
4. WHO / Leprosy. Available at: http://www.who.int/mediacentre/factsheets/fs101/en/
5. D Scott Smith. Leprosy: eMedicine Infectious disease. Available at: http://www.emedicine.medscape.com/article/220455-overview

Research Digest

Acid suppression is associated with increased enteric infections

Leonard J, Marshall JK, Moayyedi P. Systematic review of the risk of enteric infection in patients taking acid suppression. *Am J Gastroenterol.* 2007;102(9):2047-56.

This is a systematic review of 12 observational studies. There is an association between acid suppression and an increased risk of enteric infection (including Clostridium difficile, and other enteric infections).