Auto-Amputations

An 88-year-old man presented with catheter-related urinary tract infection. Deformities of both hands and feet were seen, with amputations of the digits being prominent features (Figs. 1 and 2). Radiographs of the right foot are shown in Figure 3.

What is the cause of this patient's multiple fingers and toes amputation?

A. Diabetes with peripheral neuropathy
B. Scleroderma
C. Ainhum (dactylolysis spontanea)
D. Buerger's disease
E. Leprosy

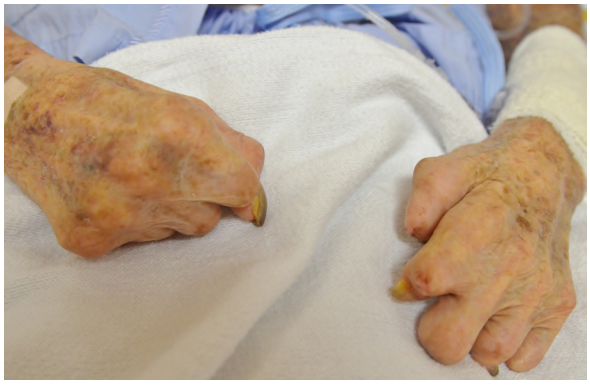

Fig. 1. Hands of the patient (dorsal view).

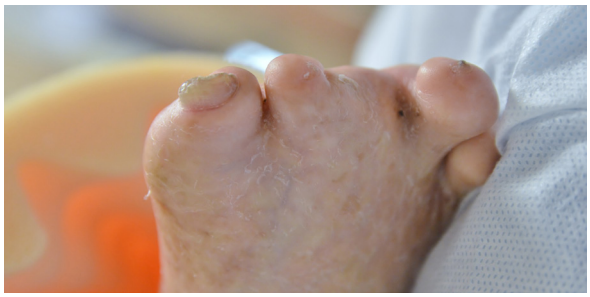

Fig. 2. Right forefoot and toes of the patient (dorsal view).

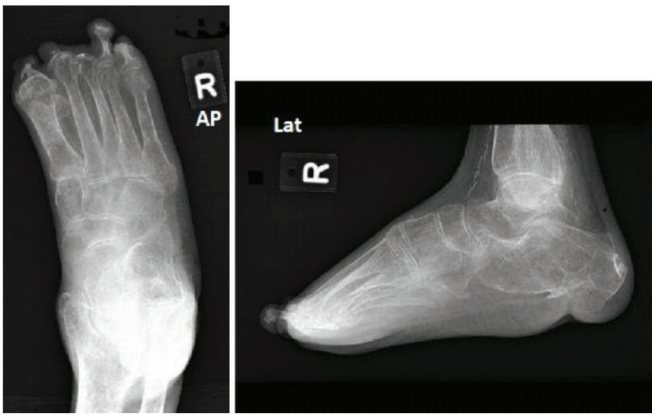

Fig. 3. Radiographs of right foot of the patient. There was diffuse resorption and acral osteolysis of the first to fourth phalanges. Charcot's deformity was also noted in the talus and possibly in the subtalar joint.

Findings and Diagnosis

Further history-taking revealed that our patient had been previously diagnosed with Hansen's disease, which was treated many years ago. He was unable to recall the details of the treatment. There had been no past history of diabetes mellitus. The patient was a non-smoker. He recalled no pain in the hands and feet prior to the progressive digital resorption.

A physical examination did not show any nodular lesion in the face to suggest leonine facies. No skin tightening, microstomia, calcinosis, nor telangiectasia were found. Greater auricular, ulnar, common peroneal and sural nerves were not palpable. Neurological examination showed normal deep tendon reflexes. There was loss of pain sensation, with anaesthesia involving gloves and stocking distribution bilaterally.

The patient's fasting sugar and glycated haemoglobin (HbA1c) levels were within normal limits, and hence these excluded the differential diagnosis of diabetes peripheral neuropathy. There were no clinical signs to suggest scleroderma such as Raynaud's phenomenon, gastro-oesophageal reflux or dysmotility or respiratory problems. Systemic sclerosis tends to affect women and the degree of auto-amputation in this patient was too extensive compared to what is usually seen in scleroderma.

Answer: E

The absence of a history of heavy smoking and limb pain from claudication make the differential diagnosis of Buerger's disease less likely (this typically causes ischaemic pain in the digits that progressively leads to skin ulcerations and gangrene). The patient did not have any gangrene at the digital tips preceding the shortening of the digits.

The patient, being an elderly Chinese man, did not fit the typical patient profile for ainhum which affects mainly African males aged between 30 and 50 years.[1] Moreover, there was no history of bulbous enlargement of the affected toes and no previous constricting bands prior to an amputation which is the main feature in ainhum.[1] Additionally, his auto-amputations involved multiple toes in comparison to ainhum which usually involves only the fifth toe.

Discussion

The incidence of leprosy in Singapore has greatly diminished over the past 5 decades—from 21.3 per 100,000 populations in 1960 to 0.1 per 100,000 populations in 2013.[2] The highest prevalence of the disease is observed in India, Africa and South America, and leprosy still occurs frequently in Southeast Asia.

Although the incidence of leprosy is lower than other causes of neuropathic arthropathy and auto-amputations, we would like to highlight this case as an important differential diagnosis not to be forgotten especially in the context of presentation in an elderly person living in Southeast Asia.

Transmission of *Mycobacterium leprae* (*M. leprae*) is through inhalation of the bacilli contained in nasal secretion and droplets.[3] Upon entering the body, the bacilli migrates and infects mainly Schwann cells and macrophages. Endothelial cells involvement has been described in leprosy, although specific endothelial cells and ligands mediating the uptake of the bacilli remains to be expounded.[4] Endothelial cells have also been identified to be a potential delivery system of viable bacilli to Schwann cells.[5] Proliferation of endothelial cells containing bacilli is found to be responsible in the pathogenesis of necrotising vasculitis.[6]

The manifestation of the disease is a spectrum determined by the ability of the host to mount cellular immunity response to *M. leprae*. When cell-mediated immunity (CMI) response is effective in eliminating the bacilli, skin lesions and peripheral nerves heal spontaneously, or the infection produces pauci-bacillary (PB) type of leprosy. If CMI is impaired, the disease spreads and produces multi-bacillary (MB) leprosy whereby in addition to skin and nerves, the eyes, testes, kidney, voluntary and smooth muscles, reticuloendothelial system and vascular endothelium can be infiltrated.

Infiltration with bacilli, inflammation from immune-modulatory cascades and granuloma within the epineurium sheath results in segmental demyelination, infarcts, oedema, and axonal damage.[7,8] This leads to impairment in sensation, autonomic and motor functions. Long-standing insult causes nerve destruction and replacement by fibrous tissue.

Acral osteolysis refers to resorption of the distal phalanx. The terminal tuft is most commonly affected and the shaft of the distal phalanx can also be affected. The mechanism of acral osteolysis of the digits is poorly understood; possibly from hypaesthesia from nerve involvement, compounded poor healing from vascular deficit and secondary infection from trophic ulcers.[9]

Firstline medications include dapsone and rifampin for tuberculoid leprosy, with addition of clofazimine for lepromatous leprosy. National Hansen's Disease Program (NHDP) advocated a treatment duration of 12 and 24 months for tuberculoid and lepromatous leprosy, respectively while the World Health Organisation (WHO) favours a shorter duration of treatment (6 and 12 months, respectively). Whilst the complex mechanism of acral osteolysis in leprosy is not completely understood, the implication of amputation is clearly understood—loss of function, leading to handicap and dependency. Early occupational therapy and podiatry care can help to delay the downhill progression of arthropathy.

REFERENCES

1. de Araujo DB, Lima SM, Giorgi RD, Chahade WH. Ainhum (dactylolysis spontanea): a case with hands and feet involvement. J Clin Rheumatol 2013;19:277-9.
2. Ministry of Health, Singapore. HIV/AIDS, STIs, Tuberculosis and Leprosy. In: Communicable Diseases Surveilance in Singapore, 2013. Ministry of Health, Singapore; 2013. p. 138-41.
3. Lastória JC, Abreu MA. Leprosy: review of the epidemiological, clinical, and etiopathogenic aspects-part 1. An Bras Dermatol 2014;89:205-18.

4. Scollard DM. Association of Mycobacterium leprae with human endothelial cells in vitro. Lab Invest 2000;80:663-9.
5. Barker LP. Mycobacterium leprae interactions with the host cell: recent advances. Indian J Med Res 2006;123:748.
6. Lu Z, Wu J, Cheng G. Leprosy. In: Li H , editor. Radiology of Infectious Diseases, Volume 2. Netherlands: Springer; 2015. p. 133.
7. Klionze AM, Ramos-Caro FA. Visceral leprosy: review. Int J Derm 2000;39:641-58.
8. Ebenezer GJ, Polydefkis M, Scollard DM. Mechanisms of Nerve Injury in Leprosy. In: International Textbook of Leprosy, 2016. Available at: http://www.internationaltextbookofleprosy.org/. Accessed on 14 June 2017.
9. Pamer P, Reeder M. Leprosy. In: The Imaging of Tropical Diseases. Available at: http://www.isradiology.org/tropical_deseases/tmcr/chapter34/intro.htm. Accessed on 14 June 2017.

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