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PHOTO QUIZ

A tropical disease characterised by rapidly progressive skin lesions and haemolytic anaemia

E.D. van Pelt[1], J.C. den Hollander[2], H.J.C. de Vries[3], Y. van Beek[1], D.C. Melles[4], W.I. van der Meijden[5], P.J.J. van Genderen[1*]

[1]Department of Internal Medicine, Institute for Tropical Diseases, Havenziekenhuis Rotterdam, the Netherlands, [2]Department of Pathology, Erasmus Medical Centre, Rotterdam, the Netherlands, [3]Department of Dermatology, Academic Medical Centre, Amsterdam, the Netherlands, [4]Department of Medical Microbiology and Infectious Diseases, Erasmus Medical Centre, Rotterdam, the Netherlands, [5]Department of Dermatology, Institute of Tropical Medicine, Havenziekenhuis, Rotterdam, the Netherlands, *corresponding author: tel. +31 (0)40-43305; fax: +31 (0)10-4121645; e-mail:p.van.genderen@havenziekenhuis.nl

CASE REPORT

A 37-year-old Philippine sailor with an unremarkable medical history visited the outpatient clinic of the Havenziekenhuis in Rotterdam. The patient initially noticed nodules in his face which spread over his body in a period of two weeks. He had also noticed a larger skin lesion with decreased sensitivity and paresthesia on his right elbow. In the past few months he had been travelling to India and South Africa.

Clinical examination showed a generalised papulo-nodular eruption (*figure 1a*). The diameter of the lesions varied between 0.5 and 5.0 cm. On the right elbow and lower arm there was a larger erythematous plaque-like skin lesion with numbness and central hypopigmentation (*figure 1b*).

Laboratory tests revealed signs of intravascular haemolysis. Direct antiglobulin test and test for enzymatic erythrocyte disorders were negative. Skin biopsies showed a granulomatous inflammatory process, with a clear zone near the epidermis (*figure 2a*). Additional Ziehl-Neelsen and Wade-Fite staining (*figure 2b*) revealed the presence of numerous acid-fast bacilli.

WHAT IS YOUR DIAGNOSIS?

See page 404 for the answer to this photo quiz.

Figure 1. *Clinical features. A) Generalised papulo-nodular eruption, ventral view of the trunk; B) Erythematous and infiltrated plaque with central hypopigmentation at the right elbow*

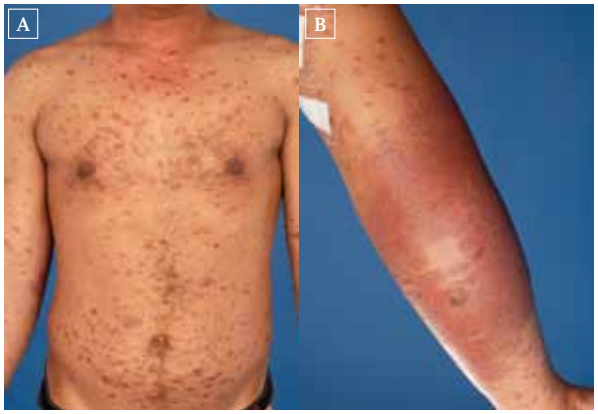
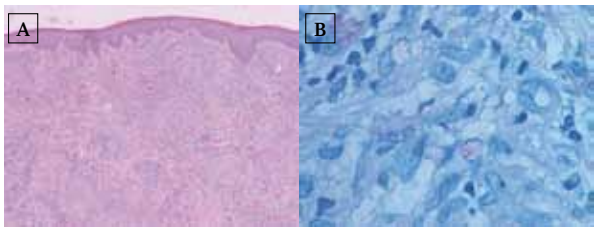


Figure 2. *Histopathology figures. A) Section from skin biopsy, showing a granulomatous inflammation process and a clear zone near the epidermis (original magnification x4); B) Wade-Fite stain revealing numerous acid-fast bacilli (original magnification x100)*



ANSWER TO PHOTO QUIZ (PAGE 400)

A TROPICAL DISEASE CHARACTERISED BY RAPIDLY PROGRESSIVE SKIN LESIONS AND HAEMOLYTIC ANAEMIA

DIAGNOSIS

A diagnosis of borderline lepromatous leprosy, also known as a type of multibacillary leprosy, was made. The diagnosis was confirmed by a leprosy-specific PCR, using *Mycobacterium leprae* specific repetitive element (RLEP). An ELISA for specific *M. leprae* anti-phenoclic glycolipid-1 (anti-PGL-1) was also positive.

DISCUSSION

Leprosy, caused by infection with *M. leprae*, is an ancient disease with a broad clinical spectrum which involves skin and peripheral nerves.[1] The specific host cellular immune response is responsible for its clinical disease manifestations. As a consequence of an impaired cellular immunity in the lepromatous type of leprosy, the leprosy bacteria can proliferate and infiltrate the tissues without restraint. The plaque-like lesion near the elbow is considered an immune area with signs of localised T-cell-mediated immune reaction, also known as an upgrade in the lepromatous spectrum towards the tuberculoid pole.[2]

Leprosy may also present with systemic features such as haemolytic anaemia, probably as a reaction to extensive infiltration with *M. leprae*. Haemolytic anaemia is a well-known complication of treatment with dapsone due to its oxidative stress effect on erythrocytes.[3] In contrast, our patient had not received prior treatment with dapsone. Additional tests revealed no other clue for the haemolysis than systemic reaction to massive infiltration with *M. leprae*. When multidrug treatment (MDT) for leprosy was initiated, which includes treatment with dapsone, frequent laboratory tests were performed to monitor progression of haemolysis. Fortunately, haemolysis did not worsen by treatment with dapsone.

Since the introduction of MDT for leprosy, the worldwide prevalence has decreased considerably.[1] Due to unfamiliarity with the disease in non-endemic industrialised countries, the diagnosis of leprosy is often not considered which may result in a significant delay in starting the appropriate treatment aiming at preventing permanent nerve damage and disability.[1,4] In the Netherlands it will take on average six years from the start of the first disease symptoms to administration of MDT.[4] Early case detection and MDT are of paramount importance to improve the clinical outcome of leprosy and are also key elements in the elimination of leprosy as a public health problem.

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