

A Case of Leprosy Mistaken for Cutaneous Sarcoidosis

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Leprosy, caused by the intracellular acid-fast bacillus *Mycobacterium leprae*, is a chronic granulomatous disease primarily affecting the skin and peripheral nerves. The infection is thought to be transmitted through droplets from the upper respiratory tract and is often indicated to have a mean incubation period of 5 years, although this is most likely much longer (1, 2). The clinical and pathological presentation of leprosy is determined by the cell-mediated immune reactivity of the host towards *M. leprae* antigenic determinants, resulting in either one of the polar stable forms of the disease; tuberculoid or lepromatous leprosy, but most frequently in one of the unstable borderline forms, called instable because the disease may upgrade or downgrade towards either pole during the natural course of the disease or during immunological reactions (type 1 leprosy reactions), which may be very harmful, resulting in permanent nerve damage (3). Another type of nerve damaging reaction that can also affect all other organs is erythema nodosum leprosum (type 2 leprosy reaction). Leprosy is often not recognized in non-endemic countries (4). We report here a case of leprosy in a Filipino woman working in Denmark, which was initially interpreted as sarcoidosis.

CASE REPORT

A 26-year-old woman from the Philippines, who had been working as an au pair in Denmark for 18 months, was referred to our hospital with a 3-week history of fever, patchy rash and conjunctivitis. Three weeks earlier she had sought treatment at a general practitioner due to fever, a rash which she described as spots on the face and transient arthralgia. The condition was initially considered as erysipelas, for which dicloxacillin

was prescribed, but a week later the macules conflated to large plaques and nodules and new elements appeared on the face and extremities. A skin biopsy was performed, which revealed numerous non-caseating epithelioid cell granulomas. These findings were interpreted as acute cutaneous sarcoidosis, and the patient was referred to a dermatologist at the local hospital. Here, the clinical findings were found to be compatible with either cutaneous sarcoidosis or Sweet's disease and a second skin biopsy from a patch on the left forearm was performed. This showed as previously, numerous non-caseating epithelioid cell granulomas consisting of histiocytes and scant giant cells surrounded by lymphocytes infiltrating the adnexa structures. At this point the patient was non-febrile and the lesions were not regressing. Since Ziehl-Neelsen staining and a PCR test for mycobacteria were negative, on the suspicion of leprosy, Wade-Fite (modified Ziehl-Neelsen stain) staining of the same biopsies was then performed, which revealed numerous acid-fast rods (Fig. 1A). Routine laboratory screening tests, including haemoglobin, leukocytes, liver and renal parameters, were all within normal range. Due to the possibility of sarcoidosis, plasma dipeptidyltransferase, calcium levels and a chest X-ray was performed; all tests were normal. The patient was referred to the Department of Infectious Diseases, where on admittance, approximately 3 weeks after the onset of symptoms, numerous macules, plaques and nodules of light coppery colour, with rough, dry, scaly, slightly elevated surfaces with well-defined borders on the face (Fig. 2), symmetrically on the extensor sides of the forearms and asymmetrically on the legs were found, while no patches were observed on the trunk or scalp (Fig. 1B). The patches were all anaesthetic to light touch and the patient was not able to discriminate between hot and cold at these locations. In addition, the patient had developed dactylitis of the right 4th finger and right foot drop. Swelling or nodular enlargement of nerves or earlobes was not present and the patient had not experienced any other neuropathy. Mild conjunctivitis, but otherwise normal eyes were observed. Slit skin smears from three different sites were performed: the left earlobe, the periphery of a plaque on the right femur, one from intact (normal-looking) skin from

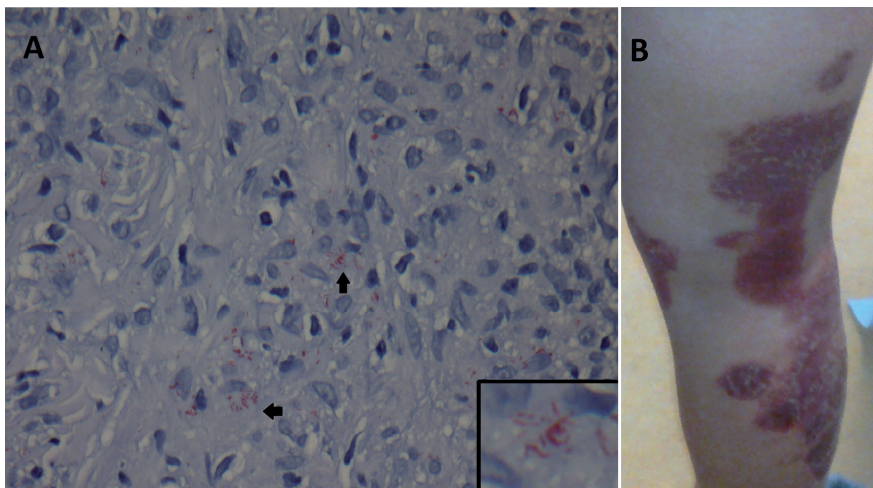


Fig. 1. (A) Wade-Fite special stain of skin biopsy showing the acid-fast *M. leprae* bacilli. (B) Slightly elevated, scaly and light-coppery coloured plaques on the right leg.



Fig. 2. Leprosy lesions on the face presenting as red/light-coppery coloured nodules and plaques.

the opposite thigh and a nasal swab were all stained for acid-fast bacilli. Microscopy of slit skin smears from the plaque and the intact skin both revealed numerous acid-fast bacilli, while no mycobacteria were observed in the smear from the earlobe and nasal mucosa. Laboratory findings showed an elevated leukocyte count $10.5 \times 10^9/l$ (normal range $3.5\text{--}8.8 \times 10^9/l$), lactate dehydrogenase 247 U/l (normal range 105–205 U/l) and C-reactive protein 16 (normal value <10). The rapid progression of the disease, with sudden onset of symptoms, clinical and histological findings designated the condition as borderline leprosy with type 1 reaction (up- or down-grading) (5, 6). The patient had never travelled outside the Philippines prior to going to Denmark, and she was not aware of any cases of leprosy in her family or any contact with known cases. The patient was categorized as a multibacillary case of leprosy and, according to WHO recommendations, was prescribed monthly dosages of 450 mg rifampicin (this dosage was adjusted to the patient's weight; the WHO recommended dosage is 600 mg), 300 mg clofazimine, and 100 mg dapsone daily (the dapsone dosage should be reduced if haemolysis is detected) and 100 mg clofazimine every other day. This regimen is recommended for 12 months (7). Furthermore, due to the leprosy reaction with nerve involvement, 50 mg of prednisolone once daily orally was added to the regimen, and was tapered to 15 mg after 3–4 weeks (8). It should be noted that the steroid regimen recommended by leading leprologists for type 1 reactions is a starting dose of 30–40 mg prednisolone tapered to zero after 5–6 months or even longer (9).

DISCUSSION

The clinico-pathological presentation of leprosy is determined by the cellular immune reaction: patients exhibiting a strong cellular response develop tuberculoid leprosy presenting with few skin lesions, and bacilli are rarely detected. At the other end of the spectrum, patients with a weak or no cellular response to *M. leprae* develop lepromatous leprosy with multiple lesions and bacilli. The borderline types lie between these two extremes (5). In our case the patient experienced a rapid deterioration involving sudden appearance of multiple elevated, erythematous skin lesions, not tender or accompanied with fever, and an almost silent nerve impairment, which designated the condition as borderline leprosy with type 1 reaction (9). Leprosy is endemic in resource-poor areas in which the infection is a great burden causing much disability (10). The diagnosis is additionally complicated by the long incubation period: patients present with symptoms long after arriving from endemic areas. In this case the suspicion of sarcoidosis was reinforced by the granulomatous skin biopsy further delaying the diagnosis of leprosy.

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