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BRIEF COMMUNICATION

A Case of Leprosy in Italy: A Multifaceted Disease Which Continues to Challenge Medical Doctors

Martina Maritati[1] · **Carlo Contini**[1]

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Abstract Leprosy is a chronic infectious disease caused by *Mycobacterium leprae*, characterized by a very long incubation period, confounding signs and symptoms and difficulty to establish the onset time. Considering the stigma associated with the diagnosis and the difficulties in detecting asymptomatic leprosy, the incidence and prevalence of this disease are underestimated. In Italy, leprosy is currently included among the rare diseases and can occur as an imported pathology in native individuals or extra-EU immigrants. Currently, given its exceptional appearance in Italy, leprosy is extremely difficult to recognize. In fact, the incomplete knowledge by the medical class of geographical epidemiology and aetiology of tropical diseases including leprosy, often delays the definitive diagnosis. Due to the increasing rate of the migration flows, in Italy and in Europe, leprosy should be considered among the differential diagnosis in patients with cutaneous and neurological signs, especially when originating from endemic countries.

Keywords Mycobacterium leprae · Borderline leprosy · Immigrants · Ulnar neuropathy

Martina Maritati and Carlo Contini have equally contributed to this work.

✉ Carlo Contini
cnc@unife.it

Martina Maritati
martina.maritati@student.unife.it

[1] Department of Medical Sciences, Section of Infectious Diseases, University of Ferrara, Via Aldo Moro, 8, 44124 Ferrara, Italy

List of Abbreviation

WHO	World Health Organisation
AIFO	Associazione Italiana Amici Raoul Follerau
EU	European
TB	Tuberculosis
SLE	Systemic lupus erythematosus

Background

Leprosy is a contagious infectious disease, often disabling, which occurs in susceptible individuals following contact with *Mycobacterium leprae*. The special affinity of *Mycobacterium leprae* for the skin, peripheral nerves (Schwann cells), extremities of the human body and the nose (cold areas of the organism), can disclose after a long period of incubation with clinical polymorphism, involving sometimes the eye, upper respiratory tract, bone and scrotum. Leprosy, rarely a direct cause of death, has a marked tendency to a spontaneous healing or to a chronic progressive evolution, often interspersed with remissions and exacerbations.

According to official reports received from 115 countries and territories, the overall registered prevalence of leprosy at the end of the first quarter of 2013 stands at 189,018 cases (excluding the small number of cases in Europe) [1]. Most countries that were previously highly endemic have achieved its elimination. However, pockets of high endemicity still remain in Angola, Brazil, the Central African Republic, India, Madagascar, Nepal, Tanzania, the Democratic Republic of the Congo and Mozambique. These countries remain highly committed to eliminating the disease [1].

In Italy, between 1990 and 2009, a total of 159 cases have been diagnosed almost exclusively in extra-EU immigrants [2].

Leprosy is currently included among the rare diseases which can occur as an imported pathology in native individuals, or in immigrants from these regions. Native outbreaks of Liguria, Puglia, Sicily and Sardinia are now virtually extinct. Herein we report the first case of Borderline Leprosy with simmetric face lesions and ulnar neuropathy, occurred in Italy since 2009.

Case Presentation

We describe the case of a 22 years old man, native from Ghana, who came to Italy after living in Libya in the previous 2 years. He was sent to the service of Rheumatology because of "pain in hands," due to a compression syndrome of the left ulnar nerve (tunnel olecranon). The patient, guest at one local Reception Center for refugees, was found positive to the tuberculin skin test and the QuantiFERON-TB Gold (Chest X-Ray negative); as a consequence, anti-TB prophylaxis with isoniazid was started.

After few months, the patient presented infiltrated and round subcutaneous lesions located on his face, interpreted by dermatologists as urticaria's manifestations. Such lesions, after a further dermatological consultancy, were subsequently described as "infiltrated, fixed, auburn and with irregular borders, associated with arthralgias and arthritis of the hands, ankles and knees". An infectious diseases consultancy described the presence of a symmetrical no scaling infiltrative dermopathy on the face, especially in the central portion (cooler part), with a thickening of facial skin and accentuation of the skin creases (Leonine facies), associated with anaesthetic areas along the edges of the lesions. There was also hair loss, involving eyebrows (lateral third) and beard. Face dermopathy was associated with apparent left ulnar neuropathy involving the fourth and fifth finger of the left hand, associated with sensory deficit (Fig. 1) [3]. Physical examination performed at general and local level (genital organs) was normal.

Suspecting a "Lepromatous Leprosy", a biopsy of the skin lesions was done. This showed the presence of "chronic granulomatous epithelioid inflammatory process with peri-neural distribution, with evidence of alcohol-acid resistant bacilli, located in macrophages and also present in Schwann cells", suggestive of "Tuberculoid Leprosy". Microbiological examination of nasal swabs and slit-skin smears for mycobacteria, was found negative. Serology for Lue, HIV, HBV and HCV was also negative, while lymphocyte T counts subpopulations and autoantibodies were normal, as well as chest X-ray and abdomen ultrasound. The Angiotensin-Converting Enzyme (ACE) was equal to 216.00 (n.v. 8.00–52.00).

A neurological consultancy confirmed the presence of ulnar mononeuropathy at the left elbow, while the ophthalmological consultancy showed no signs of active inflammation.

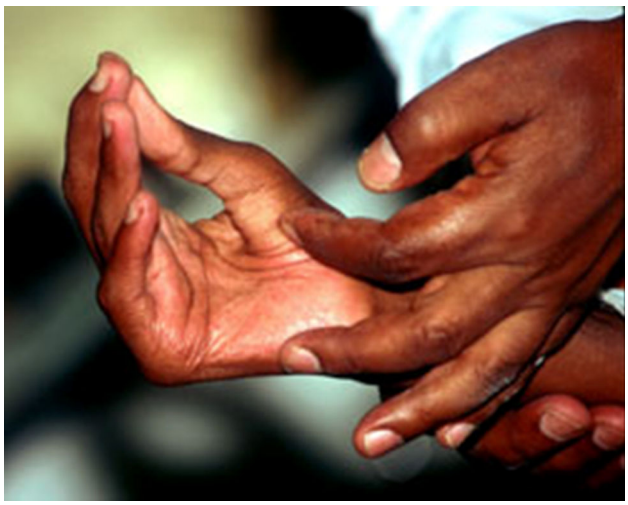

Fig. 1 Representative case of ulnar neuropathy involving the fourth and fifth finger of the right hand associated with sensory deficit [4]

The patient was addressed to the National Reference Center for Hansen's disease (S. Martino Hospital, Genoa, Italy). Following further detailed diagnostic histological picture matching with "Borderline Leprosy", he started specific therapy with rifampicin 600 mg, clofazimine 300 mg (monthly), clofazimine 50 mg and dapsone 100 mg/daily, respectively, in addition to steroid therapy (prednisolone) and vitamins (B6) to be taken every other day, as well as physiotherapic aid (left hand). After 2 months of treatment, the skin lesions improved despite the persistence of sensory, motor and autonomic deficit in charge to the left ulnar nerve, accompanied with numbness of plantar region of the right foot (due to the involvement of the rear tibial nerve). The follow-up checks were carried out every 2 months.

The treatment time was extended for 1 year according to the WHO guidelines, at the end of which the number of intact Fite-positive organisms in skin biopsies ("morphology index") resulted to be equal to zero. Currently, sensory and motor deficit still persists in the left ulnar nerve, although improved compared to the beginning of neurological symptoms. In contrast, the numbness of the plantar region of the right foot is completely solved.

Discussion

Considered a divine course for sin in the Old Testament and a karma in Buddhism, leprosy continues to be a challenge to health worldwide.

The correct incidence in Ghana is not known due to the lack of available data. In Libya (where the patient has been

living for 2 years before coming to Italy), 482 new cases have been detected, of which 357 presented Multibacillary Leprosy [4].

According to the data reported by *Hanseniology Italian Society* and the *Associazione Italiana Amici Raoul Follerau* (AIFO, Italy), there should have been approximately diagnosed from 6 to 9 new cases of leprosy, per year. A retrospective analysis performed from 2003 to 2009 has pointed out the discrepancy between the observed and the expected cases of Leprosy in Italy. Difficulty in accessing the National Health System, fear of segregation, ignorance and illegal immigrant status, would be possible explaining factors [2].

The reported data show that, despite widespread implementation of effective multidrug therapy, leprosy has not been eliminated, even if progress has been made in controlling the infection. According to WHO, the incidence rate decreased globally by 4 % during 2007 compared with 2006 [5]. A third of newly diagnosed patients have nerve damage and might develop disabilities, although this proportion varies according to several factors, including level of self-care [6].

So far, epidemiological data on disabilities are not available, but they are likely to show a worst scenario with respect to the one depicted by incidence rates only: the latest estimates compute the number of patients with leprosy-related neuropathy at over 3 million worldwide [5].

Leprosy is in fact, one of the most common causes of nontraumatic peripheral neuropathy in the developing world. *Mycobacterium leprae*, has a predilection for Schwann cells, where the pathogen multiplies unhindered by organism-specific host immunity, resulting in destruction of myelin, nerve architecture with secondary inflammatory changes.

The nerve damage affects sensory, motor, and autonomic fibers resulting in physical impairments, limitation of physical activities and social participation, as in the presented case.

Leprosy is a curable disease and treatment provided in the early stages could avoid the disabilities [7].

Cases of imported leprosy in Italy are infrequently observed. The MEDLINE and PubMed databases were searched for publications on leprosy in Italy from 2009 to 2013. Although sporadic cases were found during this period [7], the present case represents a paradigmatic example due to the simultaneous presence of cutaneous and neurologic lesions, in the absence of the positivity of skin smear.

A prompt diagnosis and the subsequent definition of a therapeutic regimen results to be extremely important, in particular in *Borderline Leprosy*, which without treatment could have the tendency to worsen toward Lepromatosus status, in contrast to the Tubercoloid form observed during or after specific treatment.

Currently, leprosy is extremely difficult to diagnose in part for the incomplete knowledge by the medical class of the geographical epidemiology of tropical diseases, which can result in delayed diagnosis. Moreover, the different "faces" of clinical illness, easily generate difficulties in differentiating leprosy from other pathologies such as sarcoidosis, fungal infections, SLE, erythema nodosum, polinevritis, syphilis and syringomyelia, particularly during the early stages of disease [8].

Conclusion

Leprosy has not been eliminated and due to the increasing migration flows, can also be observed outside endemic areas. Because of the long and unpredictable incubation time, it is possible that immigrants leave their own country without physical disabilities and develop leprosy and sequelae subsequently in their new country of residence. The present case, in view of the simultaneity of skin lesions and neuropathy, is a paradigmatic example of Hansen's disease. The critical issue is anyhow the medical expertise.

In Italy, according to a multicentric study recently published, the range of health problems in immigrants includes common infectious diseases, such as HIV infection, TB, hepatitis, and respiratory or gastrointestinal disorders that may reflect the deficiency of food, unhealthy conditions of life, and promiscuous sexual relationships [9].

Tropical diseases, although less frequent, should always be taken into account in relation to the increasing migration flows from endemic countries. In particular, in Italy and also in other parts of Europe, leprosy should be considered among the differential diagnosis in patients with suspect cutaneous and neurological signs and symptoms of this disease [10].

Informed consent Written informed consent was obtained from the patient for the publication of this case study, but not images of his face and body. A copy of a written consent is available for review by the editor-in-Chief of this journal.

Conflict of interest None.

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