



## Lepromatous leprosy presenting with erythema nodosum leprosum mimicking Behcet's disease

### ARTICLE INFO

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#### Dear editor,

Leprosy is a multisystem disease caused by *Mycobacterium leprae* (ML) infection [1]. Owing to a highly heterogeneous clinical presentation, the diagnosis of leprosy is often challenging.

A 29-year-old Moroccan male presented to our clinic because of a persistent skin rash symmetrically distributed at his face, trunk, limbs, and palmo-plantar regions.

His past medical history was remarkable for recurrent episodes of fever, arthralgia, emesis and oral aphthosis, which had appeared since his adolescence. One year before, he was hospitalized due to an acute pulmonary embolism associated with enthesitis, thrombophlebitis and a macular skin rash affecting his palms and soles. His symptoms led to a diagnosis of Behcet's disease, which was further supported by the positivity to the Human Leukocyte Antigen (HLA) B51. As a result, treatment with colchicine and prednisone was started. After 6 months, azathioprine was also added.

At our first visit, physical examination showed multiple, erythematous-brown nodules characterized by a central purulent discharge and a scaling peripheral edge, affecting symmetrically his lower limbs (Fig. 1 d,e,f). Erythematous, confluent and well-demarcated nodules on the face (leonine facies) were also observed (Fig. 1 a,b,c). Multiple lymphadenopathies localized on the neck, axillae and groins were also observed. The patient referred that he was suffering from frequent nasal bleeding and a sensory loss of the fourth and fifth fingers of the left hand.

Based on clinical findings and the absence of symptoms' improvement upon immunosuppressive treatments, a suspicion of leprosy was made; treatment with azathioprine, colchicine and oral steroids was interrupted. Two incisional biopsies were obtained from a skin lesion of the face and leg respectively. In both specimens, light microscopy examination showed dermal and hypodermal exudative-necrotic inflammation (Fig. 1 g,h). A positive Acid-fast bacillus staining was found. Polymerase chain reaction (PCR) for *Mycobacterium tuberculosis* complex was performed from a third biopsy obtained from the lower limb, but gave a negative result; PCR was conversely positive for ML. Collectively, clinical, pathological and microbiological findings were consistent with a diagnosis of lepromatous leprosy (LL) associated with erythema nodosum leprosum (ENL).

A treatment schedule including a daily intake of dapsone and clofazimine and a monthly administration of rifampicin 600 mg was commenced. The patient was lost during the follow-up.

Among the classic great imitators, including syphilis and lupus, leprosy's diagnosis is becoming challenging for medical practitioners. A long incubation period and a plethora of manifestations can delay the diagnosis, increasing the risk of developing neurological sequelae. Furthermore, leprosy is very rare in Western latitudes, while over 90% of cases of this disease occur in tropical area such as India, which is the country with the highest incidence of leprosy, and Brazil [1,2].

However, due to the forced migration of people, the disease is recently re-emerging in Western countries, including Europe [1,3].

Dermatologic signs of leprosy range from a single hypopigmented macule to diffuse nodules and might be accompanied by the involvement of autonomic, sensory, and motor fibers of the peripheral nervous system [2]. Moreover, patients may develop lepromatous reactions, that can occur before, during or after antimicrobial therapy, and include type 1 and type 2 reactions [4]. Type 1 reactions (reversal reactions – RR), involve a cell-mediated immune activation and are more frequently observed in tuberculoid and borderline leprosy; type 2 reactions, including also ENL, involve a neutrophilic immune reaction, and are more frequently observed in LL [4].

Noteworthy, clinical manifestations of leprosy may significantly overlap with several autoimmune diseases, including lupus, vasculitis and sarcoidosis [5–7]. Thrombo-embolic events have been also rarely reported in leprosy patients [7].

Because of the extreme clinical variability of this disease, it is important to remember the very early signs of LL, such as nasal bleeding, nodular thickening of earlobes and body hair loss, especially of eyebrows and lashes.

In our patient, the presence of venous thromboembolism associated with oral aphthosis and HLA B51 initially led to a diagnosis of Behcet disease. At the time of our visit the combination of facies leonine, erythema nodosum and autonomic symptoms was highly suggestive of Hansen disease. Arguably, either the long disease course or previous immunosuppressive treatments, including azathioprine, contributed to the shift from the original clinical phenotype toward ENL.

Our case indeed highlights the importance of improving

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**Fig. 1.** (a,b,c) Well-demarcated erythematous nodules on the face with variable size collectively configuring the so called leonine facies (d,e,f) Multiple, 0.5–1 cm in diameter, erythematous-brown scaling nodules with undefined edges and a purulent discharge affecting both legs and feet. Histopathological analysis showed: (g) Granulomatous inflammation with colliquative central necrosis of dermis with giant polymorphonuclear cells and neutrophils; (h) Dermal and hypodermal inflammation predominantly characterized by eosinophils, neutrophils, histiocytes and plasma cells. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

dermatologists' awareness of both pathognomonic and rare clinical signs of leprosy, whose incidence is gradually increasing in developed countries.

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#### Authors contributions

Lavinia Quintarelli: Conceptualization, Writing- Original draft preparation. Elisabetta Magnaterra: Conceptualization, Writing - Review & Editing. Roberto Maglie: Conceptualization, Writing - Review & Editing. Alberto Corrà: Conceptualization. Valentina Ruffo di Calabria: Visualization. Cristina Aimo: Visualization. Elena Biancamaria Mariotti: Visualization. Alice Verdelli: Supervision. Vincenza Maio: Visualization, Supervision. Emiliano Antiga: Supervision. Marzia Caproni: Supervision.

#### Declaration of competing interest

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