Facial rash in a 48-year-old woman

Case report of suspected leprosy in the emergency department

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Leprosy is a chronic infection caused by *Mycobacterium leprae* and *Mycobacterium lepromatosis*.[1-5] The reported global prevalence of leprosy has decreased from more than 5 million cases in the mid-1980s to 175 554 chronic cases in early 2015 and 213 899 new cases in 2014.[6] With most cases arising in India, Indonesia, Brazil, and the Democratic Republic of the Congo,[6] leprosy remains a rare disease in Canada, largely originating from immigration. Physicians' lack of familiarity with the disease and low clinical suspicion make diagnosing leprosy challenging.[7,8] Additionally, this illness is highly stigmatized across various societies, resulting in discrimination and social isolation.[9,10] Cultural beliefs range from deeming those infected as being inferior people to being punished by God. As such, many steps have been taken to reduce stigmatization, such as referring to leprosy as *Hansen disease*.[9]

We report the case of a 48-year-old woman from the Philippines presenting to a Canadian emergency department (ED) with dermatologic manifestations suggestive of leprosy.

Case

A 48-year-old woman presented to the ED with increasing periorbital edema for 2 days and a facial rash for 2 months that began on her cheeks. These lesions coalesced into larger plaques, and were accompanied by 3 new forehead lesions. She denied lesions elsewhere, pain, and pruritus. A walk-in clinic prescribed 10 days of oral antibiotics, which had little effect. Findings of a review of systems were unremarkable. She denied fever, night sweats, unexplained weight loss, focal deficits, or bowel or bladder symptoms.

The patient was otherwise healthy, took no medications, did not smoke, and had minimal alcohol use. She was born in the Philippines and immigrated to Canada in 1991, but had returned repeatedly since for approximately 2 weeks at a time, with her last visit in 2012. Her only other travels outside of Canada were to New York, NY, and Buffalo, NY. There was no family history of leprosy.

Vital signs at triage were unremarkable and the patient was afebrile. Findings of a head and neck examination revealed 5 well-demarcated eroded patches over the cheeks and forehead at various stages of healing (**Figure 1**) without purulence or serous crusting. A black eschar covered 1 lesion. Sensation of the lesions was normal to pinprick and fine touch. Periorbital edema limited left eye opening. There was no conjunctivitis or cervical lymphadenopathy. Findings of cardiovascular, respiratory, and abdominal examinations were unremarkable.

The infectious and tropical disease services were consulted. Both services highly suspected leprosy given the duration, painlessness, and atrophied, hypopigmented, and well-demarcated lesions. Suspicions were reemphasized at an outpatient visit, and the patient was scheduled for a biopsy for definitive diagnosis. The patient did not undergo the biopsy, and she left the country without treatment owing to stigma surrounding the illness.

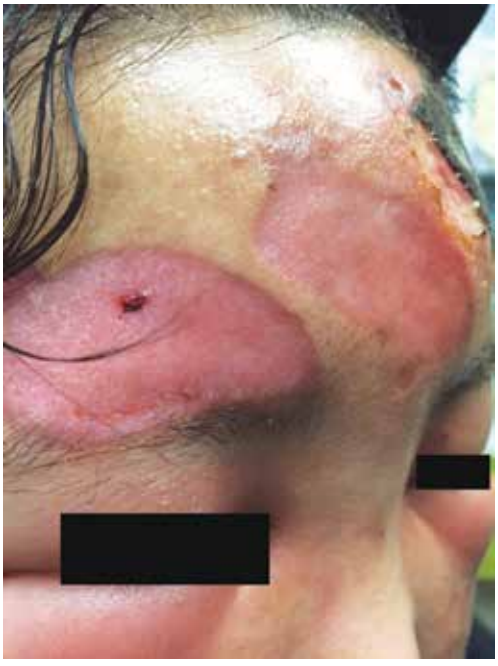
Editor's key points

- Leprosy might be more common than we realize, especially in communities with large immigrant populations.
- The variable presentations of leprosy and physician unfamiliarity with it make it difficult to diagnose, potentially resulting in treatment delays and increased morbidity.
- There is considerable stigmatization associated with leprosy, and it should be addressed in a culturally sensitive manner. Additionally, involvement of support groups and social work might be beneficial in managing the psychologic and social effects of the disease.

Points de repère du rédacteur

- La lèpre est peut-être plus courante qu'on le croit, surtout dans les collectivités où vivent d'importantes populations d'immigrants.
- Les présentations variables de la lèpre et le manque de familiarité des médecins avec la maladie compliquent le diagnostic, ce qui peut retarder le traitement et augmenter la morbidité.
- Une stigmatisation considérable est associée à la lèpre; il faut donc aborder le problème d'une manière culturellement adaptée. L'implication de groupes d'entraide et de travailleurs sociaux pourrait aussi être bénéfique dans la prise en charge des répercussions psychologiques et sociales de la maladie.

Figure 1. The patient presented with 5 well-demarcated eroded patches over the cheeks and forehead at various stages of healing without purulence or serous crusting



Discussion

PubMed and MEDLINE searches were completed using the MeSH terms *leprosy*, *mycobacterium*, and M leprae. Results were limited to articles published since 2000, with preference given to review articles. Article references were hand searched for relevant publications.

Leprosy might be more common than reports suggest.[11] Sample surveys of endemic areas have discovered several cases undetected by screening programs[12] and miscategorized owing to the use of non-standardized alternative classification systems.[13] Additionally, the World Health Organization proclaimed the global elimination of leprosy in 2000, which might have resulted in a decrease in detection activities, awareness, and training in its diagnosis.[11,14,15] This has potentially resulted in a growing number of undiagnosed cases.[16] Delays and misdiagnoses extend to North America, as cases without classic risk factors exist, including case reports of infections from armadillo exposure in the United States[17] and a Canadian man with no risk factors infected with *M lepromatosis*.[18]

The Ridley-Jopling classification divides leprosy into 3 main groups: tuberculoid, lepromatous, and borderline (**Table 1**).[5,6,19,20] These groups are related to the strength of the host's immune response to the obligate intracellular mycobacteria.[19,20] Patients with tuberculoid disease have a robust cell-mediated response and present with few skin lesions and no mycobacteria detectable on skin tests (paucibacillary).[5,19] Conversely, patients with lepromatous leprosy have a mostly antibody-mediated response and limited cell-mediated reaction, and present with multiple lesions and a high bacterial load (multibacillary).[5,19] Borderline subtypes lie between these groups, with patients having some degree of unstable immunity and multiple lesions.[19]

Risk factors for leprosy include low socioeconomic status,[5] genetic predisposition,[21] and exposure to affected household contacts.[22] Infectivity is generally low,[23] but increases with bacterial load.[24] Transmission likely occurs primarily through droplet contact with nasal mucosa.[5,20,24] The exact incubation period is unclear, but reports range from 6 months to 20 years (mean 2 to 4 years).[20]

The disease has an insidious onset and slow progression, and most commonly manifests with chronic inflammation of cutaneous and peripheral nerve tissue.[19,20] Dermatologic lesions often develop over cooler regions including the nose, earlobes, and testes, and might be macular, papular, nodular, or plaque lesions. Tuberculoid (paucibacillary) leprosy is characterized by asymmetric skin involvement with a few small, well-demarcated lesions with elevated borders associated with early development of sensory deficits.[25,26] Lepromatous (multibacillary) disease is often

Table 1. Characteristics of cutaneous leprosy: *Borderline subtypes lie between the paucibacillary and multibacillary groups, with patients having some degree of unstable immunity and multiple lesions.*

CHARACTERISTIC	PAUCIBACILLARY (TUBERCULOID)	MULTIBACILLARY (LEPROMATOUS)
World Health Organization definition[6]	1-5 patches associated with leprosy	>5 patches associated with leprosy
Bacterial count	None	Many
Severity	Mild	Can be extreme
Symmetry	Asymmetric	Symmetric
Borders	Well demarcated, elevated	Poorly demarcated
Infectivity	Very limited	Possibly more infectious
Prognosis	Good with treatment	Poor without treatment

Data from Reibel et al,[5] Rodrigues and Lockwood,[19] and Lastória and Abreu.[20]

symmetric,[27] with poorly demarcated skin lesions and late-onset sensory disturbances.[25,26]

Neurologic manifestations include nerve enlargement and dysfunction, and most commonly involve the great auricular, ulnar, radial, superficial fibular, and sural nerves.[25] Nerves with overlying skin lesions are most prone to involvement.[28] Grossly normal sensation can be seen with facial lesions, as in our patient, and might be related to the abundant innervation of the area.[29] Patients might initially experience loss of thermal sensation, followed by loss of fine touch and pain perception.[25,26] As in patients with diabetes, insensate areas are prone to injury, resulting in destructive deformities stereotypical of leprosy.[30,31] Autonomic and motor nerve involvement might present as anhidrosis, muscle atrophy, and weakness.[20,28]

Musculoskeletal involvement is common and can present as acute symmetrical polyarthritis affecting the small joints of the hands and feet.[27] Patients might develop chronic, symmetrical joint pain mimicking rheumatoid arthritis, with morning stiffness and positive test results for rheumatoid factor, affecting the wrists, metacarpophalangeal joints, proximal interphalangeal joints, knees, and metatarsophalangeal joints.[27] Ocular disease might progress despite treatment,[27] with up to 11% of cases resulting in blindness.[32,33] Other affected organs include the liver, lymph nodes, spleen,[25] testes, and ovaries.[34]

Diagnosis might be made clinically in endemic regions by physicians familiar with leprosy, but otherwise requires tissue biopsy for culture, histology, and polymerase chain reaction studies.[23] Patients require multidrug therapy, with regimens including rifampicin and dapsone for 6 months in paucibacillary disease, and rifampicin, dapsone, and clofazimine for 12 months in multibacillary disease.[5]

Conclusion

Diagnosing leprosy in Canada can be difficult owing to North American physicians' lack of familiarity with it. Multidrug regimens can provide effective therapy. Unfortunately, our patient returned to the Philippines before therapy owing to embarrassment and stigmatization. Her departure might have been avoided by more timely follow-up, and stigmatization might have been reduced through more thorough and culturally sensitive patient education. Additionally, involvement of support groups and social work might have been beneficial in managing the psychological and social effects of the disease.

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Competing interests
None declared

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References

1. Han XY, Mistry NA, Thompson EJ, Tang HL, Khanna K, Zhang L. Draft genome sequence of new leprosy agent *Mycobacterium lepromatosis*. *Genome Announc* 2015;3(3):pii:e00513-15.
2. Han XY, Aung FM, Choon SE, Werner B. Analysis of the leprosy agents *Mycobacterium leprae* and *Mycobacterium lepromatosis* in four countries. *Am J Clin Pathol* 2014;142(4):524-32.
3. Han XY, Sizer KC, Tan HH. Identification of the leprosy agent *Mycobacterium lepromatosis* in Singapore. *J Drugs Dermatol* 2012;11(2):168-72.
4. Han XY, Sizer KC, Velarde-Félix JS, Frias-Castro LO, Vargas-Ocampo F. The leprosy agents *Mycobacterium lepromatosis* and *Mycobacterium leprae* in Mexico. *Int J Dermatol* 2012;51(8):952-9.
5. Reibel F, Cambau E, Aubry A. Update on the epidemiology, diagnosis, and treatment of leprosy. *Med Mal Infect* 2015;45(9):383-93. Epub 2015 Oct 1.
6. World Health Organization. Global leprosy update, 2014: need for early case detection. *Wkly Epidemiol Rec* 2015;90(36):461-74.
7. Boggild AK, Correia JD, Keystone JS, Kain KC. Leprosy in Toronto: an analysis of 184 imported cases. *CMAJ* 2004;170(1):55-9.
8. Forno C, Häusermann P, Hatz C, Itin P, Blum J. The difficulty in diagnosis and treatment of leprosy. *J Travel Med* 2010;17(4):281-3.
9. Sermrittirong S, van Brakel WH. Stigma in leprosy: concepts, causes and determinants. *Lepr Rev* 2014;85(1):36-47.
10. Garbin CAS, Garbin AJÍ, Carloni MEOG, Rovida TAS, Martins RJ. The stigma and prejudice of leprosy: influence on the human condition. *Rev Soc Bras Med Trop* 2015;48(2):194-201.
11. Smith WC, van Brakel W, Gillis T, Saunderson P, Richardus JH. The missing millions: a threat to the elimination of leprosy. *PLoS Negl Trop Dis* 2015;9(4):e0003658.
12. Basel P, Pahan D, Moet FJ, Oskam L, Richardus JH. Leprosy incidence: six years' follow-up of a population cohort in Bangladesh. *Lepr Rev* 2014;85(3):158-69.
13. Pardillo FE, Fajardo TT, Abalos RM, Scollard D, Gelber RH. Methods for the classification of leprosy for treatment purposes. *Clin Infect Dis* 2007;44(8):1096-9. Epub 2007 Mar 5.
14. Scollard DM, Adams LB, Gillis TP, Krahenbuhl JL, Truman RW, Williams DL. The continuing challenges of leprosy. *Clin Microbiol Rev* 2006;19(2):338-81.
15. Burki T. Old problems still mar fight against ancient disease. *Lancet* 2009;373(9660):287-8.
16. World Health Organization. Global leprosy: update on the 2012 situation. *Wkly Epidemiol Rec* 2013;88(35):365-79.
17. Truman RW, Singh P, Sharma R, Busso P, Rougemont J, Paniz-Mondolfi A, et al. Probable zoonotic leprosy in the southern United States. *N Engl J Med* 2011;364(17):1626-33.
18. Jessamine PG, Desjardins M, Gillis T, Scollard D, Jamieson F, Broukhanski G, et al. Leprosy-like illness in a patient with *Mycobacterium lepromatosis* from Ontario, Canada. *J Drugs Dermatol* 2012;11(2):229-33.
19. Rodrigues LC, Lockwood DNJ. Leprosy now: epidemiology, progress, challenges, and research gaps. *Lancet Infect Dis* 2011;11(6):464-70.
20. Lastória JC, Abreu MA. Leprosy: review of the epidemiological, clinical, and etiopathogenic aspects—part 1. *An Bras Dermatol* 2014;89(2):205-18.
21. Mira MT, Alcaïs A, Nguyen VT, Moraes MO, Di Flumeri C, Vu HT, et al. Susceptibility to leprosy is associated with *PARK2* and *PACRG*. *Nature* 2004;427(6975):636-40. Epub 2004 Jan 25.
22. Sales AM, Ponce de Leon A, Düppre NC, Hacker MA, Nery JA, Sarno EN, et al. Leprosy among patient contacts: a multilevel study of risk factors. *PLoS Negl Trop Dis* 2011;5(3):e1013. Erratum in: *PLoS Negl Trop Dis* 2011;5(8).
23. Suzuki K, Akama T, Kawashima A, Yoshihara A, Yotsu RR, Ishii N. Current status of leprosy: epidemiology, basic science and clinical perspectives. *J Dermatol* 2012;39(2):121-9. Epub 2011 Oct 4.
24. Job CK, Jayakumar J, Kearney M, Gillis TP. Transmission of leprosy: a study of skin and nasal secretions of household contacts of leprosy patients using PCR. *Am J Trop Med Hyg* 2008;78(3):518-21.
25. Mackowiak PA, Schlossberg MC, Meyer CF, Kauh YC, Deng A. A 49-year-old man with a rash. *Am J Med Sci* 2005;330(3):134-8.
26. Talhari C, Talhari S, Penna GO. Clinical aspects of leprosy. *Clin Dermatol* 2015;33(1):26-37.
27. Tremblay F, Albert M. Arthritis, progressive maculopapular rash and severe peripheral neuropathy in a 60-year-old man. Diagnosis: leprosy. *CMAJ* 2014;186(3):206-9. Epub 2013 Dec 23.
28. Van Brakel WH, Nicholls PG, Das L, Barkataki P, Suneetha SK, Jadhav RS, et al. The INFIR Cohort Study: investigating prediction, detection and pathogenesis of neuropathy and reactions in leprosy. Methods and baseline results of a cohort of multibacillary leprosy patients in north India. *Lepr Rev* 2005;76(1):14-34. Erratum in: *Lepr Rev* 2005;76(3):264.
29. Britton WJ, Lockwood DNJ. Leprosy. *Lancet* 2004;363(9416):1209-19.
30. Daniel E, Rao PS, Courtright P. Facial sensory loss in multi-bacillary leprosy patients. *Lepr Rev* 2013;84(3):194-8.
31. MacMoran JW, Brand PW. Bone loss in limbs with decreased or absent sensation: ten year follow-up of the hands in leprosy. *Skeletal Radiol* 1987;16(6):452-9.
32. Courtright P, Daniel E, Sundar Rao PSS, Ravanes J, Mengistu F, Belachew M, et al. Eye disease in multibacillary leprosy patients at the time of their leprosy diagnosis: findings from the Longitudinal Study of Ocular Leprosy (LOSOL) in India, the Philippines and Ethiopia. *Lepr Rev* 2002;73(3):225-38.
33. Daniel E, Ffytche TJ, Sundar Rao PSS, Kempen JH, Diener-West M, Courtright P. Incidence of ocular morbidity among multibacillary leprosy patients during a 2 year course of multidrug therapy. *Br J Ophthalmol* 2006;90(5):568-73.
34. Singh RK, Bhasin R, Bisht YS, Kumar KV. Endocrine dysfunction in patients of leprosy. *Indian J Endocrinol Metab* 2015;19(3):369-72.

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