

Lepromatous Rhinitis an Unusual Presentation of Hansen's Disease: Case Report

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Background: Leprosy (Hansen's disease) is a chronic granulomatous infection caused by the bacteria *Mycobacterium leprae*. Classically leprosy affects the skin and peripheral nerves. The paranasal sinuses can also be infected and can serve as reservoirs for the reactivation of disease.

Case Report: A 31-year-old male patient, a private banker, presented to the authors ENT outpatient clinic with complaints of nasal obstruction, foul-smelling nasal discharge (ozaena), and intermittent epistaxis for the past one month, associated with multiple cutaneous nodules over the bilateral ear pinnae. Biopsy of the nasal mucosa revealed chronic granulomatous inflammation with multinucleated giant cells surrounded by abundant bacilli consistent with histoid leprosy.

Conclusion: Leprosy remains a major public health hazard as new cases are still being reported. The otolaryngologist should remain vigilant and have high index of suspicion for leprosy in patients with chronic nasal symptoms to prevent the potential morbidities that accompany this disease.

Keywords: Leprosy, Lepromatous rhinitis, Atrophic rhinitis, *Mycobacterium leprae*, Chronic rhinosinusitis

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Leprosy (Hansen's disease) is a chronic granulomatous infection caused by the bacteria *Mycobacterium leprae*. Classical leprosy affects the skin and peripheral nerves. The primary mode of transmission is thought to be through exposure to respiratory droplets containing *M. leprae*^(1,2). In 1994, the World Health Organization announced that Malaysia had eliminated leprosy. New cases of leprosy are rare in Malaysia, and the incidence is 0.76 per 100,000⁽²⁾. Nevertheless, the prevalence of leprosy remains high in endemic regions; worldwide, more than 200,000 new leprosy cases are detected annually⁽³⁾. Nasal and paranasal symptoms may occur early in the disease, often preceding the skin and

neural manifestations. Atrophic rhinitis is a known manifestation of the disease, characterized by mucosal crusting, ozaena, epistaxis, and paradoxical congestion of the nasal cavity. Secondary atrophic rhinitis may be caused by granulomatous infections including leprosy, tuberculosis, and syphilis, comprise approximately 1% of the total cases⁽⁴⁾. The authors presented a case of lepromatous leprosy with an unusual nasal presentation of ozaena and nasal blockage as the primary complaints.

Case Report

A 31-year-old male patient, a private banker, presented to the authors ENT outpatient clinic with complaints of nasal obstruction, foul-smelling nasal discharge (ozaena) and intermittent epistaxis for the past one month, associated with multiple cutaneous nodules over the upper limbs and bilateral ear pinnae (Figure 1). He attributed the skin lesion to scarring of an earlier episode of varicella zoster infection acquired two months prior to the onset of the skin nodules. Over the prior four months, he had visited several clinics for

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Figure 1. Right ear pinna with thickening of skin and multiple cutaneous nodules.

treatment of the skin nodules. He was prescribed oral and topical corticosteroids. The patient had no prior medical or surgical illnesses. He also denied contact with any leprosy patient or migrant workers, neither had he travelled to a leprosy-endemic region. On examination, the nose and overlying skin appeared to be normal. There was no depression over the dorsum of the nose, or a saddle nose deformity. Anterior rhinoscopy revealed excessive greenish-yellowish crusting and mucosal dryness. Nasoendoscopy revealed mucosal thickening, post-nasal drip, and crusting in the nasopharynx (Figure 2); however, there was no septal perforation. Ear examination revealed thickening of bilateral ear pinnae with several small nodules on the helix measuring 0.5×0.5 cm. Sensation over the pinna was reduced. The remainder of the ENT examination was unremarkable. Few subcutaneous nodules of various sizes, ranging from 0.5 to 1.5 cm in diameter were seen on the flexor surfaces of both forearms. Laboratory investigations, including complete blood count with differential, renal function tests and liver function tests were all normal. The erythrocyte sedimentation rate was elevated at 24 mm/hour. computed tomography (CT) of the paranasal

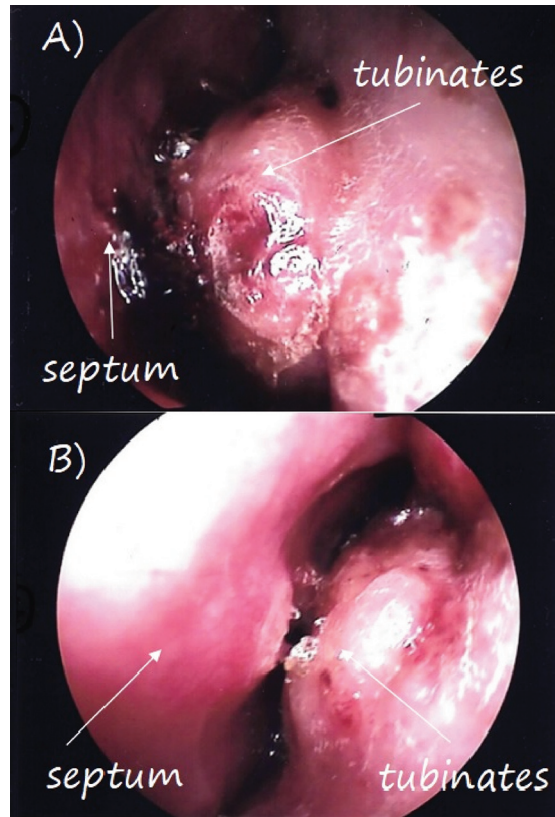


Figure 2. Nasoendoscopic view of the left and right nasal cavity. Nasal mucosa appears dry and atrophic with crusting seen over the bilateral turbinates and septal region.

sinuses revealed mucosal thickening at both maxillary sinuses, suggested of mucosal polyps. Biopsy of the nasal mucosa and punch biopsy of the skin lesion revealed chronic granulomatous inflammation with multinucleated giant cells surrounded by scanty lymphocytes and abundant bacilli consistent with histoid leprosy. Wade-Fite and Ziehl-Neelsen staining revealed numerous bacilli within and outside the histiocytes (Figure 3). Mycobacterium PCR was positive for non-tuberculosis mycobacteria. The patient was started on regimen according to the World Health Organization multidrug therapy (WHO-MDT) guidelines (1997): rifampicin 600 mg per month, clofazimine 300 mg per month, dapsone 100 mg daily, and clofazimine 50 mg daily as part of the intensive regimen. The patient was also given tualang honey for intranasal application. On follow-up two months later, the patient reported improved nasal congestion and nasal discharge. Endoscopic examination revealed near resolution of atrophic rhinitis.

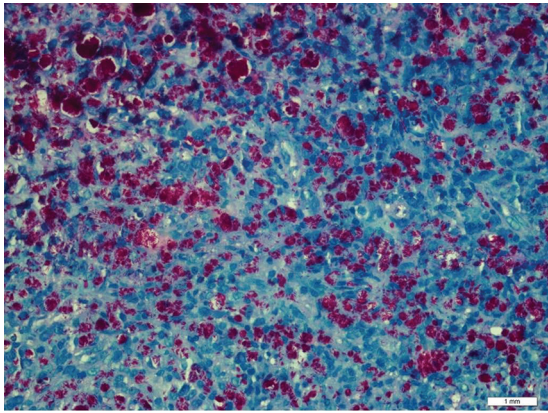


Figure 3. Ziehl-Neelsen staining of nasal mucosa with numerous bacilli within and outside the histiocytes.

Discussion

Leprosy is a chronic granulomatous disease caused by an obligate, intracellular aerobic, rod-shaped carbol fuchsin-positive acid-fast bacillus, *M. leprae*. The bacteria have tropism for skin, peripheral nerves, and the upper respiratory tract. Leprosy is classified into two types, tuberculoid and lepromatous. According to the World Health Organization, leprosy is classified into “paucibacillary” (low quantity) and “multibacillary” (large quantity) based on the number of skin lesions and proliferation of bacteria⁽³⁾. As demonstrated in the present patient, cutaneous lesion appeared four months prior to his nasal complains, however, the lesion was deemed atypical by his primary physician and therefore, treated as non-specific dermatitis with topical corticosteroids. High clinical suspicion and an early referral to the dermatologist by the primary health care professional could have prevented misdiagnosis and the delay of appropriate treatment. Histopathological studies of the nasal and skin biopsy reveal infiltration of macrophages, lymphocytes, and plasma cells with an abundance of acid-fast bacilli. The nose is the initial entry point for the bacilli via air droplets, and it is usually the initial site of the disease. However, the symptoms are mild in the initial stages of nasal involvement. Suzuki et al found that more than 90 percent of their treated ex-lepromatous leprosy patients reported feeling dryness, nasal obstruction, and crusting⁽¹⁾. Other common nasal complaints include nasal obstruction, epistaxis, crusting, ozaena, hyposmia, and nasal septal perforation. The paranasal sinuses can also be infected and serve as reservoirs for the reactivation of disease. The ethmoidal sinus is most commonly involved, and is reported to be infected in

up to 80 percent of cases⁽⁵⁾. The diagnosis is made by clinical examination and obtaining a thorough history, however, confirmation of disease is achieved by microscopic examination and histopathological studies of the skin and mucosal biopsies⁽¹⁻³⁾. Current standard treatment includes the use of three drugs, rifampicin, clofazimine, and dapsone. The multidrug therapy according to the WHO-MDT regimen (1997) aims to effectively eliminate the disease in the shortest time to prevent resistance⁽³⁾. Quality of life in this group of patients is extremely important as the disease causes after-effects including saddle nose deformity, nasal septal perforation, and atrophic rhinitis. Complications that can greatly affect quality of life⁽⁶⁾. Medical treatment should precede any surgical procedure, however, a combination of endoscopic sinus surgery and maxillary sinus irrigation can reduce the nasal symptoms of leprosy patients with chronic rhinosinusitis and atrophic rhinitis, thus, greatly improving quality of life⁽¹⁾. Menger et al reconstructed an ex-leprosy patients’ noses with saddle deformity using an external approach and achieved improved functional and aesthetic outcomes in most patients. The authors advocate reconstruction of saddle nose deformity with autologous concha cartilage grafts⁽⁷⁾. In our patient, there was no saddle nose deformity, therefore, no reconstructive surgery was performed. Atrophic rhinitis is a debilitating nasal condition characterized by progressive atrophy of the nasal mucosa, paradoxical congestion crusting, and foul-smelling nasal discharge (ozaena). Therefore, the triad of thick crusting of the nasal mucosa, foul odor, and nasal discharge should raise a suspicion of the diagnosis. The etiology of primary atrophic rhinitis remains unclear. However, secondary atrophic rhinitis is usually caused by sinus surgery (90%), radiation (2.5%), trauma (1%), granulomatous disease (1%), and infection (1%)⁽⁸⁾. Chronic granulomatous diseases such as leprosy, syphilis, tuberculosis, sarcoidosis, and Wegener’s granulomatosis are known to be associated with atrophic rhinitis⁽⁴⁾. Treatment of atrophic rhinitis remains debatable, as results of the majority of the proposed treatments are inconclusive with poor patient satisfaction. These treatments include nasal hygiene with regular intranasal irrigation of sodium bicarbonate solution, topical aminoglycoside therapy, and normal saline solution^(4,5). A study by Killera et al showed that honey was more effective in treatment of atrophic rhinitis than topical application of glucose and glycerin⁽⁹⁾. For the present patient, the authors prescribed topical application of locally-produced tualang honey as an adjunct along with sodium

bicarbonate solution nasal irrigation. This produced marked improvement in the patient symptoms of nasal dryness, crusting, and congestion.

Conclusion

Leprosy has been reported as eradicated in the majority of the developing countries including Malaysia. Nevertheless, it remains a major public health hazard as new cases are still being reported. This is partly due to the influx of the migrant workforce. The present case report emphasizes the importance that the otolaryngologist remaining vigilant and having high index of suspicion for leprosy in patients with chronic nasal symptoms to prevent the potential morbidities accompany this disease.

What is already known on this topic?

Leprosy (Hansen's disease) is a chronic granulomatous infection caused by the bacteria *M. leprae*. The pathogen has tropism towards the skin, peripheral nerves, and respiratory tract. The nasal and paranasal sinuses can also be infected and serve as reservoirs for the disease.

What this study adds?

Otolaryngologist should be vigilant and have high index of suspicion for leprosy in patients with chronic nasal symptoms to prevent the potential morbidities that accompany this disease.

Conflicts of interest

The authors declare no conflict of interest.

References

1. Suzuki J, Oshima T, Watanabe K, Suzuki H, Kobayashi T, Hashimoto S. Chronic rhinosinusitis in ex-lepromatous leprosy patients with atrophic rhinitis. *J Laryngol Otol* 2013;127:265-70.
2. Kwan Z, Pailoor J, Tan LL, Robinson S, Wong SM, Ismail R. Leprosy--an imported disease. *Lepr Rev* 2014;85:170-6.
3. World Health Organization. Global leprosy update, 2014: need for early case detection. *Wkly Epidemiol Rec* 2015;90:461-74.
4. Moore EJ, Kern EB. Atrophic rhinitis: a review of 242 cases. *Am J Rhinol* 2001;15:355-61.
5. Srinivasan S, Nehru VI, Mann SB, Sharma VK, Bapuraj JR, Das A. Study of ethmoid sinus involvement in multibacillary leprosy. *J Laryngol Otol* 1998;112:1038-41.
6. Soni NK. Microscopic rhinoscopy in lepromatous leprosy. *J Laryngol Otol* 1997;111:122-4.
7. Miller SH. Reconstructive surgery of the leprosy nose: a new approach. In: Miller SH, editor. *Yearbook of plastic and aesthetic surgery*. St Louis: Mosby; 2008. p. 46-8.
8. Chand MS, MacArthur CJ. Primary atrophic rhinitis: a summary of four cases and review of the literature. *Otolaryngol Head Neck Surg* 1997;116:554-8.
9. Killera S, Viswanatha B, Vijayashree MS. Efficacy of 25% glucose in glycerin and honey in the management of primary atrophic rhinitis: A comparative prospective study. *Res Otolaryngol* 2017;6:23-6.