

## Case Report

# Atypical ear lesion as initial presentation in Hansen disease

Akshaya Rathin Sivaji\*, Tony Kizhakkemuriyil Scaria

Department of Medicine, Panimalar Medical College Hospital and Research Institute, Poonamallee, Tamil Nadu, Chennai, India

**Received:** 02 September 2023

**Accepted:** 17 September 2023

### \*Correspondence:

Dr. Akshaya Rathin Sivaji,

E-mail: akshayrathin@gmail.com

**Copyright:** © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

## ABSTRACT

Hansen's disease is highly infectious and contagious. This disease in some individuals manifests with subtle symptoms such as isolated lesions like nodules over the ear, polyarthritis etc. These manifestations should be considered for the differential diagnosis of leprosy. We present this case with multiple cutaneous nodules over the helix and lobule of the right ear. A histopathological examination confirmed the diagnosis. Early detection and treatment of this disease prevent further complications.

**Keywords:** Buddha Ear, Hansen Disease, Mycobacterium Leprae, King's Ear

## INTRODUCTION

Leprosy also known as Hansen's disease is a chronic infectious disease caused by *Mycobacterium leprae*. Due to altered immune responses, it exhibits a wide spectrum of presentation. It commonly affects the skin, peripheral nerves, eyes and the upper respiratory tract, sometimes in advanced disease, it has a predilection to joints, lymph nodes and in some cases testes. The three cardinal signs of Hansen's disease are hypopigmented or erythematous skin lesion, loss of sensation or anaesthetic skin lesion and thickening of peripheral nerves.<sup>1</sup>

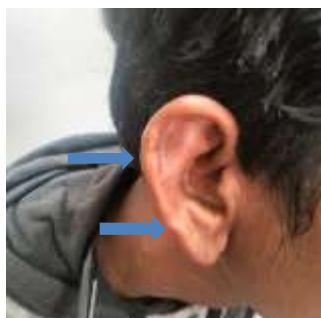
The true incidence of Hansen's disease is difficult to aggregate due to the insidious onset of initial signs and symptoms. According to the WHO 2021 report the registered global prevalence is about 1,33,781 with high rates reported in countries like India, Brazil and Indonesia.

Demographic patterns among countries vary due to geographic variations, socioeconomic status and healthcare programs in the countries. The involvement of the ear lobe in Hansen's disease during the initial stage is a rare occurrence, thus we are presenting this case.

## CASE REPORT

A 32-year-old male, a metro worker, residing in Chennai, India came to the medical outpatient department with complaints of multiple skin lesions in the right ear which has been gradually increasing in size for the past 8 months. Loss of sensation over the right ear for the past 3 months. Subsequently, the patient developed a skin lesion on the right side of the forehead. No recorded evidence of systemic hypertension, diabetes mellitus, seizure disorder, allergy, asthma and tuberculosis. The patient is not on any long-term treatment, steroid and immunosuppressive medications. No history of substance abuse, he is an occasional smoker and social drinker. The patient hailed from a low socioeconomic status. No recent history of any trauma. On general examination patient is moderately built and nourished, not anaemic, not jaundiced, no clubbing, no cyanosis, no pedal edema, no generalized lymphadenopathy and his vitals were within normal limits. On local examination, there are multiple cutaneous nodules sized about 0.5×0.5 cm present on the helix and lobule of the right ear (Figure 1). The nodules were tender, non-erythematous and hypo anaesthetic. On examination of the neck, there was a palpable thickening of the great auricular nerve on the left side (Figure 2). Routine blood

investigations were done, and all parameters were normal except raised ESR. Slit skin smear test and skin biopsy, were done.



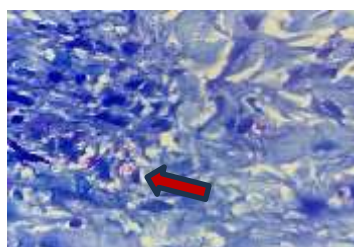
**Figure 1: Multiple cutaneous lesions tender and hypoesthetic in the right ear.**



**Figure 2: Nerve thickening- great auricular nerve.**



**Figure 3: Hypopigmented ovoid macule over the left side of the forehead.**



**Figure 4: Skin biopsy shows the presence of Lepromatous bacilli in the modified acid fast stain technique.**

## DISCUSSION

Leprosy is not uncommon in a well-advanced and protected healthy society. The manifestation of this disease occurs late due to the long incubation period, it is very difficult to identify the lesions at the earliest because the patients are unaware of the initial symptoms leading to delayed medical attention. Eventually, the patients come with florid symptoms and complications. Transmission of *Mycobacterium leprae* is highly possible after prolonged exposure to an infected individual rather than an acute transmission. In India, the annual case detection of Hansen's disease is 4.56 per 10,000 population.<sup>2</sup> But in comparison to the previous year the incidence has greatly reduced due to active early identification and intervention under the NLEP (National Leprosy Eradication Programme) launched in 1955. The classification of leprosy is done by two types: (a) WHO classification which is based on slit-skin smear. People with negative slit-skin smears are classified as paucibacillary leprosy and those with positive slit-skin smears are classified as multibacillary leprosy; and (b) Ridley and Jopling classified leprosy into six types from tuberculoid type to lepromatous type.

Leprosy has a predilection to the cooler areas of the body because low ambient temperature is required for its survival and proliferation. In this case report the patient had the involvement of his ear first and subsequent development of an ovoid hypopigmented macule over his right forehead (Figure 3). This nodular ear appearance is known as the 'Buddha Ear Sign'. This sign is more common in patients with relapse after completion of a multidrug regimen, but in this case, the Buddha Ear Sign presented as the initial finding. It is also known as king's ear, rat-bitten defect or mega-lobule deformity in leprosy.<sup>3</sup> To confirm the diagnosis slit-skin smear and skin biopsy were done. Slit skin smear with modified Ziehl - Nelson stain was positive for *Mycobacterium leprae* with a bacillary index of 3+. Biopsy showed epithelioid histiocytes showing lepromatous bacilli present as cigar-bundle shaped in modified acid fast stain with Fite-Faraco stain (Figure 4). The ear lobe is one of the easily accessible sites of the body, it can present in various forms such as nodules, ulcers and patches in the ear. The patient was diagnosed with lepromatous leprosy, paucibacillary type. Lepromatous leprosy characteristic features are erythematous nodules, madarosis (loss of hair over eyebrow or lashes) (Figure 3), and palpable thickening of the skin. In advanced disease it invades the nasal mucosa and mimics nasal stuffiness. If untreated it may lead to septal perforation or collapse (Saddle nose).<sup>5</sup> The patient was started on a multidrug regimen (dapsone 100 mg daily and a monthly dose of rifampin 600 mg) for 6 months.

## CONCLUSION

This case report makes the physician look for the atypical presentation of Hansen's disease. It should always be considered in the differential diagnosis of nodular lesions

of the external ear. Early detection of Hansen's disease reduces the risk of nerve damage, neural complications and disability.

*Funding: No funding sources*

*Conflict of interest: None declared*

*Ethical approval: Not required*

## REFERENCES

1. Loscalzo J, Fauci A, Kasper D, Hauser S, Longo D, Loscalzo J, et al. Harrison's Principles of Internal Medicine. 21st ed. New York, NY: McGrawHill; 1382-1392.
2. WHO. Supporting leprosy elimination in India, 2022. Available at: <https://www.who.int/india/news/02022supporting-leprosy->. Accessed on 22 August 2023.
3. Pruthi P, Munganda H, Bangia A, Rani U, Budhiraja R, Brajpuriya S. Leprosy with Atypical Skin Lesions Masquerading as Relapsing Polychondritis. Case Rep Infect Dis. 2016;2016:7802423.
4. Palaniappan V, Kaliaperumal K. Cauliflower Ear in Lepromatous Leprosy. Am J Trop Med Hyg. 2021;105(6):1444-5.
5. Scollard D, Stryjewska B, Dacso M. Leprosy: Epidemiology, microbiology, clinical manifestations, and diagnosis, 2023. Available at: <https://www.uptodate.com/contents/leprosyepidemiology-microbiology-clinicalmanifestationsdiagnosis>. Accessed on 22 August 2023.
6. Paudel V, Chudal D. Isolated Leprosy of Bilateral Ear Lobes, A Rare Presentation of Leprosy. Kathmandu Univ Med J (KUMJ). 2021;19(74):268-70.
7. Madke B, Nayak C. Eponymous signs in dermatology. Indian Dermatol Online J. 2012;3(3):159-65.
8. Kannan R, Kumar P, Vijayan BS, Priya YB. Lucio Leprosy in Neglected Cases of Hansen's Disease: A Series of Three Cases. JDCR. 2023;17(7):1-5.
9. Paudel D, Sah BP, Bhandary S, Shilpakar SL, Chettri ST, Mishra S. Isolated leprosy of pinna masquerading perichondritis: a rarest entity. American J Med Case Rep. 2014;2(4):87-9.
10. Bubna AK. Localized Glistening Nodules of the Right Ear: A Rare De novo Occurrence of Histoid Leprosy. Indian J Dermatol. 2016;61(6):694-6.

**Cite this article as:** Sivaji AR, Scaria TK. Atypical ear lesion as initial presentation in Hansen disease. Int J Res Med Sci 2023;11:3852-4.