Isolated Bilateral Pinna Swelling: A Rare Initial Presentation of Leprosy

Salma Yasmin Mohd Yusuf¹, Ilham Ameera Ismail¹, Rumihati Abdul Hamid², Nur Adliah Jamil¹, Mazapuspavina Md Yasin¹

¹Primary Care Medicine Discipline, Faculty of Medicine, Universiti Teknologi MARA, 47000 Sungai Buloh, Selangor, Malaysia; ²Putatan Health Clinic, Jalan Pusir Putih, 88200, Putatan, Kota Kinabalu, Sabah, Malaysia

Introduction

Leprosy, also known as Hansen disease, is a chronic, infectious disease caused by Mycobacterium leprae (M. leprae) [1]. Without timely treatment, leprosy leads to disfigurement, paralysed extremities and physical disabilities [2]. Much stigma has been associated with it and colonies of sufferers were isolated on islands and asylums in the effort to prevent the spread of the disease. M. leprae transmission occurs from prolonged close contact between susceptible and genetically predisposed individuals and untreated multibacillary patients. The nasal mucosa has been hypothesised as the main entry and exit route of M. leprae where the disease is thought to spread via respiratory droplets.

Leprosy has a systemic involvement of the dermatological, neurological and rheumatological systems [3]. Due to its systemic manifestations, it is difficult to differentiate leprosy from other systemic diseases. Incubation period also varies from 6 months to 20 years. Hence it is not uncommon for leprosy to be misdiagnosed, resulting in patients receiving late treatment [1]. The patient in this case report was subjected to repeated consultations to the healthcare professionals before a diagnosis could be established due to the unique presentation.

Up to date literature search using Pubmed/Medline, Cinahl database and Google Scholar using Mesh terms “Leprosy” [Mesh] OR “Leprosy, Multibacillary” [Mesh] OR ”Leprosy, Paucibacillary” [Mesh] OR ”Leprosy, Tuberculoid” [Mesh] OR ”Leprosy, Lepromatous” [Mesh] OR ”Leprosy, Borderline” [Mesh] revealed limited reported cases of isolated pinna swelling as an initial presentation of leprosy. Of these, five cases were of unilateral pinna involvement [3], [4], [5]. There was only one reported case of isolated bilateral pinna involvement [6].
Case Report

A 21-year-old male patient from Malaysia, with no significant past medical history, presented with complaint of painless erythematous bilateral pinna skin lesions for two weeks. He had no history of contact to any irritant or trauma and has no other skin lesions elsewhere. He was afebrile at the time of presentation and had a normal pulse rate and blood pressure. Dermatologic examination revealed erythematous and skin-coloured subcutaneous papules on both his pinna, which were painless (Figure 1). There were no other skin lesions or areas of paraesthesia on the body. Cardiovascular, respiratory and other system examinations were also normal. He was diagnosed to have infective perichondritis and received a course of broad-spectrum antibiotics, without any relief.

With the confirmation of diagnosis, the patient was treated with multi-drug therapy consisting of Rifampicin 600 mg, Clofazimine 300 mg and Dapsone 100 mg. Skin lesion over the chest resolved with gradual resolution of the ear lesions. Persistence of hyperpigmentation of the pinna, however, is a cause for despondency in the patient, warranting counselling.

Discussion

A disease once thought to be on the verge of becoming obsolete following the introduction of multi-drug treatment, leprosy is, unfortunately, still a public health concern in Malaysia and globally. The prevalence varies worldwide, but the resurgence of the incidence, particularly of multibacillary leprosy is a cause for concern [1], [7].

Leprosy has a wide spectrum of disease presentation, classified by Ridley and Jopling into six classes, ranging from tuberculoid pole on one end to the lepromatous pole on the other end [1], [3]. At the tuberculoid pole, the patients have cell-mediated immunity towards M. leprae, and elimination of mycobacteria can occur [3]. At the lepromatous pole, there is a lack of effective cell-mediated immunity to M. leprae, and thus bacilli proliferate [3]. The WHO classifies leprosy into paucibacillary and multibacillary based on the number of lesions [8]. Paucibacillary is less than 5 lesions, while multibacillary is more than 5 lesions [8].

Leprosy involvement of the ear pinna ranges from discrete nodules with minimal pain to ulceration with a “nibbled” defect [3], [9]. Megalobule of the pinna where the earlobe becomes greatly elongated or appear to wrinkle and hangs loose can also occur [9]. As mentioned, isolated pinna involvement, especially bilateral, without other systemic or cutaneous...
manifestation, is very rare.

Other differential diagnoses to pinna swellings include systemic conditions like sarcoidosis, relapsing polychondritis, and otophyma [3]. Perichondritis of the non-infective nature, for example, trauma, insect bites, piercings of the affected ear and leukaemic infiltration also cause similar pinna skin lesions [10]. Otitis externa or furunculosis of the external canal and malignant external otitis are infective causes of perichondriti that must be considered [10].

In conclusion, isolated involvement of pinna in a patient without lesions in other body parts is an unusual initial presentation of leprosy. However, leprosy should be kept as a rare differential diagnosis of perichondritis in patients not responding to conventional treatment.

Timely and proper implementation of treatment will break the chain of transmission and prevent disfigurement or physical disabilities that are responsible for the stigma associated with this disease.

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References

1. Lastória JC. Etiopathogenic aspects - Part 1. 2014; 89(2):205-18. https://doi.org/10.1590/abd1806-4841.20142450 PMid:24770495 PMCid:PMC4008049
2. Bauer B, Trautmann A. Pediatric dermatology photoquiz. 2015-4.
3. Pruthi P, Munganda H, Bangia A, Rani U, Budhiraja R, Brajputiya S. Case Report Leprosy with Atypical Skin Lesions Masquerading as Relapsing Polychondritis. 2016. https://doi.org/10.1155/2016/7802423 PMid:28116186 PMCid:PMC5220437
4. Ramos-e-silva M, Oliveira MLW, Munhoz-da-fontoura GH. Leprosy: uncommon presentations. 2005; 509-14. https://doi.org/10.1016/j.clindermatol.2005.01.012 PMid:16179185
5. Bubna AK. Localized Glistening Nodules of the Right Ear: A Rare De novo Occurrence of Histoid Leprosy. 2016; 61(6):193701-4. https://doi.org/10.4103/0019-5154.193703 PMid:27904201 PMCid:PMC512298
6. Liddell L, Holcomb M. Dermatologic Look-Alikes Nodules on the ear. (April 2015):65-70.
7. Kwan Z, Pai Loor J, Tan LL, Robinson S, Wong S-M, Ismail R. Leprosy-an imported disease. Lepr Rev. 2014; 85(3).
8. Gaschignard J, Grant AV, Thuc N Van, Orlova M, Cobat A, Huong NT, et al. Pauci- and Multibacillary Leprosy: Two Distinct, Genetically Neglected Diseases. 2016;1-20. https://doi.org/10.1371/journal.pntd.0004345 PMid:27219008 PMCid:PMC4878860
9. Ma S, Kovarik C. Erythema and induration on the right ear and maxilla. Cutis. 2016 Jul;98(1):28-9.
10. Lucena A, Espinson A. Acute atraumatic pinna (auricular) perichondritis. World J Emerg Med. 2018; 9(2):2017-8. https://doi.org/10.5847/wjem.j.1920-8642.2018.02.013 PMid:29576831 PMCid:PMC5847504