

Type 1 Lepra Reaction Presenting as Unilateral Perichondritis: A Case Report and Review of Literature

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Isolated involvement of external ear is rare in Hansen's disease. Ear involvement may present as infiltration, nodule formation, rat-bitten or cauliflower ear, mega lobule or as perichondritis. Herein, we present a case of a 65-year-old female who was being treated for infective perichondritis due to erythema and oedema over the right ear for two months. No other skin lesion was seen. Slit skin smear was negative. Histopathology from the lesion demonstrated a granuloma with epithelioid cells, giant cells and dense inflammatory infiltrate with no acid-fast bacilli. A diagnosis of borderline tuberculoid Hansen's disease was made and the patient was started on systemic steroids with multidrug therapy. We also discuss previous cases with isolated involvement of one or both the ears in leprosy. We wish to highlight that high index of suspicion of leprosy is needed in cases of auricular perichondritis who do not respond to antibiotic therapy.

Keywords : Perichondritis, Leprosy, Hansen's Disease, Lepra Reaction

Introduction

Hansen's disease is an infectious disease caused by acid-fast bacilli, *Mycobacterium leprae*, which primarily affects the superficial cooler areas of the body such as peripheral nerves, skin, nasal mucosa and external ear. The disease has a wide variety of manifestations, which have been classified as a spectrum based on the Ridley Jopling classification, with tuberculoid leprosy at one end to lepromatous leprosy at the other end with borderline forms in between. The chronic course of the disease is interrupted by episodes

of reactions that increases the risk of deformities and neuritis. A plethora of clinical manifestations seen in Hansen's disease sometimes leads to diagnostic difficulties. Involvement of the external ear is common in leprosy as it is one of the cooler parts of the body and commonly presents as infiltration seen in lepromatous leprosy (Yusuf et al 2019). Isolated involvement of the pinna is, however, very uncommon. We report a case of borderline tuberculoid Hansen's disease with Type 1 lepra reaction in a lady with isolated unilateral involvement of the pinna,

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initially leading to misdiagnosis of perichondritis.

Case Report

A 65-years-old female, native and resident of western Uttar Pradesh, was referred from the Department of Oto-Rhino-Laryngology for dermatological opinion regarding erythema and swelling in the right ear for a duration of 2 months. She was a known case of type 2 diabetes mellitus and was on oral hypoglycaemic agents. She was being treated with oral antibiotics for perichondritis of the right ear for her complaints, but there was no relief. There was no history of trauma to the site, fever, loss of sensations at any site over skin. Family history was not significant.

On cutaneous examination, an oedematous, infiltrated, and erythematous plaque was present over the right pinna (Fig.1). Mild tenderness was



Fig. 1 : Erythema and oedema over the right pinna.

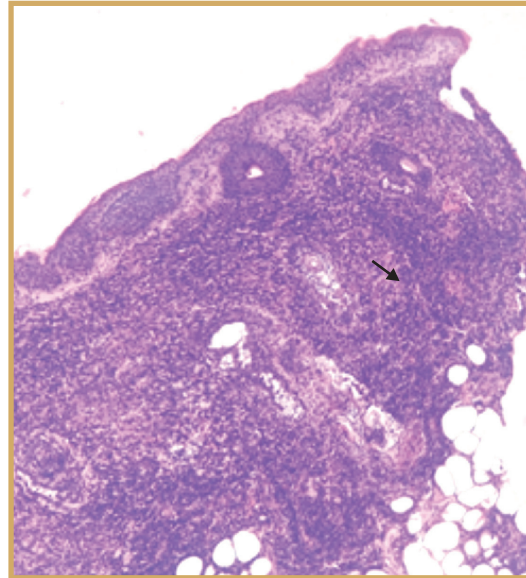


Fig. 2a : Normal epidermis with ill formed non caseating granulomas and chronic nflammatory infiltrate in the dermis (H&E, 10X)

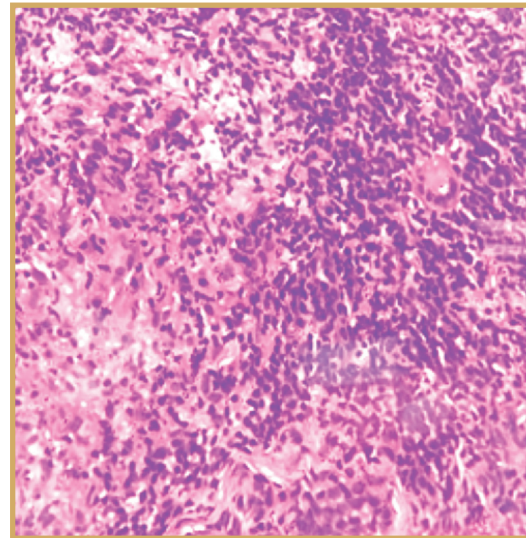


Fig. 2b : Ill-defined non-caseating granulomas with epithelioid cells and Langhans giant cells in the dermis. (H&E, 40X)

seen over the plaque. The left ear was completely normal. Sensory and motor examination was within normal limits. No nerve thickening or tenderness was found.

Slit skin smear from right ear was negative. Punch biopsy from right ear was sent for histopathological examination which revealed clusters of epithelioid cells along with giant cells and dense chronic inflammatory infiltrate in the dermis (Figs. 2(a), 2(b)). AFB was negative. Based on cutaneous examination and investigations, a diagnosis of borderline tuberculoid Hansen's disease with Type 1 lepra reaction was made.

Patient was started on systemic steroids and Paucibacillary multidrug therapy. There was marked improvement in erythema and oedema over a period of 1 week. Currently the patient is being followed up with no exacerbation of reaction.

Discussion

Hansen's disease is characterised by a chronic course and varied presentation. Due to its variable incubation period and varied systemic and cutaneous manifestations, it is often misdiagnosed resulting in delayed treatment and increased risk of neurological and functional impairment. Involvement of external ear can take place in varied clinical forms such as infiltration, nodule formation, "nibbled" or "rat-bitten" appearance, mega lobule, and auricular chondritis (Palaniappan & Kaliaperumal 2021). Majority of patients present with bilateral involvement of ear with skin lesions elsewhere on the skin as well. Isolated involvement of one or both ears is rare in leprosy.

Our patient was misdiagnosed as a case of perichondritis. Yusuf et al. described erythematous to skin coloured asymptomatic papules over both the pinnae including lobules in a 21-year-old male from Malaysia developing over two years. He was misdiagnosed as a case of infective perichondritis and was advised

cosmetic surgery. He was diagnosed as a case of lepromatous leprosy after slit skin smear and histopathology (Yusuf et al 2019).

Paudel et al (2014) reported a case of a 15-year-old male from Nepal with complaints of pain and erythematous papules over the left pinna for 15 days. History of similar lesions was present which occurred one year back. He was misdiagnosed as a case of perichondritis. Correct diagnosis was made after aggressive treatment was ineffective and a biopsy was done which showed high bacillary index of 4+.

Pruthi et al (2016) have also reported about a 21-year-old male who presented with one month history of fever, pain in small joints of hands, erythematous, scaly ulcerative rash over extensors of elbows and redness of upper two-thirds of both pinna with sparing of the lobules. There was generalized weakness, loss of weight and loss of appetite. A diagnosis of relapsing polycondritis was considered. However, skin biopsy showed ill formed granulomas and acid-fast bacilli. The patient was diagnosed to have borderline lepromatous leprosy and was relieved after 6 months of multi-drug therapy (Pruthi et al 2016).

Isolated unilateral involvement of the ear has been rarely described in literature. Palaniappan & Kaliaperumal (2021) described a case of lepromatous leprosy in a 49-year-old male who presented with skin-coloured, non-tender asymptomatic papules and nodules over the left ear involving the tragus, helix and lobule mimicking a cauliflower ear. On further examination, hypochromic macules over body and thickened ulnar nerve were found. Biopsy was consistent with a diagnosis of lepromatous leprosy (Palaniappan & Kaliaperumal 2021).

A case of histoid leprosy with only three smooth, flesh-coloured nodules over helix and lobule of one external ear was recorded by Bubna in a 21-year-old male (Bubna 2016). Shilpa et al. also

described a case of unilateral cauliflower ear in a male patient due to multibacillary leprosy (Shilpa et al 2016). Ramos-e-Silva et al (2005) reported several atypical cases of leprosy which included a 29-year-old male with a single erythematous plaque over the helix of left earlobe which had anaesthesia.

Isolated involvement of both the pinnae is also rarely seen in Hansen's disease. Narayanan & Akshay (2022) described a case of lepromatous leprosy who presented with complaints of thickening and nodularity of both the ear lobes. However, diffuse infiltration of the face and madarosis were found on examination (Narayanan & Akshay 2022). Paudel and Chudal from Nepal reported lepromatous leprosy in a 25-year-old male who presented with bilateral papulonodular lesions over both pinnae including the helix and lobule, with no skin lesions elsewhere. Slit skin smear showed high bacillary positivity of 6+ (Paudel & Chudal 2021).

Conclusion

Our case is unique as the patient presented with unilateral involvement of the pinna, which led to the incorrect presumptive diagnosis of infective perichondritis by the otorhinolaryngologist. The patient was found to have borderline tuberculoid leprosy with type 1 lepra reaction. Thus, a high index of suspicion is required for timely diagnosis and treatment in order to prevent deformities which are more common in untreated reactional states.

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