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Isolated Leprosy of Pinna Masquerading Perichondritis : A Rarest Entity

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Abstract Leprosy is a chronic bacterial disease caused by *Mycobacterium leprae*. It is the oldest known disease to human beings. Incubation period ranges from weeks to years. The prevalence of leprosy varies markedly from country to country. However, the overwhelming majority of cases are found in developing countries and 92 percent of the cases are detected in just 11 countries led by India and Brazil. Grossly leprosy is divided into lepromatous and tubercular form, based on immunological pattern of disease. Though leprosy is common condition, isolated leprosy of pinna has not been reported so far in the literature. We report a male of 15 years with clinical feature of perichondritis of left pinna due to leprosy.

Keywords: *leprosy, perichondritis, pinna*

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1. Introduction

Leprosy is an ancient deforming disease caused by *Mycobacterium leprae*, which is still poorly understood and often feared by the general public and even by some in the health care professions. Fortunately, the outlook for patients has dramatically improved over the last three decades with the introduction of multi-drug treatment and management strategies that have somewhat diminished the stigma of this diagnosis. [1,2] An estimated 1.34 million cases of leprosy occur worldwide. [3] The prevalence of leprosy varies markedly from country to country. However, the overwhelming majority of cases are found in developing countries, and 92 percent of the cases are detected in just 11 countries led by India and Brazil. [4] Globally, Nepal is among the six major endemic countries which account for 23% of all new cases detected during 2005 and 24% of registered cases at the beginning of 2006. [5] Leprosy is a chronic infectious disease caused by *Mycobacterium leprae.* Leprosy exhibits a wide spectrum of presentation, varying from the tuberculoid (TT) to the lepromatous pole, with the immunologically unstable borderline forms in-between, depending upon the immune status of the individual. At the lepromatous pole, the patients lack effective cell-mediated immunity to *M. leprae* and bacilli proliferate, while at the tuberculoid pole, the patients have cell mediated immunity towards *M. leprae* and there is elimination of mycobacteria. *Mycobacterium leprae* is a unique organism with capacity to invade not only Schwann cells but also other parenchymal tissues such as testis, lymph node, larynx, liver, bone, muscle and cartilage. [3] Though pinna is one of the sites for taking for slit skin smear as it harbors the *Mycobacterium leprae* being cooler place, but isolated lesion of leprosy of pinna has not been reported so far in the literature. To the best of our knowledge this is the first case of isolated leprosy of pinna.

2. Case

A 15 years male from Terai region of Nepal came to our ENT OPD with complain of pain and small multiple pinkish lesions in left pinna for 15 days. The lesions were associated with mild dull aching, continuous pain and itching sensation. There was no history of fever and trauma to the ear or any similar lesions in other parts of the body. There was history of similar lesion 1 year back which was relieved by taking oral antibiotics. Physical examination revealed thickening of skin over pinna with maculopapular rashes and telangiectesia.

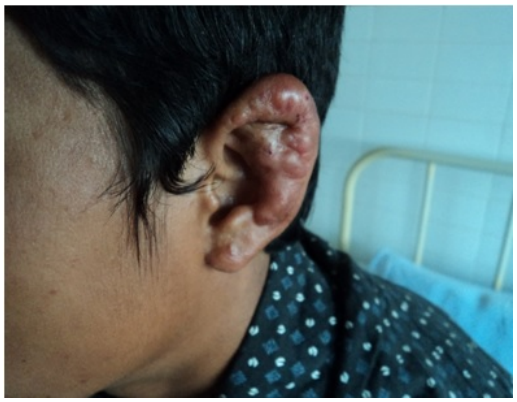

Figure 1. Lesion of left pinna before treatment

On clinical basis we made provisional diagnosis of the perichondritis of left pinna and patient was treated with intravenous ciprofloxacin for two weeks. Despite aggressive treatment patient did not improve and hence we sent tissue biopsy from left pinna. Biopsy specimen showed numerous acid fast bacilli, macrophages and foamy cells suggestive of lepromatous leprosy with bacillary index of 4.

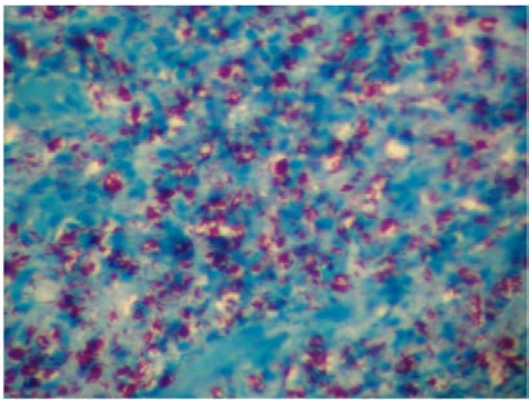

Figure 2. Histological section showing golbi of acid fast bacilli (AFB stain; X 100 magnification)

On the basis of this, diagnosis of Borderline tubercular Hansen's disease was made and the patient was treated with multibacillary (MB) Multi Drug Therapy (MDT) regimen comprising Rifampicin 600 mg once a month, Clofazimine 300 mg once a month followed by 50 mg daily and Dapsone 100 mg daily for 12 months.

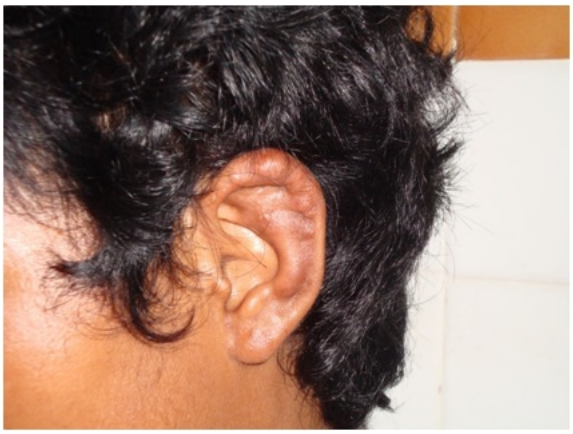

Figure 3. Resolving lesion of left pinna after 1 month of starting treatment

After one month of treatment on follow-up patient was well, lesion of pinna was disappearing. The lesions disappeared after completion of the treatment at 12 months.

3. Discussion

Leprosy is an ancient deforming disease caused by *Mycobacterium leprae*, which is still poorly understood and often feared by the general public and even by some in the health care professions. Fortunately, the outlook for patients has dramatically improved over the last three decades with the introduction of multi-drug treatment and management strategies that have somewhat diminished the stigma of this diagnosis. Leprosy is a chronic infectious disease caused by *Mycobacterium leprae* and is an ancient scourge that still affects ≥ 4 persons per 10,000 in Brazil, India, Madagascar, Mozambique and Nepal. Globally, Nepal is among the six major endemic countries which account for 23% of all new cases detected during 2005 and 24% of registered cases at the beginning of 2006. [5] The patient belongs to endemic zone of leprosy.

Mycobacterium leprae infection usually presents with cutaneous and neurological involvement. However, initial clinical manifestation in muscle, lymph node, larynx, liver, spleen, bone marrow, bone and testis (proven by biopsy) has been infrequently reported. [6] Isolated leprosy of pinna is not yet reported in literature and one of the rare form of disease. In our case the disease presented as a diagnostic dilemma and delayed the appropriate treatment of the patient.

Leprosy exhibits a wide spectrum of presentation, varying from the tuberculoid to the lepromatous pole, with the immunologically unstable borderline forms in-between, depending upon the immune status of the individual. At the lepromatous pole, the patients lack effective cell-mediated immunity to *M. leprae* and bacilli proliferate, while at the tuberculoid pole, the patients have cell mediated immunity towards *M. leprae* and there is elimination of mycobacteria. On this basis different form of leprosy are categorized. In 1997, a case of leprosy was defined as an individual who has not completed a course of treatment and has one or more of the three cardinal signs: [7]

- Hypopigmented or reddish skin lesions with loss of sensation
- Involvement of the peripheral nerves as demonstrated by their thickening and associated loss of sensation
- Skin-smear positive for acid-fast bacilli

Leprosy can present as a purely neural disease without skin lesions; the so-called neuritic leprosy. Nerve biopsy is confirmatory. The incidence is 0.5% in Ethiopia, 4.6% in India and 8.7% in Nepal [2].

The current WHO recommended multi drug treatment (MDT) for adults is as follows: [8]

1. For paucibacillary disease (PB), Rifampicin: 600 mg once a month Dapsone: 100 mg daily Duration = 6 months
2. For multibacillary disease (MB), Rifampicin: 600 mg once a month Dapsone: 100 mg daily Clofazimine: 300 mg once a month and 50 mg daily Duration = 12 months
3. For single PB lesion, rifampin 600 mg, ofloxacin 400 mg, minocycline 100 mg a single dose

Leprosy in otorhinolaryangology isn't unusual disease. Oral cavity and nose are the most common sites. It can present rarely in larynx, epiglottis being the most common site and sometimes requiring emergency tracheostomy. [9] Leprosy also can present as facial nerve paralysis to otolaryangologists. Our case belongs to one of the rarest form of leprosy, only involving the pinna, and presenting as perichondritis. The clinical presentation of our patient was consistent with TT, but based on the evidence from histopathology and bacteriology, the patient was shown to be borderline tuberculoid and was treated as multi bacillary regimen and we got a good response.

4. Conclusion

Thickening of skin, painful lesion over pinna are common complaints especially following trauma. Though perichondritis of pinna due to leprosy in a patient without lesions in other body parts is unusual presentation, leprosy should be kept as a rare differential diagnosis of perichondritis in patient not responding to conventional treatment if he belongs to endemic zone.

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