CASE REPORT

Difficult diagnosis and challenging treatment – a report on leprosy

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Accepted for publication 4 August 2016

Summary  Histoid leprosy is an unusual variant of lepromatous leprosy posing difficulties for diagnosis as well as treatment even to the experts. We report a case of a 73 year old male from the hilly region of Nepal, who presented with multiple asymptomatic scrotal nodules for 3 months. Though clinical diagnoses were pilomatrixoma and steatocystoma multiplex; histopathological examination was consistent with that of histoid leprosy. After 10 months, he had unusual Type 2 lepra reaction with constitutional symptoms, perichondritis, epididymo-orchitis but without erythema nodosum leprosum. Two years later, his three family members were also affected with leprosy. Hence, this case is reported to highlight the diagnostic dilemma, complicated disease course and infectivity of histoid leprosy.

Keywords: Bacillary index; Epididymo-orchitis; Histoid leprosy

Introduction

‘Histoid leprosy’, first described by Wade, is an unusual variant of bacilliferous leprosy with classical clinical, histopathological and bacteriological findings with 3-6% incidence.1 Its rarity demands a high level of suspicion even to expert eyes. Because of the diagnostic dilemma and the highly bacilliferous nature, an affected person may transmit the disease to contacts.

Case Report

A 73 year old male from the hilly region of Nepal, presented to the dermatology Outpatient Department with multiple, gradually progressing, asymptomatic, scrotal lesions for 3 months.
Examination revealed multiple, erythematous, round-oval, some discrete and some coalesced nodules over the scrotum (Figure 1). They were smooth, 0.5 × 0.5 cm-2 × 2 cm, non-tender and firm. On clinical bases, a differential diagnose of pilomatricoma and steatocystoma multiplex were kept. However, histopathological findings were of histoid leprosy with 6+ bacillary index (BI) (Figure 2); hence, diagnosis of histoid leprosy was made.

After 10 months of treatment with multibacillary multi drug therapy (MB-MDT); he had an unusual Type 2 lepra reaction (T2R) with constitutional symptoms, perichondritis, epididymo-orchitis, bilateral pedal edema and tender inguinal lymph nodes (Figure: 3); but without erythema nodosum leprosum (ENL) and neuritis.
FNAC from the scrotum showed features of leprosy. This episode was successfully treated with systemic corticosteroids for 8 weeks. His skin lesions resolved completely (Figure 4) and BI = 0 on slit skin smear examinations at 24 months. He was completely normal with no disease related complications even after 3 years of treatment. However, three of his family members had leprosy after 2, 3 and 5 years of his successful treatment.

Discussion

Histoid leprosy is a rare variant of bacilliferous leprosy with classical clinical, histopathological and bacteriological findings. The term ‘histoid leprosy’ was first described by Wade in 1960. Though the exact prevalence of histoid leprosy in not known in our country, in India, it ranges from 1.8–3.6%. Though the disease is more common in third to fifth decades, occasionally, extremes of the ages can also be victimised as in our case. Common causes of histoid leprosy are reported to be dapsone monotherapy, irregular or inadequate treatment. But these days, there are few reports even on de novo presentation without a past history as in our case. Since leprosy had already been eliminated from our
country, occurrence of *de novo* histoid leprosy in post elimination era also raises a genuine question, “are we able to diagnose all leprosy patients?” Moreover, the difficulty in detecting this infectious condition might raise a question on the sustainability of elimination. So, we would like to emphasise the high level of suspicion for disease diagnosis.

Site most usually affected are the face, back, buttocks, extremities, and bony prominences.\(^4\) Scrotal skin involvement is rare with only a few published case reports.\(^9,10\) Type 2 lepra reaction (T2R), an acute complication mainly seen in lepromatous leprosy, is also called ENL because it is the most common skin manifestation.\(^11\) Though it is possible to have T2R without ENL, till now only one case report has been reported from India.\(^12\) T2R is rarely seen in histoid leprosy. Bhutani et al. had reported three cases of T2R among 20 histoid patients;\(^5\) but there is no report of T2R without ENL in histoid leprosy. Since our patient of histoid leprosy had an unusual episode of T2R reaction with fever, constitutional symptoms, bilateral pedal edema, perichondritis, epididymo-orchitis and enlarged tender inguinal lymph nodes but without ENL, we believe this is the first published case report.

### Conclusion

This case report is to highlight the diagnostic dilemma, complicated disease course and infectivity of histoid leprosy in a leprosy eliminated country.

### References