A Clinico-Epidemiological Profile of Histoid Leprosy in a Tertiary Care Hospital - North India

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Abstract

Background: Histoid leprosy (HL) cases are a reservoir of disease as it is a rare variant of lepromatous leprosy lead to further spread of leprosy. Therefore, early diagnosis and management of these cases needed to eradicate leprosy. The main objective of our present case series is to show different clinical features, histology findings and bacteriological findings in patients who attended tertiary care hospital outpatient department over last 6 years.

Methods: It is a retrospective 6 year study (January 2014-January 2019). Particular of patients were obtained and clinical findings are noted in histoid leprosy patients in Mahatma Gandhi Medical College and Hospital, Jaipur, Rajasthan (India).

Results: In this study, there were 7 cases of HL. All cases were males in the age group ranging from 30 to 65 years with shorter duration of illness. 3 patients (42.80%) were de novo cases of HL. Nodule (71.5%) were the commonest skin lesions while arm (78%) were the most common sites involved. Earlobe infiltration was seen in 2 cases (28.5%). Lepra reactions were seen in one (14.2%) cases. No deformity/disability were noted. Most common nerve involved was ulnar nerve. The bacteriological index (B.I) of Histoid leprosy, +3/+4 were mainly seen in borderline leprosy (BB, BL), B.I +1/+2 mainly in borderline tuberculoid (BT) leprosy.

Conclusion: In this study majority of cases were de novo three out of seven. It also develop in patient who took inadequate or irregular treatment. So, early detection of Histoid Hansen is needed for awareness and to eradicate leprosy.

Keywords: Histoid Leprosy, De novo, Clinical profile.

INTRODUCTION

Histoid hansen’s disease is an uncommon form of lepromatous leprosy (LL), first described by Wade [1] in 1963 with high bacillary load, which clinically present as spontaneous asymptomatic multiple discrete, smooth, shiny, dome shaped, nontender, skin coloured papules or cutaneous or subcutaneous nodule and plaque over apparently normal looking skin of lepromatous patients, who received irregular or inadequate treatment or on dapsone monotherapy rarely de novo [2]. In India, National leprosy control programme (NLCP) was launched in 1954 and in 1983, it was switched to National leprosy Elimination programme (NLEP). India has achieved the leprosy elimination goal in 2005. Recently several cases of histoid leprosy were reported including Bhukya & Reddy et al. [3], Nair et al., [1], Meena et al., [4], R Subha et al., [2]. The main objective of our present case series is to show different clinical features, histology findings and bacteriological findings in tertiary care hospital who had attended outpatient department over last 6 years.

MATERIAL AND METHODS

This is an 6-year duration retrospective study. In our study we included all the cases of leprosy who attended the leprosy centre during study period of 6 year duration (January 2014 to January 2019) were included, among them seven cases of Histoid Hansen were clinically diagnosed and studied. The data were recorded in a performa such as age, sex, duration of illness, previous treatment taken, history of treatment and family history, mode of acquired transmission. Clinical examination including general physical examination and palm, soles, hair, nail and mucous membrane were examined and positive findings were noted. A thorough cutaneous examination including lepra reaction and peripheral nerve examination (for...
thickness, tenderness or any abscess) were noted. Deformities like facial palsy, claw hand, wrist drop, foot drop, plantar ulcer according to WHO grading were noted. Slit skin smear (SSS) and histopathological slides were also reviewed and were treated with MB MDT.

RESULTS
The incidence of Histoid Leprosy among the registered patients was 3.41 % (7 of 205) of all leprosy cases, during 6 year of duration (June 2014 to January 2019). All cases were males in the age group ranging from 30 to 65 years with shorter duration of illness.

Out of these, three patients were de novo cases (development of Histoid lesion without previous evidence of Hansen’s disease), one patient have finding of LL with histoid with type 1 reaction and Earlobe infiltration, one patient who was diagnosed as BL- leprosy, took irregular treatment and presented with histoid lesion after 3 year, interestingly his two brothers were also diagnosed as BT-leprosy and one patient had prior inadequate treatment with dapsone for few months.

Cutaneous and subcutaneous nodules were the commonest skin lesions found in 5 patients (71.5%). Papules in 4 patients (57.1%), plaques in 3 patients (42.8%) hypo pigmented patches in 3 patients (42.8 %) and No mucosal lesions and ulcerations were seen in any of our patients (Figure-1).

The commonest anatomical site involved as arms (78%) followed by back (67%), abdomen (45%), thigh (45%), forearms (36%), chest (27%), face (21%), legs (20%), hands (18%) and elbow (9%) (Table-1).

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Skin lesions</th>
<th>Site involved</th>
<th>Nerve involvement B/L</th>
<th>Trophic change</th>
<th>Glove &amp; stocking anesthesia</th>
<th>Deformity</th>
<th>De novo/ LL with Histoid</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Nodules, plaques</td>
<td>Arms, back, thigh, face</td>
<td>Ulnar, LPN</td>
<td>_</td>
<td>_</td>
<td>_</td>
<td>De novo</td>
</tr>
<tr>
<td>2.</td>
<td>Nodule, papules</td>
<td>Arm, forearm, abdomen, face</td>
<td>Ulnar, RCN, GAN</td>
<td>_</td>
<td>_</td>
<td>_</td>
<td>De novo</td>
</tr>
<tr>
<td>3.</td>
<td>Hypopigmented patches, nodule, papule</td>
<td>Back, legs, face, abdomen</td>
<td>Ulnar, LPN</td>
<td>_</td>
<td>_</td>
<td>_</td>
<td>De novo</td>
</tr>
<tr>
<td>4.</td>
<td>Nodule, papules</td>
<td>Face, forearm, arm, legs, back</td>
<td>GAN, Ulnar, Median, LPN, PTN</td>
<td>_</td>
<td>+</td>
<td>_</td>
<td>LL with histoid</td>
</tr>
<tr>
<td>5.</td>
<td>Nodule, hypopigmented patches</td>
<td>Face, back, abdomen, elbow</td>
<td>Ulnar, median</td>
<td>_</td>
<td>+</td>
<td>_</td>
<td>BL with histoid</td>
</tr>
<tr>
<td>6.</td>
<td>Plaque, Hypopigmented patches</td>
<td>Chest, abdomen, legs, thigh</td>
<td>LPN, Ulnar, PTN</td>
<td>_</td>
<td>_</td>
<td>_</td>
<td>BT with histoid</td>
</tr>
<tr>
<td>7.</td>
<td>Papule, plaques</td>
<td>Legs, hands, forearm, back</td>
<td>Ulnar, Median, LPN, PTN</td>
<td>_</td>
<td>_</td>
<td>_</td>
<td>BT with histoid</td>
</tr>
</tbody>
</table>

On nerve examination of all patients there were thickening of two or more peripheral nerves in which Ulnar nerve was the most commonly affected which was followed by lateral popliteal nerve. Two patients had glove and stocking type of anesthesia with sensory loss. No deformity were noticed in any patient.

Peripheral nerve were thickened in 6 (85.7 %) and tenderness present in 5 (71.4%) patients, nerve abscess not found in these patients. Most common nerve involved was ulnar nerve in all 7 patients followed by lateral popliteal nerve in 5 (71.4%) patients (Table-2).

<table>
<thead>
<tr>
<th>Peripheral nerve</th>
<th>Thickened</th>
<th>Tender</th>
<th>Nerve abscess</th>
</tr>
</thead>
<tbody>
<tr>
<td>Great auricular</td>
<td>2</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Radial</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Ulnar</td>
<td>7</td>
<td>5</td>
<td>-</td>
</tr>
<tr>
<td>Median</td>
<td>3</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Common peroneal</td>
<td>5</td>
<td>2</td>
<td>-</td>
</tr>
<tr>
<td>Sural</td>
<td>0</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Posterior tibial</td>
<td>2</td>
<td>-</td>
<td>-</td>
</tr>
</tbody>
</table>

In all patients, slit skin smear (by Ziehl Neelsen stain) revealed abundant uniformly stained organisms with tapering ends seen singly or in clusters. The bacteriological index (B.I) of Histoid leprosy, +3/ +4 were mainly seen in borderline leprosy.
(BB, BL), B.I +1/ +2 mainly in borderline tuberculoid (BT) leprosy (Table-3).

### Table-3: Bacteriological index

<table>
<thead>
<tr>
<th>Bacteriological index</th>
<th>0</th>
<th>+1</th>
<th>+2</th>
<th>+3</th>
<th>+4</th>
<th>+5</th>
<th>+6</th>
</tr>
</thead>
<tbody>
<tr>
<td>Histoid leprosy</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>1</td>
<td>1</td>
<td>3</td>
<td>2</td>
</tr>
</tbody>
</table>

Histopathology showed thinned out epidermis, positive Fite-Faraco stain for lepra bacilli, thin spindle-shaped histiocytes forming interlacing bands, Grenz zone (band of una) and collection of spindle shaped histiocytes arranged in a whorl like pattern.

Histoid lesion have lymphocytes and polymorphonuclear leukocytes at periphery of nodules, Acid fast bacilli in histoid lesion is longer than ordinary leprosy bacilli.

Repeat SSS after completing one year MB MDT are necessary if it showed long slender solid bacilli with tapering ends and not responded clinically. Then MB MDT was continued for one more year till SSS showed fragmented bacilli and skin lesions resolved.

DISCUSSION

Histoid Hansen may be consider as a variant of lepromatous leprosy and as a distinct entity first described by wade as a bacillary rich leproma in 1963 [2]. Histoid term is derived from appearance of nodule that showing spindle shaped cells resembling dermatofibroma [5]. Among total leprosy patients, incidence in India vary from 2.79 % to 3.60% reported, with male predominance and in various studies by Singh et al., [6] & Schgar et al., [7], we find that 20-40 year age group was most common affected [2]. As compared to our study we find case in age group of 30-65 years. Clinically, it present as superficially placed, subcutaneous or deeply fixed cutaneous solid nodules and plaque which are multiple, painless, firm, globular, smooth, discrete, skin coloured to dark brown, with surrounding normal skin [8]. Most common site are posterior and lateral aspects of the arms followed by buttocks, thighs, dorsum of the hands, lower back and over bony prominence such as elbow and knees.

In our study nodule found in 71.5 % cases. Papules in 57.1%, plaque in 42.8% and hypopigmented patches in 42.8% as compared to study by R Subha et al., [2] in which nodule found in 91%, papules in 72%, plaque 36% and hypopigmented patches in 18%, ulceration 9% seen which are nearly same findings.

Many factors like immune response, genetic factors and treatment play an important role in deciding manifestations of histoid leprosy [3]. Mostly it occur De novo and in untreated cases [8].

In histoid leprosy cell-mediated immunity is higher than in active cases of lepromatous patients as established by increased CD36 expression by keratinocytes, CD4 lymphocytes predominant over CD8 lymphocytes and activated lymphocytes and macrophages are increased in number in the lesions [9].

In slit skin smear (SSS), Large bacillary load present in histoid lesions and acid fast bacilli are longer, arranged in groups or parallel bundles in spindle-shaped histiocytes than ordinary lepra bacilli [10].

In histoid lesion, electron microscopic features is absence of electron transparent zone or foam, which interferes with bacterial metabolism , leads to cell
death, unlike in lepromatous lesions. This absence of zone responsible for preservation of bacilli within histoid lesions.

Classical Histopathologic findings are epidermal atrophy due to dermal expansion of underlying leproma and acellular band below epidermis. Leproma consists of fusiform histiocytes arranged in whorled containing AFB. Sometimes clinically, histoid leprosy mimics lepromatous leprosy or ENL reactions, by histopathology it differentiated from LL type showing macrophage granuloma with foamy changes with innumerable bacilli and globi and ENL reactions showing acute inflammation mostly neutrophils with edema along granular AFB.

Immunohistochemistry studies showing number of dendrite epidermal cells, which are identified by antibodies against CD1, HLA-DR, CD45, CD36 as in histoid leprosy these were reduced as compared to tuberculoid (TL) and lepromatous leprosy (LL) [9].

Histoid leprosy managed initially by ROM therapy (Rifampicin 600 mg, Ofloxacin 400 mg, and Minocycline 200 mg) once, followed by MB-MDT for 2 years [5]. Repeat SSS after completing one year MB MDT are necessary if it showed long slender solid bacilli with tapering ends and not responded clinically. Then MB MDT was continued for one more year till SSS showed fragmented bacilli and skin lesions resolved.

As histoid leprosy cases lead to further spread of leprosy as it act as reservoir of disease. Therefore, early diagnosis and management of these cases needed to eradicate leprosy.

CONCLUSION
Histoid leprosy is a multibacillary infective form, with male predominance, mostly seen De novo and in patient who took inadequate or irregular treatment. Histoid leprosy cases lead to further spread of leprosy as it act as a reservoir of disease. Therefore, clinical features, characteristic histology and early detection is needed for awareness and to eradicate leprosy and at the end of the treatment it is essential to do bacteriological and morphological indices for deciding treatment guideline.

REFERENCES