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Clinico-pathological study of cases of histoid leprosy

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ABSTRACT

Background: Rare variants of leprosy pose a diagnostic challenge and histoid leprosy is one such form of disease with unique clinical and histopathological features. The objective of the study was to study clinical and histopathological features of histoid leprosy.

Methods: A prospective observational study was conducted at M. R. Medical College and Basveshwar teaching and general hospital, Kalaburagi from July 2018 to October 2019. Newly diagnosed and histologically proven cases were included.

Results: Histologically confirmed histoid leprosy accounted for 8 cases (7 males and 1 female). Mean age of the patients was 44.5 years. Patients presented with papules, plaques and nodules over an apparently normal skin. Bacterial index ranged from 4+ to 6+ according to Ridley index. Histopathology showed circumscribed lesion with free sub-epidermal grenz zone and spindle shaped cells.

Conclusions: As bacillary load is very high in these patients, they can form a potential reservoir of infection in the community especially in post leprosy elimination era.

Keywords: Histoid leprosy, Grenz zone, Histiocytes

INTRODUCTION

Histoid leprosy was first described by Wade in 1960. It is an expression of multibacillary (MB) leprosy. Histoid lesions are seen in lepromatous leprosy (LL). Histoid lesions may be occasionally seen in borderline and indeterminate leprosy. It usually manifests in patients on diaminodiphenylsulfone (DDS) for a long time, reflecting initial improvement followed by relapse. However, it is now well-known that the disease may occur *de novo* in patients who have never been exposed to any antileprosy therapy.

Histoid leprosy is characterized by typical cutaneous and/or subcutaneous nodules and plaques over an

apparently normal skin with unique histopathology and characteristic bacterial morphology.⁴

Well demarcated, cutaneous nodule is its cardinal feature.²

Histoid lesions can be classified as subcutaneous nodules, deeply fixed or superficially placed cutaneous nodules, soft nodules and plaques. In a single patient 3 to >50 such lesions may be seen. They present with firm, reddish or skin coloured dome shaped or oval papules with shiny and stretched overlying skin. They may become elevated, protruberant and even pedunculated. The skin surrounding lesion is apparently normal. So, a study was undertaken to observe the findings of clinical and histopathological features of histoid leprosy.

METHODS

This study is a prospective observational study conducted at M. R. Medical College and Basveshwar Teaching and General Hospital, Kalaburagi from July 2018 to October 2019. Newly diagnosed and histologically proven cases were included. Patients who refused to give consent for biopsy and pregnant women were excluded from the study.

A detailed history of the patient including age, sex, occupation, onset and duration of lesions, history of decreased sensations, constitutional symptoms (fever, malaise, joint pain) and prior history of drug intake (dapsone) was taken.

After that, written and informed consent from the patient was taken. Under aseptic precautions the papulo-nodular area was locally infiltrated with 2% lignocaine and punch biopsy was done using 4 mm punch and subjected to pathological examination.

The study was done after taking approval from the institutional ethics committee.

Statistical analysis

All statistical analyses were conducted by using Statistical Package for the Social Sciences (SPSS V.17.0). The categorical variables were expressed in terms of percentages and Chi-square test was used for the analysis of these variables. Continuous variables were analyzed by measures such as sample mean, standard deviation, and statistical significance was tested by independent sample T test and ANOVA test was applied for significance (p<0.05 was considered as significant).

RESULTS

Confirmed histoid leprosy accounted for 8 cases including 7 male and 1 female (Figure 1). Patients in the study were adults with mean age of 44.5 years. Most of the patients belonged to low socioeconomic status being either farmers or daily wage workers.

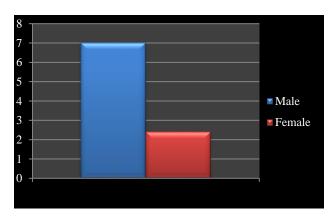


Figure 1: Gender.

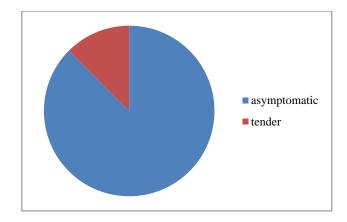


Figure 2: Papules and nodules.



Figure 3 (A-C): Multiple subcutaneous asymptomatic nodules distributed over upper limb and trunk.



Figure 4 (A and B): Ear lobe infiltration.

Seven (87.5%) patients (Figure 2) presented with multiple asymptomatic firm reddish to skin coloured dome shaped papules to plaques distributed over upper limbs, lower limbs and trunk (Figure 3). Duration of the disease ranged from 6 months to 1 year. 1 male patient presented with tender nodules which subsided on treatment. None of the patients had lesions over palms, scalp, axilla, groin and mucosa. Number of lesions ranged from 30 to 150. None of the patients had history of contact with leprosy patients. All the cases were *de novo* in presentation. Ear lobe infiltration (Figure 4) was present in 7 patients (87.5%). Multiple nerves (ulnar, superficial radial and lateral popliteal) were enlarged bilaterally and non-tender. Ulnar nerve being most commonly involved in 6 patients (75%).

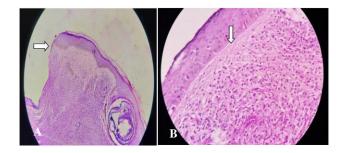


Figure 5 (A and B): Arrow mark indicates subepidermal free Grenz zone in low and high power respectively.

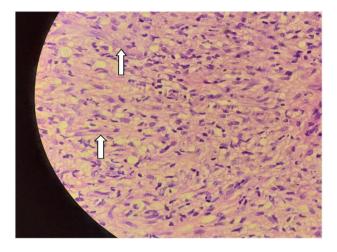


Figure 6: Arrow mark indicates spindle shaped cells called histiocytes.

Histopathology

In all the cases free subepidermal grenz zone (band of Unna) was present (Figure 5). Spindle-shaped histiocytes arranged in groups or parallel bundles were present (Figure 6). Lymphocytic infiltration was seen in all cases.

Fite ferraco stain was done which showed plenty of long slender solid staining acid-fast bacilli arranged in clusters or globi (Figure 7). Bacterial index (BI) ranged from +4 to +6 (2 patients +4, 3 patients +5 and 3 patients +6) (Figure 8). Mean BI was 5.12.

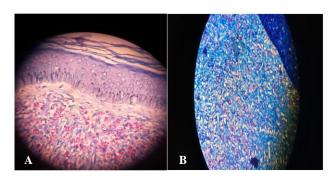


Figure 7 (A and B): Fite ferraco stain shows plenty of long slender solid staining acid-fast bacilli arranged in clusers or globi in high and low power respectively.

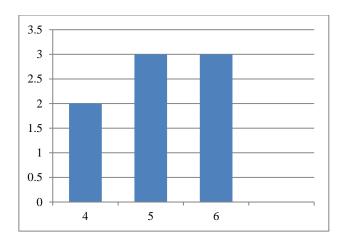


Figure 8: Bacterial index.





Figure 9 (A and B): Arrow marks indicates lesions of scrofuloderma.

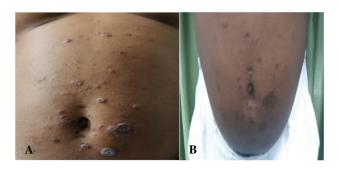


Figure 10 (A and B): Before and after 7 months of multi drug treatment.

One patient in our study also had scrofuloderma along with histoid leprosy which is also a very rare presentation. Patient was started on antitubercular drugs along with antileprosy drugs and he responded well to the treatment (Figure 9 shows images of before and after treatment with antitubercular drugs for 2 months respectively).

All the patients were started on multi drug treatment (MDT) and all patients showed clinical improvement with MDT (Figure 10) of which 3 patients lost follow up after 1 year.

DISCUSSION

Histoid leprosy is considered as a well-recognized expression of multibacillary leprosy characterized by typical clinical, histopathological, immunological and bacteriological findings.

Bacteriology

Solid staining bacilli, uniformly stained long rods with tapering ends, longer than (ordinary) leprosy bacilli.⁵ Occurs singly or sometimes in clusters or as globi.

Histopathology

Histoid lepromas, unlike ordinary lepromatous lesion grow in expansile rather than an infiltrative fashion. ^{4,5,7} It resembles tissue forming process rather than granuloma. Classical feature is presence of numerous, thin, spindle shaped histiocytes forming interlacing bands, whorls and at times tight curlicules. In most of the cases, a free subepidermal grenz zone (band of Unna) is present. ^{4,6,8} Histoid lesions can also have lymphocytes and occasionally, polymorphonuclear leukocytes, especially at the periphery of the lesion. ⁸

The total number of cases in this study was 8over a period of 1 and half year. The male: female ratio in this study was 7:1. Male preponderance was seen in this study which was similar to studies done by Kaur et al, (5.7:1), Nair et al, (16:1) and Sehgal et al (8.2:1).⁹⁻¹¹ The 40 to 50 year-old age group accounted for the maximum number of cases in this study which is similar to the studies by Kaur et al, and Nair et al, and in contrast to studies by Sehgal et al and Kalla et al where the 20 to 40 year-old age group was most commonly affected.^{9,10,12}

Seven (87.5%) patients presented with multiple asymptomatic firm reddish to skin coloured dome shaped papules to plaques distributed over upper limbs, lower limbs and trunk similar to study of Nair et al, while only 1 patient had tender nodules without any constitutional symptoms which subsided on treatment in this study. 10 A unique highlight of this study is all the cases were de novo in nature whereas study by Nair et al, accounted for 64.70% and Kaur et al, accounted for only 12.5% de novo cases.9 Erythema nodosum leprosum (ENL) is considered to be extremely uncommon in histoid leprosy. There are only a few published case reports of ENL developing in patients with histoid leprosy. 13-15 None of the patients in this study reported with type 1 or 2 reactions. Study by Nair et al, 4 out of 17, Kaur et al, 6 out of 40, Kalla et al, 2 out of 25 histoid leprosy patients had ENL.^{9,10,12}

None of the patients in this study had any deformities compared to the study of Kaur et al, where 10 patients (25%) and Nair et al, 2 patients (11.7%) had deformities. 9,10

Bacterial index of our study ranged from +4 to +6 mean being 5.12 similar to study by Nair et al, having Bacterial index of 5.17 (mean). Tuberculoid contamination means the presence of epitheloid cells in the infiltrate which was not observed histologically in this study.

MDT in histoid leprosy is similar to multibacillary leprosy i.e. rifampicin 600 mg once a month supervised, clofazimine 300 mg once a month supervised, dapsone 100 mg dailyself-administered and clofazimine 50 mg daily self-administered. All the three drugs for at least 2 years; preferably till smear negativity should be given.

CONCLUSION

As bacillary load is very high in these patients, they can form a potential reservoir of infection in the community. It should be ensured that this multibacillary form of the disease be identified and treated promptly to decrease the chances of transmission to the general population, as such cases will, in all likelihood, probably continue to occur in the post elimination era.

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Ethical approval: The study was approved by the

institutional ethics committee

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