

De Novo Histoid Leprosy with unique feature of Transmigration: A case report.

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Abstract: Introduction: To report a case of de Novo Histoid Leprosy with unique microscopic features in an individual who presented with asymptomatic raised nodules.

Method: - A 18 year old student presented with history of asymptomatic raised skin lesions over bilateral arms and chest since 6 months. There was no history of fever, pain, itching, loss of sensation or any drug intake. On clinical examination there were multiple nontender raised skin colored papules and nodules which were firm in consistency ranging in size from 0.3 cm to 1.5 cm. There was no loss of pain, touch, temperature or significant past history. Hematoxylin and eosin stained skin biopsy revealed histiocytic granulomas with heavy bacillary load on FiteFaraco stain.

Result : - FiteFaraco stain revealed plenty of lepra bacilli in the dermis as well as few lepra bacilli transmigrating from dermis to epidermis.

Conclusion:-It is important to make a diagnosis of histoid leprosy which is a rare variant of lepromatous leprosy as its management is different. The presence of lepra bacilli in the stratum spinosum and stratum corneum of epidermis indicates the possibility of desquamating epithelium being the source for the shed of lepra bacilli.

Keywords:-de novo, FiteFaraco, grenz zone, histoid leprosy, transmigration.

I. Introduction

Histoid leprosy is a clinical expression of multibacillary leprosy characterized by the occurrence of cutaneous & subcutaneous nodules and plaques with a distinctive histopathology¹. The term 'histoid leprosy' was first coined by Wade in 1960 as a histological concept of bacillary rich leproma composed of spindle shaped cells. It is so called because the microscopic appearance of nodule shows spindle shaped cells resembling those in dermatofibroma². It is an uncommon variant of lepromatous leprosy and occurs in lepromatous patients who relapse after dapsone monotherapy, in the presence of dapsone resistance or at times de novo³. The histoid lesions commonly appear as smooth, shiny, hemispherical, dome-shaped nontender soft to firm nodules which may be superficial, subcutaneous or fixed deeply under the skin and plaques or pads appearing on otherwise normal- looking skin. Histoid lesions have a preferential centofacial distribution over the forehead, tip of the nose, chin and cheeks⁴. In our case, upper chest and arms were the predominant sites involved with papules being the dominant morphological pattern.

The classic histomorphological features of histoid leprosy include epidermal atrophy, dermal expansion by the underlying histiocytic granulomas and a grenz zone immediately located below the epidermis.⁴ The lesion consist of fusiform histiocytes arranged in whorled, criss-cross or storiform pattern. These histiocytes resemble fibroblasts and is suggested to have arisen from tissue histiocytes.¹⁰ Within these histiocytes abundance of acid fast lepra bacilli are seen. The enormous bacillary population in histoid lesions is suggested to be due to focal loss of immunity.¹¹

II. Case Report

A 18 year old student presented with of small, asymptomatic, red raised lesions on outer surface of right forearm, just above the wrist, arm and chest since 6 months which gradually increased in size. There is presence of similar lesions on other arm and chest since 5 months. There was no history of fever, pain, itching, loss of sensation or any drug intake.

On clinical examination, there were multiple non tender raised skin colored papules (Fig. 1) and nodules (Fig. 2) which were firm in consistency and ranging in size from 0.3 cm to 1.5 cm in size. There was no loss of pain, touch or temperature bilaterally.

Histopathological examination of H&E stained skin biopsy (Fig. 3) revealed epidermal atrophy, subepidermal grenz zone confirmed by Masson's trichrome stain (Fig. 4) and bundles of spindle shaped histiocytes in dermis arranged in storiform pattern (Fig. 5) and sheets. Also seen were histiocytic granulomas (Fig. 6) comprising of rounded, spindle shaped as well as vacuolated histiocytes in the lower dermis. FiteFaraco stained slide shows plenty of lepra bacilli in the dermis (Fig 7) & its appendages including the sweat glands. These lepra bacilli were longer than usual lepra bacilli and were having tapering ends hence called histoid lepra

bacilli. Typical globi were not seen. They were present extracellularly as well intracellularly in clusters as well singly. There was an exclusive finding of presence of lepra bacilli in the stratum spinosum as well as corneum (Fig. 8, Fig. 9) (transmigration).

III. Discussion

Histoid leprosy is considered as a well-recognized expression of multibacillary leprosy characterized by typical clinical, histopathological, immunological and bacteriological findings.¹ Its incidence in India is estimated to be 2.79 to 3.60%. The average age at diagnosis is between 21 and 40 years.¹² Our case has presented to us at even younger age. Male to female ratio in most parts of the world is 2:1⁹.

Histopathological features of histoid leprosy can resemble fibromas or fibrosarcomas.⁸ Since histoid lesions can arise de novo, as in our case, it is important to do FiteFaraco stain in all the cases who are showing histiocytic granulomas along with storiform arrangement of histiocytes in the dermis even if the patient is not giving any history of contact or previous drug therapy. It is very important to diagnose this variant of lepromatous leprosy as the management is entirely different. Rifampicin 600 mg, Oloxacin 400 mg and Minocycline 200 mg is followed by WHO MBMDT therapy for 2 years.¹² It is a general belief that the therapy could be continued till the patient becomes Split skin smear AFB negative.

The pathogenesis of this rare and unusual variant of leprosy still remains unresolved. Although histoid leprosy is considered to be variant of lepromatous leprosy there exists an enhanced immune response against *Mycobacterium Leprae* in these patients compared with lepromatous leprosy with respect to both cell mediated immunity and humoral immunity. Despite the presence of adequate number of macrophages it has been claimed that they lack the functional property to kill bacilli that exists in high numbers in histoid lesion.⁵

In our case we found the presence of lepra bacilli not only in dermis and dermal appendages but also in all the layers of epidermis including the prickle cell layer as well as stratum corneum. The presence of lepra bacilli in desquamating epithelium and the possibility of sweat being the source for the shedding of lepra bacilli has been previously described.⁹ Ghorpade published a case report of leprosy and suggested that possibly skin could be a portal of both exit and entry for the bacillus and should be given due recognition for its contributory role in leprosy transmission.⁶ Okada et al.⁷ found the presence of lepra bacilli in the keratinocytes of epidermis by electron microscopy. Histopathological examination of the lesion is extremely helpful in typing the leprosy and hence playing vital role in the management of patient.

Presentation of young student as a de novo case of histoid leprosy with presence of intra epidermal bacilli can be an extensive source of spread. So, important steps to diagnose and treat all the leprosy cases should be taken. Histopathology & special stain plays important role in further typing of the lesion and managing the patients in a better way and as such must be done in all cases.

IV. Figures And Tables

Figure 1 Papule



Figure 1 Photograph showing multiple papules on upper chest.

Figure 2 Nodular mass

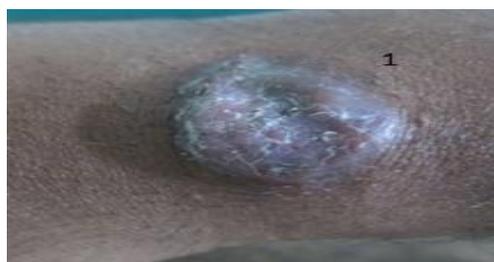


Figure 2 Nodular mass 5 cm in diameter on lateral surface of right wrist

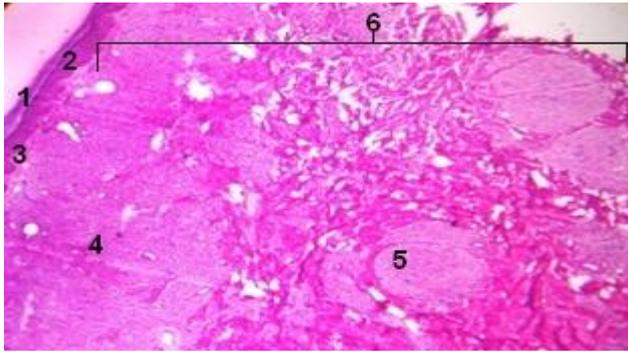


Figure 3 Skin Biopsy (H&E x4)

Figure 3

1. Hyperkeratosis
2. Atrophic epidermis
3. Grenz zone
4. Sheets of histiocytes in storiform pattern
5. Granulomas
6. Dermal expansion



Figure 4 Grenz zone Masson's trichrome Stain x40

Figure 4

1. Hyperkeratosis
2. Atrophic epidermis
3. Grenz zone
4. Granuloma encompassing dermal appendages

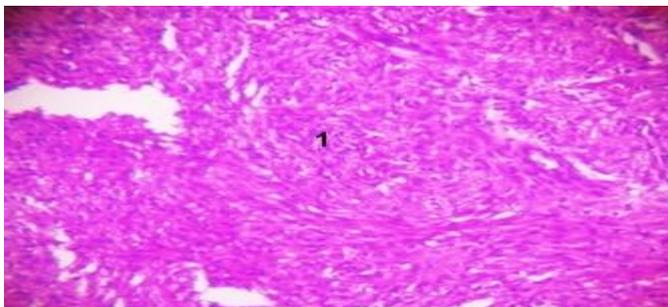


Figure 5 Storiform Pattern of histiocytes (H&E x40)

Figure 5

1. Storiform Pattern of histiocytes

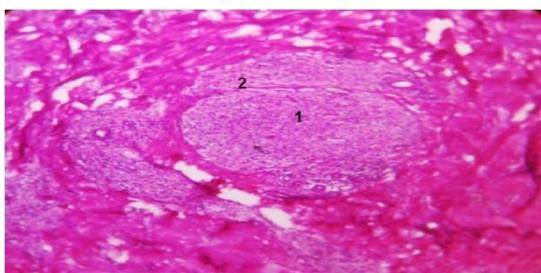


Figure 6 The dermis show histiocytic granulomas encompassing the dermal appendages. (H&E x40)

Figure 6

1. Granuloma encompassing dermal appendages in dermis
2. Pseudocapsule

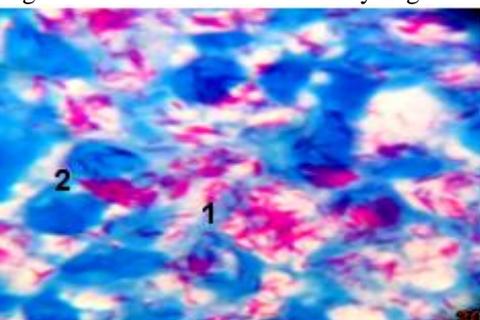


Figure 7 Plenty of Acid fast histoid lepra bacilli in singles & clusters (Fite-Faraco x100)

Figure 7

1. Histoid lepra bacilli singly scattered
2. Lepra bacilli in clusters

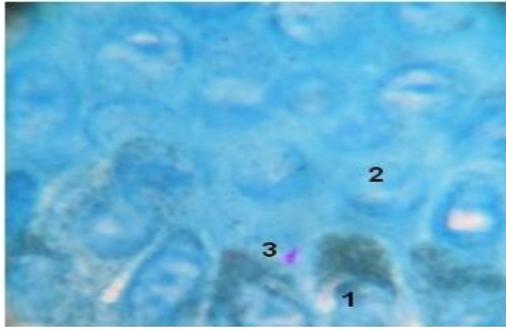


Figure 8
1. Stratum basalis
2. Stratum spinosum
3. Histoidlepra bacilli in stratum spinosum

Figure 8 The epidermis shows transmigration of bacilli in stratum spinosum (Fite Faraco stain x100)

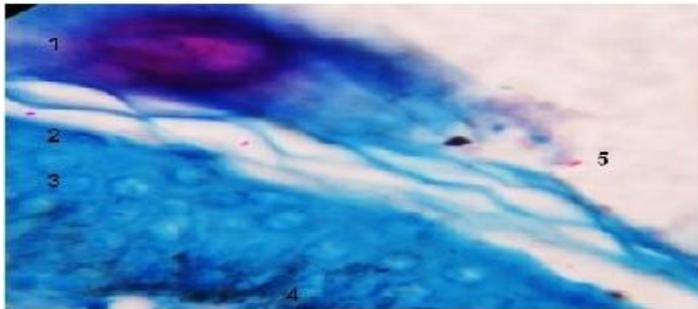


Figure 9
1. Stratum corneum
2. Stratum granulosum
3. Stratum spinosum
4. Stratum basalis
5. Histoid lepra bacilli in stratum corneum

Figure 9 The epidermis shows transmigration of bacilli in stratum corneum. (Fite Faraco stain x100)

V. Conclusion

It is very important to diagnose histoid leprosy because the immunopathology as well as the treatment differs from the lepromatous leprosy. Histoid leprosy is believed to occur due to mutant strain of leprosy bacillus called as histoid bacillus which is longer than usual lepra bacillus and has got tapering ends.¹³ So our case was unique in the sense that the patient was a de Novo case of histoid leprosy with transmigration of histoid bacilli in & out of the epidermis. De Novo presentation of histoid leprosy in a young student with spread of lepra bacilli through desquamating epithelium can be a source of spread as well as a potential reservoir of infection.

Therefore it is suggested that along with nasal, cutaneous mode is new mode of spread and should be considered and in near future managed accordingly for any such similar lesions to avoid further spread, otherwise our goal of eliminating leprosy from the world can never be fulfilled.

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