CASE REPORT

Histoid Leprosy: A Case Report

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Introduction:

Histoid Leprosy is rare but most ancient bacterial disease in the history of mankind. It is a chronic granulomatous inflammatory disease, primarily of the peripheral nervous system, skin, and reticuloendothelial system. It was first reported by Wade in 1963. Leprosy (Hansen's disease) is a chronic infectious disease which is diagnosable and curable if recognized early and treated adequately.The causativeagent of leprosy is Mycobacterium leprae which is only bacteria to infect peripheral nerves. Leprosy is characterized by a variety of abnormal immune responses. It depends on the integrity of the host's specific CMI response to the M. leprae and it may be genetically determined. Anti-leprotic Multidrugs Therapy (MDT) as recommended by WHO is now the standard and accepted method for leprosy control. Leprosy cannot be completely eradicated; it will disappear when the economic and cultural factors change, because leprosy is the thermometer of civilization. It commonly affects buttocks, back, face and extremities. It presents clinically as an erythematous or hypopigmented anesthetic patch and a thickened and/or tender cutaneous nerve trunk. Leprosy is also called Hansen disease. Leprosy is a great imitator of other skin diseases, and it can present with different morphological lesions, this is why an expert eye is needed to diagnose it. One of the important clinical presentations of leprosy is histoid leprosy, which is very difficult to diagnose due to different clinical and histopathological findings that mimic, e.g., a fibromatous disorder.

Case Report:

A 27year-old young unmarried male, electrician, from Mirpur, Dhaka, presented to us with the complaints of multiple, asymptomatic, shiny papules and nodules all over the body. The lesions were present for the last one and half year. The condition was deteriorated in last 6 months. He was suffering from multiple abscess formation in both handsand feet for last 6 months and treated locally. He had one episodes of epistaxis in the last 6 months that were treated symptomatically. The nodules were distributed predominantly over the trunk and upper extremity. Family history was non-contributory. He did not take any form of treatment for his skin lesions. Clinical examination revealed multiple discrete, skin-colored, shiny cutaneous and subcutaneous papules and nodules over the trunk and limbs. The nodules varied in size from 0.5 to 1.0 cm; there was no impairment of pain, touch, or temperature sensation. There marginal thickening of ulnar nerves. Thener muscles of both the hands were atrophied. There was no more deformity or ulcer found in the body. Systemic examination did not show any abnormality. Routine laboratory tests did not show any abnormality.

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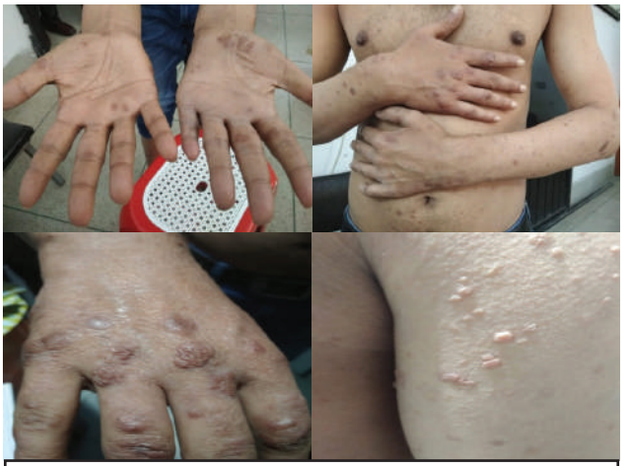
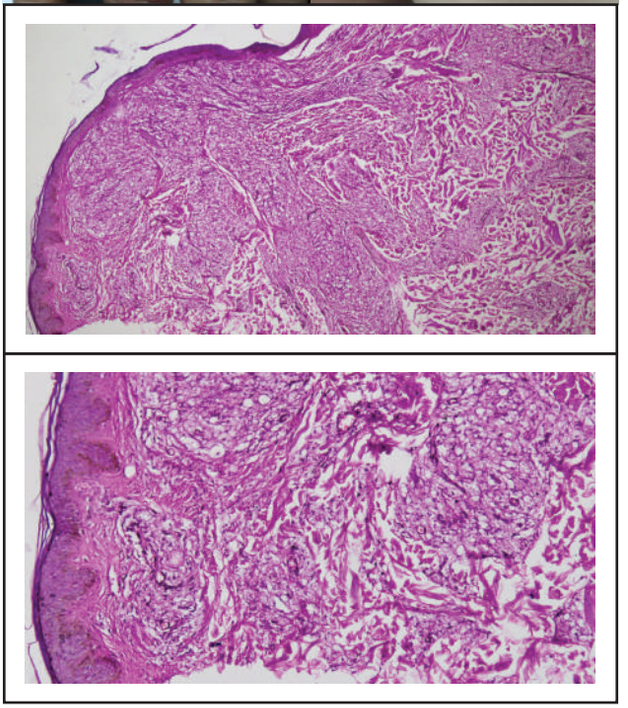
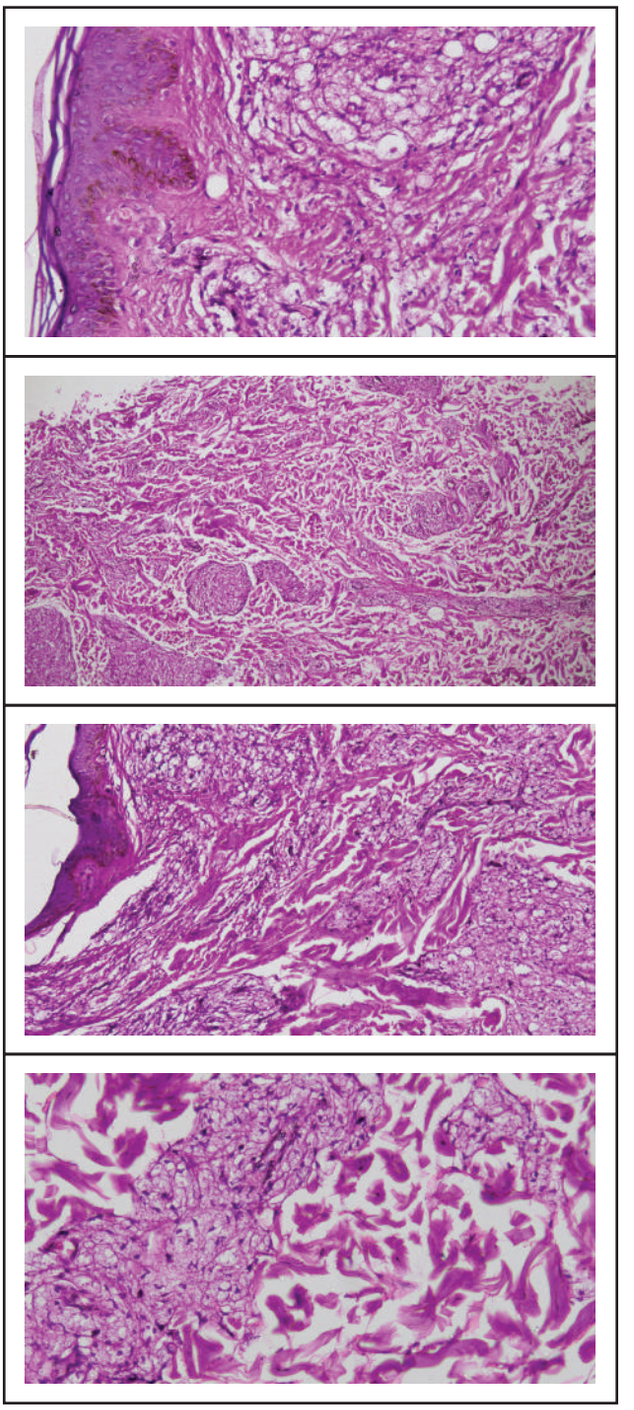
Histopathology of the nodule showed that the dermis revealed many foamy histiocytes and small amount of lymphocytes. The lymphocyte contains many sheets of spindle cells arranged in a whorled, crisscross/pstoriform pattern. Fite'sFaraco stain showed numerous acid fast bacilli. The patient was diagnosed asthe histoid variant of lepromatous leprosy. Now he isgetting treatmentwith multidrug therapy as multibacillary patient– (MB-MDT) comprising of monthly rifampicin and clofazimine; and daily dapsone and clofazimine,which will be continued for a period of 1 year.







Discussion:

Histoid leprosy is considered a variant of lepromatous leprosy1 and by others as a distinct entity2. The incidence has been reported to vary from 1% to 2% amongst total leprosy patients3. Its exact etiopathogenesis is not well understood as it may develop after an inadequate and irregular

treatment with dapsone monotherapy or MDT[4]. In Bangladesh, its incidence among leprosy patients is estimated to be 2.5 to 3.9%. There is a male preponderance, and the average age at diagnosis is between 21 and 40 years. Histoid leprosy has characteristic clinical, histo-pathologic and bacterial morphological features[5,6]. Clinically,it is characterized by cutaneous and/or subcutaneous lesions on apparently normal skin. The lesions are usually located on the posterior and lateral aspects of the arms, buttocks, thighs, dorsum of hands, on the upper and lower part of the back and over the bony prominences, especially over the elbows, knees and knuckles.

There are three histologic variants of histoid leprosy: pure fusocellular, fusocellular with epitheloid component, and fusocellular with vacuolated cells. The third pattern is most commonly observed[7].

Histoid leprosy might represent an enhanced response of the multibacillary disease in localizing the disease process. An increase in both cell-mediated and humoral immunity against Mycobacteriumleprae, as in lepromatous leprosy, has been hypothesized.

Clinical differential diagnoses include Neurofibromatosis, Eruptive xanthoma, Mucinosis. Each of them can be differentiated from histoid leprosy on the basis of the characteristic histopathology and absence of mycobacteria in slit skin smear[8].

Ears may be unaffected. Histoid lesions have also been reported to present along the course of the peripheral nerve trunks and cutaneous nerves.

However, whether histoid leprosy should be treated as other multibacillary forms or other immunotherapies should be added to the treatment regimen deserves consideration.Classical histopathological findings include epidermal atrophy as a result of dermal expansion of the underlying leproma and a grenz zone located immediately below the epidermis. The lesion consists of fusiform histiocytes arranged in a whorled, criss-cross or storiform pattern. These histiocytes resemble fibroblasts and it is suggested that fibroblast-like macrophages may have arisen from the tissue histiocytes rather than blood monocytes. Within these histiocytes, an abundance of acid-fast bacilli can be seen.

The AFB are not found in globi formation, as they do not secrete any glial substance. They are longer than the normal bacilli, uniform in length, and are arranged in parallel bundles along the long axis of histiocytes. Within the histiocytoid collections, there can also be islands of tuberculoid granulomas, which are called by Wade as, ‘contaminating tuberculoid bacilli’.

Histoid leprosy is managed by initially giving the range of motion therapy, with Rifampicin 600 mg, Ofloxacin 400 mg, and Minocycline 200 mg, which is followed by WHO MBMDT therapy[9].

Fite's stain from FNAC can be positive in case of histoid leprosy, which must stimulate interest for further studies. FNAC is a simple cost-effective method of investigation that is more useful in tuberculoid, lepromatous, and histoid leprosy patients. In our case, a correct cytological diagnosis was possible on the basis of a positive Fite-Faraco stain. This case highlights the importance of FNAC in all similar cases where spindle cell nodular lesions of the

skin are encountered. It was good to carry out a Fite-Faraco stain for ruling out or confirming the diagnosis, especially as there was little clinical suspicion in our case.

Conclusion:

Histoid leprosy is a distinct andrare form of multibacillary leprosy with characteristic clinical, bacteriological and histopathologic features. It may arise de novo or as a relapse after inadequate leprosy treatment. It is treated as multibacillary forms.

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