CASE REPORT

Hansen disease presenting as multiple shiny papules on face mimicking acne: A case report

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ABSTRACT

The present case report describes a patient with lepromatous leprosy acquired within a contagious family setting during adolescence. The young girl presented with multiple skin colored, nontender papules and few nodules, especially over face and arms, which mimicked acne, since 4 months.

INTRODUCTION

Leprosy is a contagious and chronic systemic granulomatous disease caused by *Mycobacterium leprae* (Hansen's bacillus). Characterized by transmission from person to person and has a long incubation period (between two and six years). The mycobacterium prefers peripheral tissue, as it survive better at a temperature close to 30ºC rather than 37ºC. Hence it affects the skin, peripheral nerves, the mucosa of the upper airways and other tissues such as bone and some viscera.[1] The diagnosis, based on clinical suspicion, is confirmed through bacteriological and histopathological analyses, as well as by means of the lepromin test (intradermal reaction that is usually negative in lepromatous leprosy form and positive in the tuberculoid form).[2] Depending upon the immune response against the bacilli, the disease presents with a broad clinical spectrum. At one pole are the tuberculoid (TT) patients, with effective T cell-mediated immunity resulting in very low bacterial numbers. While at the other, the lepromatous (LL) patients mount an ineffective humoral response and exhibit a high bacillary load. Other unstable forms, with characteristics between these poles, can also be observed. Family members, and especially children, who have family members with the disease are at highest risk. The disease is believed to spread through respiratory droplets in close quarters like its relative *Mycobacterium tuberculosis,* and similarly requires extended exposure to an individual in most situations, so healthcare workers are normally not infected (except with the most infective individuals such as those in the most progressed lepromatous forms, as those patients have the highest bacterial loads).[3]

CASE REPORT

A 27 years old female patient working as a maid came to our dermatology outpatient clinic complaining of asymptomatic skin raised lesions affecting face, ears, arms and hands 4 months ago,

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(Fig. 1,2) on history taking patient was diagnosed before in other dermatology clinic as inflammatory skin disease and was treated with oral antibiotics, topical steroid and emollients without improvement then in the follow up visit the treatment was changed to anti acne treatment in the form of topical and systemic medication without any improvement also. She had no past history of similar skin lesions, no history of fever, joint pains or epistaxis. There was history of Leprosy in her Uncle. Skin examination revealed multiple skin colored nontender papules and nodules of size ranging from 0.5 cm-1.5 cm, bilaterally symmetrical over both arms, hands, ears and face, peripheral nerve examination revealed thickened ulnar nerve on both sides, sensations were also affected, eye examination was normal, nail and mucous membranes were not affected and showed no significant abnormalities. Routine blood investigations did not show any significant abnormality. Skin biopsy was done from skin lesions and revealed nodular granulomatous infiltrate. This infiltrate involved the entire dermis but was clearly separated from the epidermis by a small grenz zone of collagen. The infiltrate was around the appendages and also extended into the subcutaneous fat. The involved monocyte-macrophages exhibited a rather eosinophilic cytoplasm in which bacilli could be seen, many of which were fragmented. In these chronically persistent lesions, one can observe clumps of bacteria and bacterial fragments that are called globi. Modified Ziehl-Neelsen stain (Wade-Fite) stain was done and was confirmed positive for the presence of numerous acid-fast bacilli. (Fig. 3,4) Final diagnosis of Lepromatous Leprosy was made and patient was started on WHO recommended multidrug therapy.

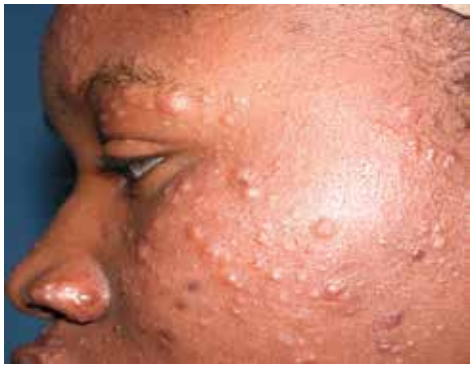

Fig. 1 Skin colored and shiny papular eruption in face.

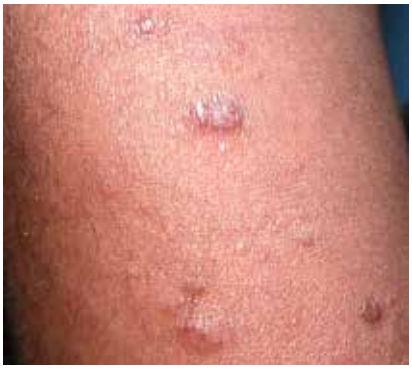

Fig. 2 Flesh colored and shiny papules and nodules in right arm.

DISCUSSION

Lepromatous leprosy is the more severe form of Hansen disease. It present with widespread skin rashes (called multibacillary leprosy), numbness, and muscle weakness. The nose, kidneys, and male reproductive organs may also be affected. It is more contagious than tuberculoid leprosy and close contact with one of family members can infect patient as in our case, which had a positive family history in her uncle. Leprosy results from the failure of Th1 cell activation which is necessary to eradicate the mycobacteria (Th1 response is required to activate macrophages that engulf and contain the disease). In lepromatous leprosy, TH2 response is turned on, and because of reciprocal inhibition (IL-4; IL-10), the cell-mediated response (TH1) is depressed. Early detection of the disease is the most importance, since severe physical and neurological damage are irreversible

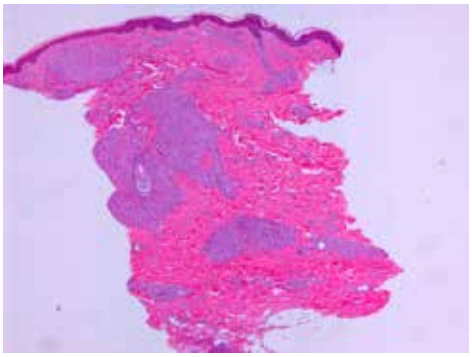

Fig. 3 Nodular granulomatous infilterate separated from epidermis by grenz zone and scattered all over the dermis.

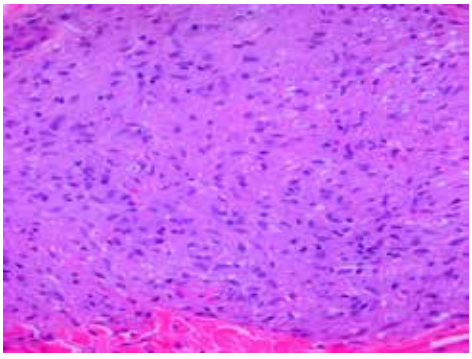

Fig. 4 Granulomatous infiltrate formed of epitheliod histiocytes, foamy hitiocytes, histiocytes with globi and few lymphocytes.

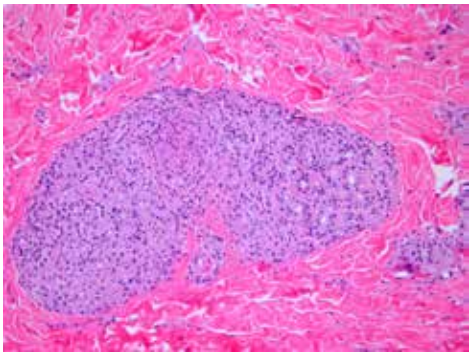

Fig. 5 Granulomatous infiltrate around eccrine glands and nerve.

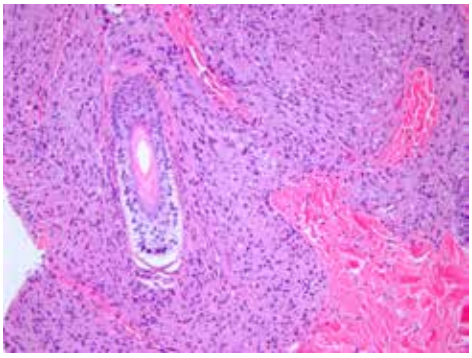

Fig. 6 Perifollicular granulomatous infiltrate.

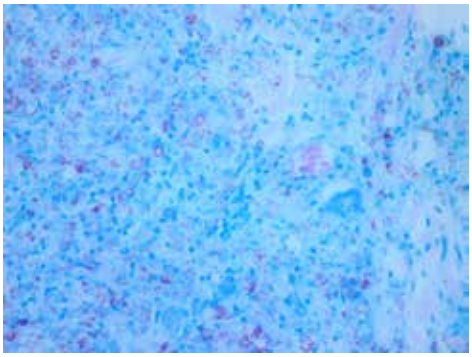

Fig. 7 Fite Stain revealed numerous lepra bacilli within histiocytes.

even if cured (e.g. blindness, loss of digits/limbs/sensation). This early presentation is the same for both tuberculous and lepromatous forms of leprosy as they are a spectrum of the same disease (lepromatous being the more contagious and severe form in patients with impaired Th1 response). In our case there was complete loss of sensation to warm and cold, also sensation to fine touch was affected which is not common in this type of leprosy. Lepromatous leprosy can present as plaques, macules, papules, and nodules affecting the face, ears, trunk, and extremities.[4] The skin nodules can clinically mimic infective or soft-tissue nodules as in our case firstly it was treated as skin infection then was treated like acne. Disease progression is extremely slow, and signs of infection may not appear for years.[5] This debilitating form of leprosy begins to spread causing the eyebrows to disappear and spongy tumor like swellings appear on the face and body. Additional signs and late complications include madarosis, saddle nose, infiltration of both earlobes, and acquired ichthyosis on the lower extremities. Anesthesia in a stocking or glove distribution may develop, often together with enlarged peripheral nerves and neuropathic changes. Ocular manifestations such as lagophthalmos (inability to completely close the eyes) and corneal and conjunctival anesthesia

due to involvement of branches of the facial and trigeminal nerves, respectively, are sometimes present in severe cases.[6,7] Histoid leprosy is a clinically distinct form of multibacillary disease that some authors classify as a variant of lepromatous leprosy, it is characterized by the development of dermatofibroma-like papules and nodules.[8] One of the major problems faced by clinicians and patients, especially during treatment, is the development of reactions. These are characterized by acute inflammation that appears suddenly. Type 1 reactions can affect patients with any form of leprosy (except for the early indeterminate form), but there is a predilection for those in borderline categories. Type 2 reactions most often occur in patients with lepromatous or borderline lepromatous leprosy.[9] Type 1 (reversal) reactions are due to a change in the immunologic state of the patient and are often associated with neuritis. Type 2 reactions are due to the formation of immune complexes in association with an excessive humoral reaction. They usually occur when patients with lepromatous forms of leprosy undergo treatment. A type 2 reaction represents a cutaneous and systemic small vessel vasculitis, and the most common clinical manifestation is erythema nodosum leprosum.[10,11,12] Patients with the diffuse form of lepromatous leprosy, who are usually from Central or South America, may develop the Lucio phenomenon, a reactional state characterized by thrombotic phenomena in addition to necrotizing cutaneous small vessel vasculitis. On a worldwide basis, leprosy is a common cause of cutaneous vasculitis, especially in low-income countries.[13,14] The disease attacks the internal organs, bones, joints and marrow of the body resulting in physical degeneration. The result is deformity with loss of feeling in the fingers and toes which eventually fall off.

When leprosy is suspected, the diagnosis of several forms (e.g. LL, BL) can be confirmed by finding bacilli in cutaneous smear, lymph nodes, or nasal secretions. Samples for bacilloscopy may be obtained from the earlobes, forehead, chin, extensor forearms, and dorsal fingers, as well as the buttocks and trunk. To avoid bleeding, a fold of skin is firmly squeezed between the finger and thumb of the examiner or with forceps, and a small incision is made with a scalpel blade. The liquid obtained is smeared onto a slide and allowed to dry. The smear is usually stained by the Fite (or Ziehl-Neelsen) method and a search is made for red rods (against a blue background) at 100× with oil immersion.[15] Organisms are found in 100% of patients with lepromatous leprosy. By PCR, a number of genes encoding antigenic proteins (e.g. 36 kD aproline-rich antigen, Ag85B) can be amplified, as can *M. leprae*-specific repetitive repeat sequences (RLEP region). This molecular technique is of particular help in paucibacillary leprosy and can be performed on slit-skin smears and fresh, frozen,[16] or paraffin-embedded skin biopsy specimens. In a study utilizing RLEP real-time PCR, *M. leprae* DNA was detected in 38 (75%) of 51 paraffin-embedded skin biopsy specimens from patients with paucibacillaryleprosy.[17] In addition, immunohistochemical staining of biopsy specimens for the PGL-1 antigen may prove helpful in paucibacillary disease.

Pathological evaluation is also helpful in skin diseases that may be confused with leprosy and different cutaneous presentations, in the lepromatous pattern, an infiltrate is seen in the dermis, subcutis, lymph nodes, abdominal organs (e.g. kidney, liver), testicles, and bone marrow. The infiltrate contains Virchow cells, which are macrophages with

numerous bacilli as well as lipid droplets in their cytoplasm. In H&E-stained sections, these cells have a foamy appearance. The bacilli in leprosy can be detected by a Gram, Ziehl-Neelsen, or Fite (the most commonly used) stain, all of which stain the bacilli a bright red color. Methenamine silver stains are also useful for detecting fragmented acid-fast bacilli. It is recommended that at least six sections be examined before declaring them negative.[19,20] A band of normal-appearing dermis (Unna band or Grenz zone) separates the epidermis from the infiltrate, which is composed of plasma cells and lymphocytes in addition to Virchow cells. Both isolated bacilli and globi (clumps of bacilli) are also seen in the dermis.

Nowadays, three classic skin tests for leprosy - histamine, pilocarpine, and lepromin (Mitsuda) - are performed infrequently.[18] The lepromin test provides prognostic, but not diagnostic, information. It is positive in TT and BT leprosy.[15]

Contrary to popular belief, both forms of leprosy are curable, with the lepromatous form classically treated with Dapsone, Rifampin and Clofazimine. But, if left untreated the person may die up to 20 or 30 years from its inception.Leprosy can be cured. In the last two decades, 16 million people with leprosy have been cured. The World Health Organization provides free treatment for all people with leprosy.

Treatment depends on the type of leprosy that you have. Antibiotics are used to treat the infection. Long-term treatment with two or more antibiotics is recommended, usually from six months to 2 years.[21,22] Antibiotics cannot treat the nerve damage. Multidrug therapy is extremely effective. Importantly, after the first dose, the patient is no longer infectious to others. All patients who complete the prescribed regimen are considered cured, as there are virtually no relapses. Bacilli may be found but they are non-viable. Drugs that may play a significant role in the treatment of leprosy in the future include other quinolones (e.g. moxifloxacin), clarithromycin, and ansamycins.[23,24] Rifapentine (a rifampin derivative) has higher peak serum concentrations, a longer serum half-life, and more bactericidal activity against *M. leprae* than does rifampin. In addition, moxifloxacin appears to be more bactericidal than ofloxacin. Thus, the combination of rifapentine, moxifloxacin, and minocycline (PMM) may be superior to rifampin, ofloxacin, and minocycline (ROM).[24] Anti-inflammatory drugs are used to control nerve pain and damage related to leprosy. This may include steroids, such as prednisone.Patients with leprosy may also be given thalidomide, a potent medication that suppresses the body's immune system. It helps treat leprosy skin nodules. Thalidomide is known to cause severe, life-threatening birth defects and should never be taken by women who are pregnant or women who may become pregnant.[23,24]

Although leprosy is now considered a curable disease with a good prognosis and excellent survival rate, it can still be incapacitating and stigmatizing. It is very important to make the diagnosis as soon as possible and to examine contacts, since early treatment prevents development of disabilities. Recognition of leprosy in its initial phases by healthcare professionals as well as the general population in endemic countries is essential in order to reduce the impact of this disease.

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