Leprosy Specific Orofacial Aspects

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ABSTRACT

Leprosy is a chronic infection caused by Mycobacterium leprae, GHA. Hansen first identified the organism in 1873, so called Hansen disease. Mycobacterium leprae is a bacillus that presents a peculiar tropism for the skin and peripheral nerves. The upper airway has a great importance as a route of M. Leprae infection. The clinical spectrum of leprosy ranges from the tuberculoid form (TT) to the disseminative and progressive lepromatous form (LL). Cell-mediated immunity is considered to be the crucial defence against the disease and the magnitude of this immunity defines the extent of the disease. Facial lesions in leprosy can occur in all form of the disease and also in lepra reaction, oral lesions are rare but, when present, occur in the lepromatous form.

Keywords: Leprosy, Oral lesion, Mycobacterium leprae, Treatment.

INTRODUCTION

Leprosy is a chronic infectious disease caused by Mycobacterium leprae, an acid-fast bacillus that presents a peculiar tropism for peripheral nerves and the skin. The prevalence of leprosy in the world has declined since the introduction of the multi-drug therapy (MDT) recommended by the World Health Organization (WHO). India accounts for 80% of the detection of leprosy cases in the world. The annual case detection rate in India is highest among the world (53 per 100,000). The global prevalence rate is around 1.25 per 10,000 persons. The predominant route of leprosy transmission is from the contaminated patient’s upper airways to a new host, thus emphasizing the importance of mucosal lesion control. The nasal mucosa can be affected in the early stages of the disease and oral involvement appears in the advanced stages, a fact suggesting the hematogenic or lymphatic dissemination of M. leprae. Leprosy-specific oral lesions are generally asymptomatic ulcers or nodules sometimes rich in M. leprae resembling nonspecific oral lesions, but they can maintain the focus of infection in endemic areas.

CLASSIFICATION

There are two types of classification widely referred; Indian classification and Ridley Jopling classification. Such a classification is necessary since the disease exhibits wide range of clinical manifestations, basically both classifications are same but in Indian classification there are indeterminate and neuritic types which do not appear in Ridley-Jopling’s.

- Indian classification includes:
  - Tuberculoid
  - Borderline

- Ridley-Jopling classification includes:
  - Tuberculoid (TT)
  - Borderline tuberculoid (BT)
  - Mid-borderline (BB)
  - Borderline-Lepromatous (BL)
  - Lepromatous (LL).

PATHOPHYSIOLOGY

M. leprae favors temperatures a little below the body temperature for its multiplication. Based on this fact, a pathophysiologic mechanism is postulated for oral involvement: A nasal lesion with obstruction of the air flow leads to oral breathing (mouth breathing), which is very common in lepromatous leprosy. This causes a decrease in the intraoral temperature, mainly in sites near the air intake, the anterior areas, facilitating the harbouring of the bacillus.

FOR SCHWANN CELLS

The unique predilection of M. leprae for Schwann cells is probably determined by the mycobacterium’s binding to the G domain of the 2 chain of laminin 2, which is a component of the basal lamina of Schwann cells. This form of laminin is restricted to peripheral nerves, which explains the specific tropism of M. leprae.

OROFACIAL MANIFESTATIONS OF LEPROSY

Facial lesions of leprosy do not differ from lesions in other parts of body, but there are some features and deformities.
LEPRA REACTION

Lepra reaction, the hypersensitivity reactions to the presence of the pathogen in the tissues, may occur any time during the course of the disease. There are two types of reactions: Type I and type II.9,10,4,15

Type I

It is a delayed type of hypersensitivity reaction, i.e. type-IV. It is usually seen in borderline leprosy. Antigens from breaking down lepra react with T lymphocytes leading to rapid change in cell mediated immunity. Clinically some or all skin lesions become erythematous, edematous, warm to touch and tender. Necrosis with supervening ulcerations may also occur. Rapid swelling of one or more nerves due to acute neuritis may lead to motor deficit. Early recognition is necessary to prevent the consequences of acute neuritis. It is managed with systemic steroids or other anti-inflammatory drugs in addition to anti-leprosy drugs.9,10,4,15

Type II

It is an immune complex disease (type III hypersensitivity reaction). It is seen usually in lepromatous or borderline lepromatous. It is characterized by appearance of crops of brightly erythematous, tender nodules or plaque that may ulcerate. Fever and arthralgia is common. Such episodes may occur even after the completion of treatment. It is managed with thalidomide, systemic steroids and antimalarials.9,10,4,15

TUBERCULOID LEPROSY

It is a benign form involving the skin, nerves and regional lymph nodes. Lesions are divided into early, intermediate and developed lesions. Early lesions includes hypopigmented macules, sharply demarcated and hypoesthetic. Intermediate lesions become larger with elevated and circinate margin, peripheral spread, and irritation of nerve ending, persistent or recurrent paresthesia and numbness. Developed lesions are densely anesthetic and loose normal skin organ (sweat gland, hair follicle), neuritic pain. Facial lesions includes regional nerve thickening, usually greater auricular nerve or supra orbital nerve. Loss of eyebrows and eyelashes were noticed.10,15

BODERLINE TYPE

It is the most common form of leprosy. It resembles tuberculoid leprosy disease except that the skin lesions are usually larger, greater in number, the edges of the lesions are less defined and sometime there are satellite lesions near the edges of the lesion. It resembles lepromatous disease except that some of the skin lesions are anesthetic, number of lesions are lesser and more asymmetrical. Oral lesion resembling those of lepromatous leprosy can rarely occur.9,10

LEPROMATOUS TYPE

It is more commonly seen in children and females. This is a malignant form of the disease, involvement of body skin, peripheral nerves, mucous membrane, lymph nodes, eyes, skeleton, testis and other internal organs. There are no early symptoms of nerve damage, patient does not notice these lesions to seek medical attention and continue to infect others. Common lesions seen on the face includes macule, papules, nodules, and diffuse thickening of the skin. Thickening of the skin on the face can cause deepening of the natural line on the forehead and rest of the face (leonine face). Ear lobes are also thickened, eyebrows may be lost and nose may collapse (due to involvement of anterior nasal spine). Skin lesions are numerous and have bilateral and symmetrical distribution. Nerve involvement occurs in lateral stage of disease. Incidence of facial palsy has been varying from 3 to 24.5%. Facial nerve paralysis is thought to be due to bacterial invasion in the extracranial part of facial nerve. Incidence of bilateral facial palsy is uncommon and occurs in 0.3 to 2% cases. Oral lesions are mostly seen in this type. The main dangers for the eye in leprosy are lagophthalmosos, corneal hypoesthesia and iridocyclitis.10,16-19

INDETERMINATE TYPE

It clinically presents as a hypopigmented ill-defined macule usually seen in covered areas of body but also present on the face. It is very rare as compared to other types and difficult to diagnose clinically so should be diagnosed histologically.13,20

ORAL MANIFESTATIONS

The oral lesions in leprosy develop insidiously, are generally asymptomatic and are secondary to nasal changes.21-23 The most frequently affected site is the hard palate.13,22,24,25 The greater prevalence in men could be explained by the fact that women seek doctor’s advice earlier, perhaps for esthetical reasons.23,26 In the advanced stages, there may be deformities and functional alterations, such as fibrosis and retraction of the soft palate or perforation of the hard palate, with serious disturbances in phonation, and nasal regurgitation of food.21,23,27

Palate

Although most authors have found more serious changes in the mid-forward portions, some have found the soft palate to be more commonly affected area.23,28,29 The most varied types of lesions are observed: Infiltration, ulceration, perforation, and reddish or yellow-reddish nodules, sessile or pedunculated, varying from 2 to 10 mm, some confluent, and prone to ulceration.23,30,31
**Tongue**

It is affected in 17 to 25% of the cases, mainly the dorsal surface, especially the anterior two-thirds. Changes from superficial erosions with loss of the papillae and longitudinal fissures have been described to nodular infiltration, that could lead to a 'paving stone appearance'. Scarring can also occur. Unlike other subcutaneous muscles, in which a great number of bacilli are observed, the muscles of the tongue do not exhibit significant numbers. Mukjerhee and Bucci et al. suggest that the lesions of the base of the tongue could originate from highly infectious nasal secretions, which pass from the nasal to the oral cavity.

**Uvula**

In extreme cases there is intense fibrosis with partial loss or even complete destruction of the uvula.

**Lips**

There may be macrocheilia (caused by infiltration) or microstomia (caused by ulceration and subsequent repair with fibrosis of perioral or lip lepromas).

**Gingiva**

They are usually affected in the area behind the upper central incisors, often by contiguity, of lesions of the hard palate. Chronic gingivitis, periodontitis may occur.

**Dental Changes**

Dental changes of leprosy are described as odontodysplasia leprosa. There is early and severe granulomatous involvement of premaxilla in childhood. Other features include circumferential hypoplasia, shortening of roots, usually involving maxillary anterior teeth. Longstanding lepromatous lesions may show granulomatous invasion of pulp and pinkish discoloration of crowns.

**LABORATORY FEATURES**

a. **Mistuda reaction:** It is complementary test for the diagnosis of leprosy. It is a delayed type of hypersensitivity reaction in which erythematous papular nodules develop at the site of intradermal injection of mistuda agent.

b. **Biopsy:** Histopathologic examination of the skin shows an atrophic epidermis with loss of the rete ridges in all patients. Oral lesions showed epithelial hyperplasia with areas of ulceration and a dense lymphohistiocytic inflammatory infiltrate with the presence of histiocytes showing foamy cytoplasm vacuoles with large amounts of acid-fast bacilli in all patients.

c. **Determination of antibodies to phenolic glycolipid I (anti-PGLI):** The PGL-1 fraction is part of the cell envelope of *M. leprae* and induces the production of the humoral specific response against PGL-1 detected in patient serum. When the antibody is present at high levels, the infection supposed to be active, especially during the reactional episodes, which constitute a very common complication in the evolution of leprosy.

d. **Skin smears:** Can be obtained by scraping the dermis with scalpel or razor blade and are examined microscopically both the density of organism (bacteriological index) and the percentage of organisms that stain evenly (morphological index).

e. **Bacilloscopy:** To identify *Mycobacterium leprae* in ocular conjunctivae.

f. **Polymerase chain reaction:** But not used routinely.

**DIAGNOSIS**

Traditionally presence of two out of three cardinal signs has been considered as diagnostic of leprosy. These include:

1. Diminution or loss of sensation in a typical skin lesion or in an area supplied by peripheral nerve.
2. Enlargement and/or tenderness of a peripheral nerve.
3. Finding of acid fast bacilli in smears.

**DIFFERENTIAL DIAGNOSIS**

Differential diagnosis of leprosy is divided into three types:

a. Differential diagnosis of flat lesion
b. Differential diagnosis of raised lesions
c. Differential diagnosis of nodular lesions.

**Differential Diagnosis of Flat Lesion**

1. **Birth mark or naevoid:** Present since birth, edges sharply defined saw tooth appearance, normal sweating, sensations intact
2. **Vitiligo:** Depigmented lesion, sweating normal, sensations intact, white hairs on lesion.

**Differential Diagnosis of Raised Lesions**

1. **Granuloma annulare:** Annular (ring shaped orientation) in extremities, no sensory impairment or nerve changes, history/family h/o diabetes
2. **Lymphoma:** Sensations normal, long standing lesion, associated lymphadenopathy hepato- splenomegaly, nerves normal

**Differential Diagnosis of Nodular Lesions**

1. **Neurofibromatosis:** Coffee-ground coloured macules, soft multiple nodules, painful on pressure (sometimes), skin smears negative for *M. leprae*, iris lesions, familial, present since birth/early childhood.
2. **Dermal leishmaniasis:** Nodules/infiltration on face and ear like LL, history of kala-azar, skin smears negative for *M. leprae*, no sensory changes, no nerve abnormalities.

**TREATMENT**

- Multidrug therapy (MDT) is the cornerstone of the leprosy elimination strategy as it cures patients, reduces the reservoir
of infection and thereby interrupts its transmission. MDT also prevents disabilities through early cure.

- For purposes of treatment, leprosy is divided into two types as follows:
  - *Pauci-bacillary (PB) leprosy*: 1 to 5 skin lesions. Regimen of two drugs—Rifampicin and Dapsone for 6 months.
  - *Multi-bacillary (MB) leprosy*: >5 skin lesions. Regimen of three drugs—Rifampicin, Clofazimine, and Dapsone for 12 months.
  - Paucibacillary include all intermediate, tuberculoid and borderline tuberculoid cases.10
  - Multibacillary include mid borderline, borderline lepromatous and lepromatous cases.10
  - Regimen for paucibacillary leprosy 600 mg rifampicin monthly, supervised, and 100 mg dapsone daily, unsupervised, for 6 months. It can be treated with a single therapeutic dose consisting of 600 mg rifampicin, 400 mg ofloxacin, and 100 mg minocycline.10,41-45
  - Regimen for multibacillary consist of 600 mg rifampicin and 300 mg clofazimine monthly, supervised, and 100 mg dapsone and 50 mg clofazimine daily, for 12 months.10,41-45
  - Reduced doses of the above regimen are appropriately determined for children.

As according to the literature oral lesion are always present with skin lesions so, as the treatment of leprosy should be done with MDT and with MDT oral lesions are also subsided.

**TREATMENT FOR ORAL LESIONS**

In patients undergoing MDT, most of the oral lesions will subside by itself. If ulcer persists then symptomatic treatment is the primary approach. Adequate oral hygiene helps in relieving symptoms. Typical antihistamines, antacids, corticosteroids or applications meant to soothe the painful ulcers will be helpful, oral analgesics such as paracetamol or ibuprofen and local anesthetic lozenges, paints or mouth rinses such as benzocaine, avoiding spicy or hot foods will reduce pain.

Debacterol, is an acidic agent which chemically cauterizes the ulcer surface, sterilizing it, and covering the painful nerve endings. A healthy diet with vitamin supplementation is recommended. Excellent oral hygiene, including use of bacterial rinses (Rx chlorhexidine or OTC listerine), has been shown to reduce frequency of attacks. Reducing stress is important as well. Topical steroid treatments (Kenalog, Lidex gel, Decadron rinses) are quite useful, reducing the pain and duration of the lesions. Usually a weaker steroid is used first and stronger varieties are attempted until results are found. In severe cases a short course of systemic steroid (prednisone, and has been shown to have some beneficial effects as well.

For Fissured Tongue

No definitive therapy or medication is required for fissured tongue.9 If symptomatic, patients with fissured tongue are encouraged to brush the dorsum of the tongue to eliminate debris that may serve as an irritant.

**For Gingivitis**

The first step is to thoroughly clean your teeth, removing all traces of plaque and tartar—a procedure known as scaling. The cleaning may be uncomfortable, especially if your gums are already sensitive or you have extensive plaque and tartar buildup. Meticulous oral hygiene is necessary after professional tooth cleaning. Professional tooth cleaning in addition to brushing and flossing may be recommended twice per year or more frequently for severe cases. Antibacterial mouth rinses or other aids may be recommended in addition to frequent, careful, tooth brushing and flossing.

**For Periodontitis**

In the earlier stages of the disease, most of the treatment involves root planing and curettage (cleaning) under the gum margins. It involves the removal of plaque and inflamed soft tissue in the pockets around the tooth with an instrument called a curette. Its purpose is to remove the bacterial colonies and the mechanical and chemical irritants that cause inflammation in hopes that the disease can be eradicated. The goal is that the gum will reattach itself to the tooth or will shrink enough to eliminate the pocket.

**For Palatal Perforation**

For palatal perforation obturator can be given and surgical treatment with repositioning of flap may be done if necessary.

**REFERENCES**