The Leonine Face of Leprosy: An International Exploration

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International Exploration
India, Ethiopia, Brazil
History and Prevalence

- **600 and 200 BC**: First case of Leprosy reported in India/China.
- **1873**: G.H. Armoeur Hansen discovered Leprosy.
- Oldest pathogen after *Pseudomonas aeruginosa* and *Bacillus Anthracis*.
- **1980**: 11-15 million worldwide
- **2010**: 212,000 cases
Mycobacterium Leprae

- Acid-fast, obligate intracellular curved rod.
- Slow Reproducer: 14 days to double!
- Incubation Period: 4-10 years
- Likes cold temperatures
- Found in armadillos, mice foot pads, and monkeys.
Etiology, Epidemiology, and Transmission

• **Three requirements:** contagious patient, susceptible patient, and close intimate contact.

• 25% transmission in domestic contacts.

• **Respiratory:** Most common mode of transmission (followed by dermatologic then GI).

• HLA DR 2 and DR 3: Tuberculoid forms

• HLA DQ1: Lepromatous form
The clinical-immunologic spectrum of leprosy.

**FIG. 75.2** The clinical-immunologic spectrum of leprosy.
# Ridley Joplin Spectrum of Leprosy

## Classification of Leprosy

<table>
<thead>
<tr>
<th>Clinical findings</th>
<th>LL</th>
<th>BL</th>
<th>BB</th>
<th>BT</th>
<th>TT</th>
<th>I</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type of lesions</td>
<td>Macules, papules, nodules, diffuse infiltration</td>
<td>Macules, papules, plaques, infiltration</td>
<td>Plaques and dome-shaped, punched-out lesions</td>
<td>Infiltrated plaques</td>
<td>Infiltrated plaques, often hypopigmented</td>
<td>Macules, often hypopigmented</td>
</tr>
<tr>
<td>Number of lesions</td>
<td>Numerous</td>
<td>Many</td>
<td>Many</td>
<td>Single, usually with satellite lesions, to more than 5</td>
<td>One or few (up to 5)</td>
<td>One or few</td>
</tr>
<tr>
<td>Distribution</td>
<td>Symmetric</td>
<td>Tendency to symmetry</td>
<td>Evident asymmetry</td>
<td>Asymmetric</td>
<td>Localized, asymmetric</td>
<td>Variable</td>
</tr>
<tr>
<td>Definition</td>
<td>Vague, difficult to distinguish normal versus affected skin</td>
<td>Less well-defined borders</td>
<td>Less well-defined borders</td>
<td>Well-defined, sharp borders</td>
<td>Well-defined, sharp borders</td>
<td>Not always defined</td>
</tr>
<tr>
<td>Sensation</td>
<td>Not affected</td>
<td>Diminished</td>
<td>Diminished</td>
<td>Absent</td>
<td>Absent</td>
<td>Impaired</td>
</tr>
<tr>
<td>Bacilli in skin lesions</td>
<td>Many (globi)</td>
<td>Many</td>
<td>Many</td>
<td>Few (1+), if any, detected</td>
<td>None detected</td>
<td>Usually none detected</td>
</tr>
</tbody>
</table>
Indeterminate Leprosy

- Early stage of leprosy that may progress into other types.
- Seen in children.
- Single, ill-defined hypopigmented patch with slight anesthesia.
Tuberculoid Leprosy/TT

Clinical:
1/2 well-defined plaques less than 10 cm with decreased sensation/sweating.

Bacteriological: Negative

Histopathological:
  Granuloma in the dermis

Immunological: Lepromin Test: ++
Tuberculoid Leprosy
Tuberculoid Leprosy Histopathology
Borderline forms of Leprosy - UNSTABLE

• BT- Borderline Tuberculoid
• BB- Borderline Borderline
• BL- Borderline Lepromatous
BR Hansen’s/ Borderline Tuberculoid Leprosy

• **Clinical:** 1-2 lesions not as well defined lesions that are greater than 10-20 cm with decreased sensation/sweating.

• **Bacteriological:** Negative

• **Histopathological:** Tuberculoid granuloma with epitheliod cells and lymphocytes.

• **Immunological:** Lepromin test weakly positive.
Borderline Borderline/Mid-Borderline

- **Clinical**: Multiple asymmetric plaques with possible decreased sensation/sweating.
- **Bacteriological**: 1+ to 3+
- **Histopathological**: Loosely arranged granuloma of epithelioid like histiocytes and lymphocytes. May start to have Grenz Zone.
- **Immunological**: Lepromin Negative
Borderline Lepromatous/ BL Hansen’s

- **Clinical:** Multiple small plaques with normal sweat/sensation tending towards symmetry.
- **Bacteriological:** 2+ to 4+.
- **Histopathological:** Histiocytes and lymphocytes are clustered within poorly defined and patchy granuloma Grenz zone, deep dermal granulomas.
- **Immunological:** Lepromin Negative.
Lepromatous Leprosy

- **Clinical**: Leonine facies, madarosis, saddle nose, etc.
- **Bacteriologically**: 3+ to 6+.
- **Histopathological**: Grenz zone with diffuse granuloma of monomorphous histiocytes.
- **Immunological**: Lepromin Negative.
Lepromatous Leprosy
Histopathology
Our LL Patient

- Icthyotic Patients on Shins
- Bullae on Right Palm
- Leonine Faces
- Madriosis
- Dactylitis
- Pedal Edema
- Nasal Stuffiness/Epistaxis
- Infiltration of earlobe
- Glove and Stocking Sensory loss
Diagnosis of Leprosy

- Both Neurologic and Dermatologic
Dermatologic Diagnosis of Leprosy

- **Skin Biopsy**: Looking for acid fast bacilli
- **Ear slit skin**: distinguishes multibacillary from paucibacillary
- **Histamine test**: to test nerve damage
- **Antibody testing including**: FLA-ABS and PGL-L
- **Lepromin skin test**: (not useful) tests for cell-mediated unity.
Earslit Test
Neurologic Testing

Muscle Testing:
• Thenar Atrophy
• Hypothenar Atrophy
• Guttering of the dorsal interossei
• Pen Test
• Card Test
• Book Test
• EMG for definitive diagnosis

Peripheral Nerve Testing: Palpation
Leprosy Reactions

- **SUDDEN** exacerbation in clinical activity that requires **EARLY** intervention.
- **Causes:** Antibiotics for leprosy, infections, vaccination, pregnancy, medications.
- **Warning signs:** pain of existing lesions and new lesions.

**Two Types:**
- **Type I:** cell mediated immunity within existing skin lesions for borderline types
- **Type 2:** is a mediated by immune complexes seen in LL and BL types.
Type 1 Reaction

- Sudden change in EXISTING LESIONS in borderline patients
- MCC seen in BL.
- Can get better (reversal) or worse (downgrade)
- Not associated with constitutional symptoms.
- **Cause:** Cell mediated attack of M. Leprae antigen.
- Can cause local neuritis.
Type 2 Reaction/Erythema Nodosum Leprosy

- Erythema Nodosum-like lesions that occur in either BL or LL.
- Due to circulating immune complexes.
- Associated with constitutional symptoms.
- Conjunctivitis, neuritis, synovitis, nephritis, hepatosplenomegaly, orchitis, lymphadenopathy.
- Histologically- leukocytoclastic vasculitis!
## Treatment of Leprosy

<table>
<thead>
<tr>
<th></th>
<th>Rifampin</th>
<th>Clofazimine</th>
<th>Dapsone</th>
<th>Ofloxacin</th>
<th>Minocycline</th>
<th>Therapy duration</th>
</tr>
</thead>
<tbody>
<tr>
<td><em><em>MB (&gt;5 lesions</em>)</em>*</td>
<td>600 mg once monthly</td>
<td>300 mg once monthly and 50 mg daily</td>
<td>100 mg daily</td>
<td>–</td>
<td>–</td>
<td>12 blister packs over 12 to 18 months</td>
</tr>
<tr>
<td><em><em>PB (2–5 lesions</em>)</em>*</td>
<td>600 mg once monthly</td>
<td>–</td>
<td>100 mg daily</td>
<td>–</td>
<td>–</td>
<td>6 blister packs over 6 to 9 months</td>
</tr>
<tr>
<td><em><em>PB (single lesion</em>)</em>*</td>
<td>600 mg x 1</td>
<td>–</td>
<td>–</td>
<td>400 mg x 1</td>
<td>100 mg x 1</td>
<td>Single dose</td>
</tr>
</tbody>
</table>
Major Side Effects of Treatment

**Rifampin:**
- Orange secretions in tears that stain contact lens

**Clofazamine:**
- Red-brown discoloration due to lipofuscin.

**Dapsone:**
- Hemolytic anemia and methemoglobinemia
Thank you
References


• Google Images
