Leprosy (caused by mycobacterium leprae)
by Brooks David Kimmis, MD

<table>
<thead>
<tr>
<th>Diagnosis/Form of disease</th>
<th>Clinical features</th>
<th>Histopathology</th>
<th>Laboratory evaluation</th>
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<tr>
<td>Lepromatous Leprosy</td>
<td>Multiple, ill-defined, erythematous macules, papules, nodules, and plaques</td>
<td>Virchow cells (foamy-appearing macrophages containing bacilli and lipid droplets)</td>
<td>PCR</td>
<td>2018 WHO Guidelines: 1) For paucibacillary disease (TT &amp; BT), 6-month course of: • Rifampicin 600 mg Qmonth • Clofazamine 300 mg Qmonth and 50 mg daily • Dapsone 100 mg daily 2) For multibacillary disease (LL, BL, BB), same regimen as above, but for 12 months</td>
<td>Leprosy exists on a spectrum from the lepromatous to the tuberculoid form.</td>
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<td></td>
<td>Widespread</td>
<td>Bacilli stain with Gram, Ziehl-Neelsen, or Fite</td>
<td>Slit-skin smear (incision at lesional site with microscopic evaluation of obtained fluid with Fite or Ziehl-Neelsen stain)</td>
<td>2018 WHO Guidelines: 1) For paucibacillary disease (TT &amp; BT), 6-month course of: • Rifampicin 600 mg Qmonth • Clofazamine 300 mg Qmonth and 50 mg daily • Dapsone 100 mg daily 2) For multibacillary disease (LL, BL, BB), same regimen as above, but for 12 months</td>
<td>Tuberculoid leprosy results from a Th1 predominant response and lepromatous leprosy from a Th2 response.</td>
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<td></td>
<td>Symmetric</td>
<td>Grenz zone often present</td>
<td>Organisms are found in 100% of patients with lepromatous leprosy, 75% of borderline leprosy, and 5% of tuberculoid leprosy patients</td>
<td>2018 WHO Guidelines: 1) For paucibacillary disease (TT &amp; BT), 6-month course of: • Rifampicin 600 mg Qmonth • Clofazamine 300 mg Qmonth and 50 mg daily • Dapsone 100 mg daily 2) For multibacillary disease (LL, BL, BB), same regimen as above, but for 12 months</td>
<td>Ridley and Jopling classification: lepromatous, tuberculoid, dimorphous and indeterminant forms</td>
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<td>Favors face, buttocks, lower extremities—requires cool temperatures for growth (30-35°C)</td>
<td>Onion-skin appearance to cutaneous nerves</td>
<td>Nerve conduction studies and peripheral nerve ultrasound may be helpful</td>
<td>2018 WHO Guidelines: 1) For paucibacillary disease (TT &amp; BT), 6-month course of: • Rifampicin 600 mg Qmonth • Clofazamine 300 mg Qmonth and 50 mg daily • Dapsone 100 mg daily 2) For multibacillary disease (LL, BL, BB), same regimen as above, but for 12 months</td>
<td>National Hansen Disease Program Recommendations (for the most part, the US follows these guidelines)</td>
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<td>Sensation unaffected</td>
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<td></td>
<td>Can result in Leonine facies, madarosis, saddle nose, earlobe infiltration, acquired ichthyosis, orchitis</td>
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<td>Enlarged, inflamed, palpable peripheral nerves</td>
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Leprosy exists on a spectrum from the lepromatous to the tuberculoid form.

Tuberculoid leprosy results from a Th1 predominant response and lepromatous leprosy from a Th2 response.

Ridley and Jopling classification: lepromatous, tuberculoid, dimorphous and indeterminant forms

National Hansen Disease Program Recommendations (for the most part, the US follows these guidelines)

1) For paucibacillary disease (TT & BT), 12-month course of:
- Rifampicin 600 mg daily
- Dapsone 100 mg daily

2) For multibacillary disease (LL, BL, & BB), same regimen as above, but for 12 months

Leonine facies differential
- Multicentric reticulohistiocytosis
- Scleromyxedema
- Mycosis fungoides
- Lepromatous Leprosy
- Sarcoidosis
- Nodular mastocytosis
- Systemic Amyloidosis
- Leishmaniasis

Brooks David Kimmis, MD, is a PGY-3 at University of Kansas Medical Center.
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<td>Tuberculoid</td>
<td>Few well-demarcated plaques, which can be erythematous or hypopigmented</td>
<td>Dermal granulomas which may be linear and represents tracking along nerve fibers (“lavender sausages”)</td>
<td>Epithelioid cells and Langhans giant cells surrounded by lymphocytes</td>
<td>Edematous cutaneous nerves</td>
<td>Absent organisms even with staining Nerve involvement distinguishes from other granulomatous processes</td>
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<td>Asymmetric</td>
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<td>Anesthesia and alopecia of lesions</td>
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<td></td>
<td>Neuropathic changes such as neurotrophic ulcers and bone resorption of digits</td>
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<td>Borderline Leprosy</td>
<td>Cutaneous and peripheral nerve involvement related to the predominance of Th1 vs Th2 response</td>
<td>Lepromatous pole: increased bacilli on pathology Tuberculoid pole: decreased bacilli on pathology</td>
<td>Combination of findings seen in lepromatous and tuberculoid leprosy. Can see both Virchow cells and granulomas.</td>
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Leprosy reactions. These clinical findings represent reactive immunologic changes, often in response to treatment.

Type 1 reactions (reversal reactions)
- Change in cell-mediated immunity and Th1 cytokine pattern, often during or following treatment, highest risk in borderline forms
- May be downgrading (borderline leprosy that downgrades towards lepromatous pole) or upgrading (with increase in cell mediated immunity)
- Increased inflammation of existing skin lesions, onset of new lesions, acute neuritis (*emergency), and progressive neurologic impairment. Lacks systemic symptoms (unlike Type 2 reactions)
- Treatment: Prednisone

Type 2 reactions (erythema nodosum leprosum)
- Enhanced humoral immunity and Th2 pattern with immune complex formation
- Occurs in the setting of treatment of leprosy with high bacterial load, including lepromatous and borderline lepromatous leprosy
- Nodules (erythema nodosum-like lesions, which is referred to as erythema nodosum leprosum), and systemic symptoms including fever, myalgias, malaise, joint swelling and pain, lymphadenitis, hepatosplenomegaly, orchitis, glomerulonephritis
- Treatment: Thalidomide

Lucio Phenomenon
- Primarily affecting patients of Central or South American origin
- Thrombosis and necrotizing cutaneous small vessel vasculitis
- Seen in diffuse lepromatous leprosy
- Distal lower extremities with purpura and ulcerative bullae
- Treatment: prednisone

References:
2. Guidelines for the diagnosis, treatment and prevention of leprosy. New Delhi: World Health Organization, Regional Office for South-East Asia; 2017. License: CC BY-NC-SA 3.0 IGO