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Leprosy (caused by mycobacterium leprae)

by Brooks David Kimmis, MD

Diagnosis/ Form of disease	Clinical features	Histopathology	Laboratory evaluation	Treatment	Comments
Lepromatous Leprosy	Multiple, ill-defined, erythematous macules, papules, nodules, and plaques Widespread Symmetric Favors face, buttocks, lower extremities- requires cool temperatures for growth (30-35°C) Sensation unaffected Can result in Leonine facies, madarosis, saddle nose, earlobe infiltration, acquired ichthyosis, orchitis Enlarged, inflamed, palpable peripheral nerves	Virchow cells (foamy-appearing macrophages containing bacilli and lipid droplets) Bacilli stain with Gram, Ziehl-Neelsen, or Fite **Grenz zone** often present Globi (aggregates of bacilli) Onion-skin appearance to cutaneous nerves	PCR Slit-skin smear (incision at lesional site with microscopic evaluation of obtained fluid with Fite or Ziehl-Neelsen stain) Organisms are found in 100% of patients with lepromatous leprosy, 75% of borderline leprosy, and 5% of tuberculoid leprosy patients Nerve conduction studies and peripheral nerve ultrasound may be helpful	2018 WHO Guidelines: 1) For paucibacillary disease (TT & 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 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 (LL, BL, & BB) disease, 24-month course of • Rifampicin 600 mg daily • Clofazamine 50 mg/day • Dapsone 100 mg/day	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. Rabello classification: lepromatous, tuberculoid, dimorphous and indeterminant forms Ridley and Jopling classification: Lepromatous Leprosy (LL), Borderline Lepromatous Leprosy (BL), Mid-borderline Leprosy (BB), Borderline Tuberculous Leprosy (BT), Tuberculoid Leprosy (TT), Indeterminate Leprosy Leonine facies differential • Multicentric reticulohistiocytosis • Scleromyxedema • Mycosis fungoides • Lepromatous Leprosy • Sarcoidosis • Nodular mastocytosis • Systemic Amyloidosis • Leishmaniasis

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Tuberculoid	Few well-demarcated plaques, which can be erythematous or hypopigmented Asymmetric Anesthesia and alopecia of lesions Neuropathic changes such as neurotrophic ulcers and bone resorption of digits	Dermal granulomas which may be linear and represents tracking along nerve fibers ("lavender sausages") Epithelioid cells and Langhans giant cells surrounded by lymphocytes Edematous cutaneous nerves Absent organisms even with staining Nerve involvement distinguishes from other granulomatous processes			
Borderline Leprosy	Cutaneous and peripheral nerve involvement related to the predominance of Th1 vs Th2 response	Lepromatous pole: increased bacilli on pathology Tuberculoid pole: decreased bacilli on pathology Combination of findings seen in lepromatous and tuberculoid leprosy. Can see both Virchow cells and granulomas.			

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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

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