



Overview

- Definition
 - Commonly known as “river blindness,” this eye and skin disease is caused by filaria (worm) *Onchocerca volvulus*, transmitted by female *Simulium* blackfly, which breeds in fast-flowing rivers
 - Second to trachoma as the most common infectious cause of blindness worldwide: >15 million people infected
- Symptoms
 - Photophobia, tearing, foreign body sensation
 - Decreased vision
 - Visual field constriction
 - Nyctalopia
- Laterality
 - Bilateral and symmetric
- Course
 - Several years may separate initial infection and clinical presentation
 - Untreated disease results in blindness
- Age of onset
 - Rare in children and teenagers, but increases in the third decade due to rising microfilarial load
- Gender/race
 - Males more commonly affected due to outdoor responsibilities
 - Most common in equatorial Africa, but also in Central/South America and Eastern Mediterranean
- Systemic association
 - Skin disease
 - 20% of patients are co-infected with *Loa Loa* (Chap. 54), a fact bearing significant treatment implications

Exam: Ocular

Skin and eye diseases are caused by microfilaria (offspring) and not macrofilaria (mother). Ocular disease develops due to dead microfilariae, as living microfilariae are, in fact, well tolerated in the eye.

Anterior Segment

- Conjunctivitis and 0.5- to 2-mm diameter conjunctival nodules
- Limbal edema and hyperemia
- Dead microfilariae can be seen directly in the cornea, appearing straight and opaque
- Punctate or sclerosing keratitis
- Anterior uveitis is rare and does not correlate with microfilaria load, and varies from low-grade, non-granulomatous to severe, turbid, granulomatous
- Pupillary seclusion leading to angle closure glaucoma
- Cataract

Posterior Segment

- Pigment clumping and RPE atrophy, either diffuse or geographic with distinct borders, located temporal to the macula or nasal to the optic nerve
- Cotton wool spots and intraretinal hemorrhage
- Intraretinal worms can be seen as reflective opacities with green tint
- Vascular sheathing
- Optic neuritis resulting in optic atrophy and peripapillary hyperpigmentation

Exam: Systemic

- Dermatitis papules is most common with pruritus at acute stage
- Pretibial skin depigmentation (“leopard skin”)
- Chronic disease results in skin lichenification and scarring, atrophy, pigment changes, especially on buttocks, waist, shoulders
- Facial skin eruption and purplish lesions on upper body are rare but seen in Central America
- Groups of round, painless, subcutaneous nodules with firm fibrous capsule containing adult worms (15–40 mm, lifespan 10 years)
- Lymphatic obstruction with microfilariae, generally inguinal or femoral

Imaging

- FA: mottled fluorescence around RPE atrophy
- Visual field: diffuse constriction

Laboratory and Radiographic Testing

- Sensitive, low-cost detection of antigens in tears, urine, dermal fluid
- PCR assay with superficial skin scratch, or microscopic skin snip evaluation
- Ultrasonography to detect and evaluate nodules
- ELISA and radioimmunoassay test for parasite specific antibodies

Differential Diagnosis

- Contact dermatitis
- Scabies
- Prickly heat
- Insect bites
- Leprosy, yaws, or superficial mycosis
- Nodule differentiation from lymph node, lipoma, fibroma, dermal cyst, ganglia

Treatment

- Ivermectin is the treatment of choice
 - Kills microfilariae but not macrofilariae
 - Given at 150 mcg/kg in one oral dose every 6–12 months, both adults and children
 - Length of treatment depends on disease activity (continuous skin manifestation is a good gauge) and whether the patient still lives in endemic areas
 - However, serious and sometimes fatal adverse reactions occur in those co-infected with loiasis
- Doxycycline is emerging as a treatment to kill or sterilize macrofilariae
 - Kills *Wolbachia*, an endosymbiotic bacteria required for the survival of *O. volvulus* macrofilariae and embryogenesis
 - Given at 200 mg PO QD for 6 weeks
 - Since it does not kill microfilariae, ivermectin may still need to be given to reduce symptoms
 - Limited data suggest it may be safe in loiasis co-infection
- Topical corticosteroids and cycloplegia to reduce ocular inflammation and positive pupillary seclusion and resultant angle closure glaucoma

Referral/Co-management

- Dermatology and/or Interventional Radiology for nodulectomy