Mucocutaneous leishmaniasis

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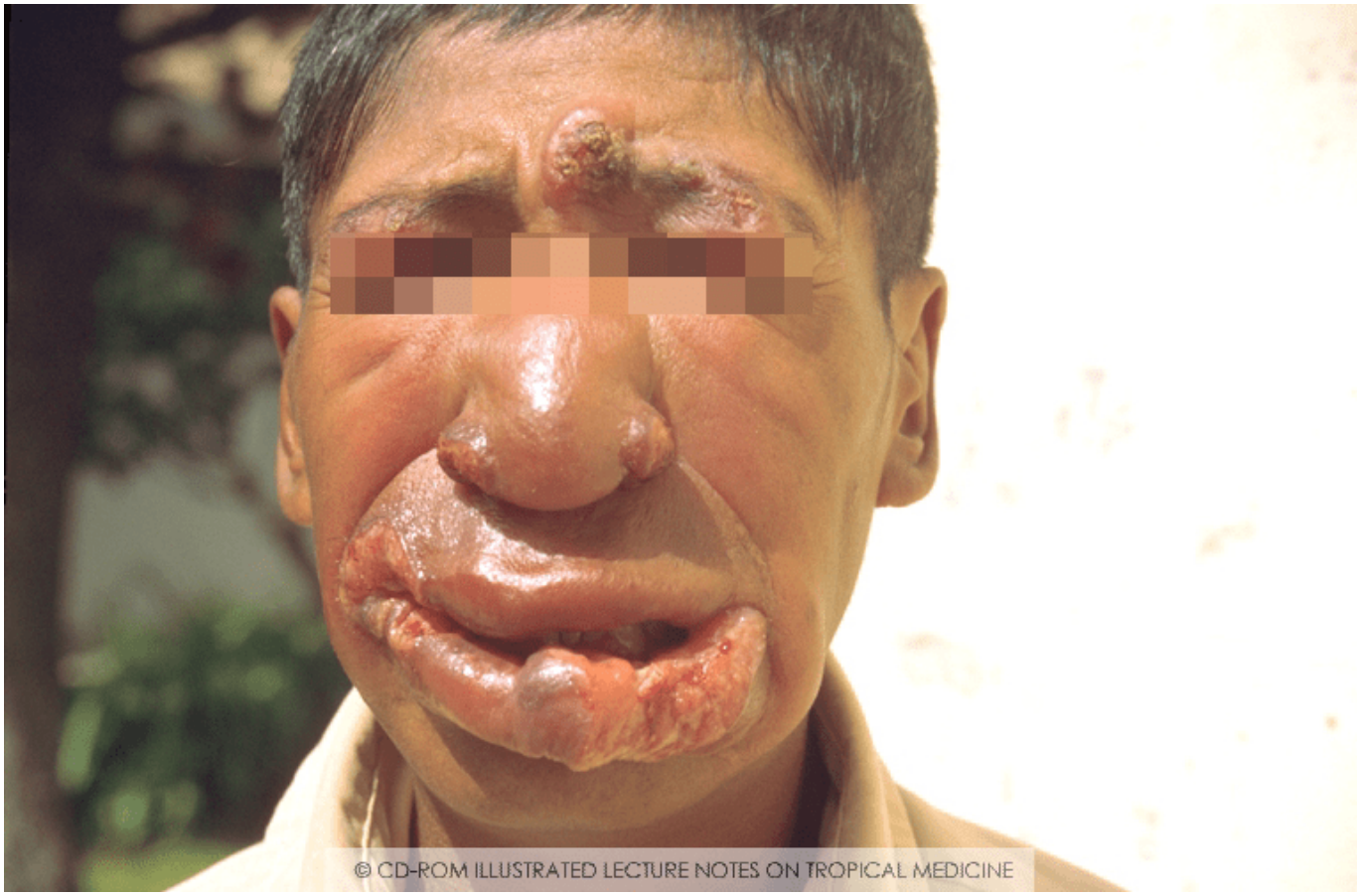
Distribution

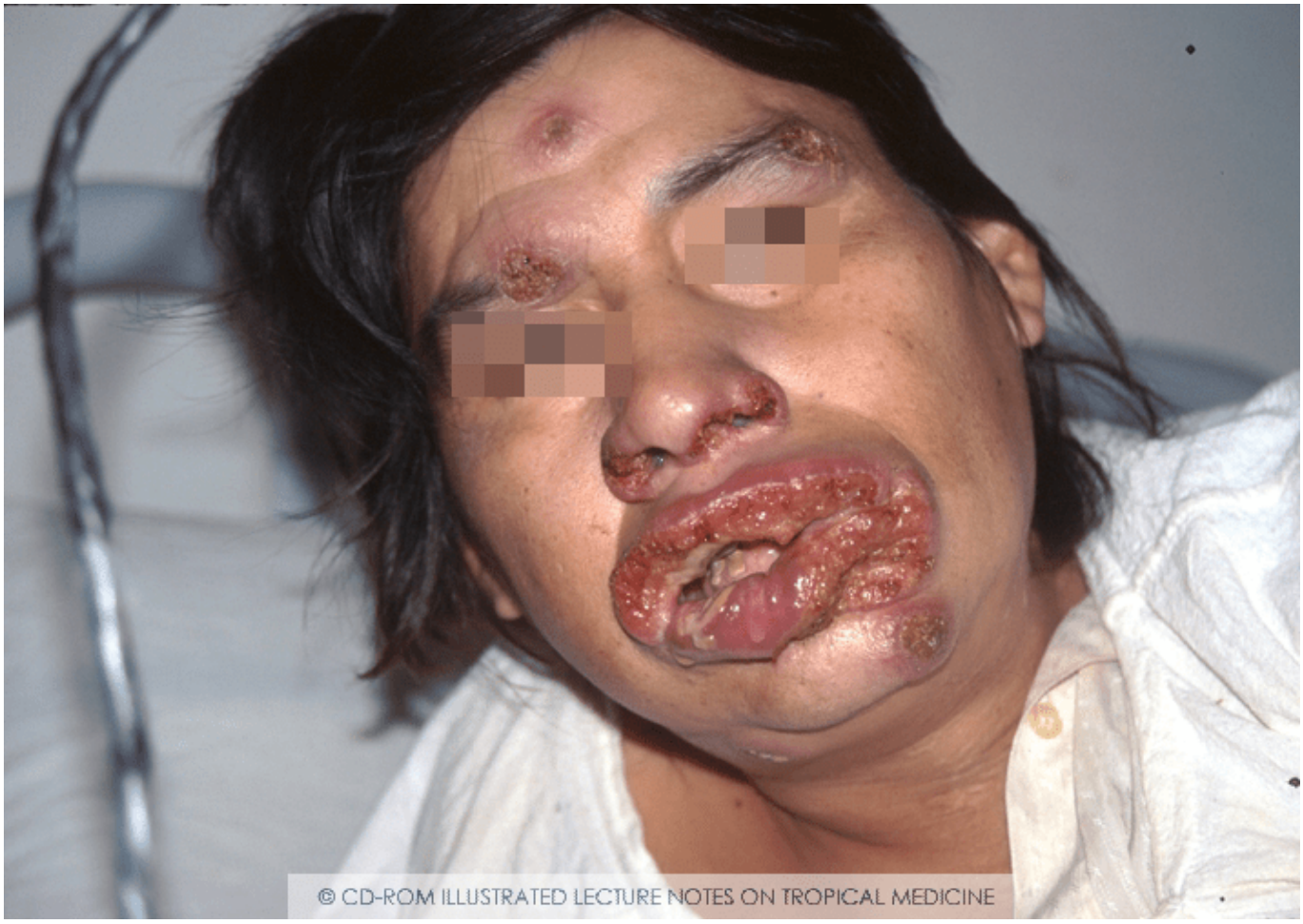
At present 90% of all mucocutaneous leishmaniasis occurs in Bolivia, Peru and Brazil. Illustrations of skin lesions and disfigurements suggestive of leishmaniasis are encountered on pre-Inca earthenware. These indicate that the disease was already in existence in Peru and Ecuador in the 1st century AD. Texts dating from the 15-16th century Inca period and the Spanish conquest mention the risk of cutaneous ulcers in seasonal farmers. Espundia was also described as “white leprosy”.

Clinical aspects

When skin and mucosae are affected the disease is known as mucocutaneous leishmaniasis. This is very rare in East Africa but frequent in South America, where it is known as “espundia”. After an initial skin lesion, that slowly but spontaneously heals, chronic ulcers appear after months or years on the skin, mouth and nose, with destruction of underlying tissue (nasal cartilage, for example). Tissue destruction with disfigurement can be very severe. Parasites are usually rare in the lesions. A substantial part of the disfigurement is possibly due to immunological mechanisms. One hypothesis is a relationship between the occurrence of mucocutaneous lesions and the presence of certain alleles of polymorphic tumour necrosis factor a and b genes.



Espundia or mucocutaneous leishmaniasis often results from infection with Leishmania brasiliensis. Photo Cochabamba, Bolivia



Espundia or mucocutaneous leishmaniasis often results from infection with Leishmania brasiliensis. Photo Cochabamba, Bolivia

Diagnosis

The lesions often contain few parasites. Diagnosis is sometimes made solely on a clinical basis. Culture of the parasites is possible, but not really feasible in primitive rural conditions. Serology in espundia can be positive or negative (the quality of the antigen is of crucial importance). A practical problem in South America is whether a certain skin lesion with *Leishmania* amastigotes is caused by *L. braziliensis* or not. The geographical origin of the lesion or PCR and/or zymodeme analyses may give an answer here, though these laboratory techniques are not available in rural areas.

Mucocutaneous leishmaniasis, differential diagnosis:

Differential diagnosis includes skin cancer, tertiary syphilis and yaws, leprosy, rhinoscleroma (a very chronic granulomatous infection with *Klebsiella rhinoscleromatis*), rhinosporidiosis, midline granuloma (a form of T-cell lymphoma), Wegener's granulomatosis, sarcoidosis, skin tuberculosis, infection with the free-living amoeba *Balamuthia mandrillaris*, chronic nasal cocaine abuse, noma, and fungal infections such cryptococcosis, histoplasmosis and South American blastomycosis (paracoccidioidomycosis). With this last disease, which is a very chronic infection, the lungs are frequently affected in a manner that can mimic tuberculosis. The yeast has typical oval cells with ectospores and can be detected in sputum.

Overview: Differential diagnosis of nasal ulcers:

1. Mucocutaneous leishmaniasis (espundia)
2. Fungal infections, such as paracoccidioidomycosis (syn. South American blastomycosis), histoplasmosis, cryptococcosis, coccidioidomycosis
3. Actinomycosis
4. Treponematoses (syphilis, yaws, bejel)
5. Leprosy
6. Tuberculosis
7. Rhinosporidiosis
8. Rhinoscleroma (chronic infection with *Klebsiella rhinoscleromatis*)
9. Balamuthiasis (infection with free-living amoeba)

Non-infectious

1. Granulomatosis with Polyangitis (formerly Wegener granulomatosis)
2. Midline granuloma (a form of T-cell lymphoma)
3. Other non-Hodgkin lymphoma
4. Squamous cell carcinoma
5. Sarcoidosis
6. Relapsing polychondritis
7. Cocaine abuse

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