Patient History

A 23-year-old woman originally from a rural area in Paraguay, South America, consulted for a lesion of the upper lip that had been present for 1 year and was progressively increasing in size. She had no prior history of injury.

Physical Examination

The examination revealed thickening of the upper lip, with marked infiltration and induration. The mucosa of the upper lip, hard palate, and upper gums showed multiple eroded erythematous papules with a cobblestone or mulberry-like appearance (Figure 1).

Additional Tests

The blood tests and chest radiograph were normal. Direct microscopic examination of the exudate of the lesion revealed round fungal structures with double-contoured walls and budding. Cultures in enriched Sabouraud medium at 37°C showed slow-growing rough, cream-colored colonies.

Histopathology

The histopathologic study showed a severe granulomatous reaction in the dermis, with numerous multinucleated giant cells that contained small structures with a rounded morphology and thick, double-contoured walls, some with superficial budding (Figures 2 and 3).

What is your diagnosis?
Diagnosis
Paracoccidioidomycosis

Course and Treatment
Treatment was initiated with itraconazole, 200 mg/d, and a good therapeutic response was observed at 2 months. At the time of writing, the patient had not returned for further follow-up.

Comment
Paracoccidioidomycosis is a systemic deep mycosis that is endemic in the rural areas of Central and South America. The causative agent is *Paracoccidioides brasiliensis*, a dimorphic fungus that can present as a saprophytic mycelium or as a pathogenic yeast. The fungus usually enters humans through the lungs, producing an initial infection that is often asymptomatic, particularly in young subjects. Afterwards, blood-borne dissemination can occur, affecting the skin, mucosa, and other organs. On occasion, the fungus enters through a skin or mucosal injury.\(^1\),\(^2\),\(^4\),\(^5\)

Mucocutaneous manifestations predominate in the oral mucosa. These consist of ulcerated and hypertrophic papular erosive lesions (mulberry-like stomatitis) that cause a thrombus-like lip. Periodontal abscesses occur on occasion, with tooth loss and involvement of the tonsils, epiglottis, and larynx. The nasal region is also affected with ulcers and scabby lesions and destruction of the nasal septum, simulating American mucocutaneous leishmaniasis. Ulcerated or crusted nodules with a bleeding and granulomatous base have also been reported; these nodules can affect any part of the integument, but usually appear around the natural orifices of the body. The enlarged lymph nodes can suppurate and necrose, leading to fistulae similar to those observed in tuberculosis.\(^1\),\(^4\)

The condition is diagnosed by direct microscopic examination of lesion exudate using potassium hydroxide, Giemsa, or Grocott stain, in which the round, double-walled yeast cells with multiple budding (characteristic helm-wheel appearance) are seen. Culturing in Sabouraud agar or blood agar confirms the diagnosis.\(^4\),\(^6\) The histopathology is also conclusive if the "helm-wheel" structures are observed inside the multinucleated giant cells or free in the suppurative areas.\(^1\),\(^4\)

In severe forms, the treatment of choice is intravenous amphotericin B. Oral itraconazole therapy consisting of 200 to 400 mg/d is preferable in all other clinical forms. The duration of itraconazole therapy varies from 6 to 12 months for mild cases and 12 to 18 months for moderate cases.\(^4\)

Conflicts of Interest
The authors declare no conflicts of interest.

References