Pathologic Quiz Case

An Unusual Infection in a Human Immunodeficiency Virus–Positive Man

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The patient was a 33-year-old African American man who had tested positive for human immunodeficiency virus type 1 (HIV-1) approximately 14 months prior to presentation. He was referred to our hospital for persistent sinusitis. His medical history included previous intravenous drug use, positive hepatitis C titers, Candida esophagitis, and presumptive *Pneumocystis carinii* pneumonia.

The patient had been hospitalized 3 times because of his sinusitis and had been placed on ceftriaxone, cefaclor, amoxicillin/clavulanate potassium, ceftazidime, and ciprofloxacin. He improved with each intravenous antibiotic; however, once placed on an oral dosage, his symptoms would return. On one admission, *Pseudomonas aeruginosa* was cultured from his nasal discharge.

On physical examination, we found approximately 80% destruction of the septum, as well as extensive necrosis of the inferior turbinate, middle turbinate, and the ethmoid region (Figure 1). There were several crusted draining skin lesions diffusely present on the extremities and trunk, which measured approximately 0.5 to 1.0 cm in diameter (Figure 2). Radiographic examination was unremarkable.

Cultures of the nasal tissue grew *Pseudomonas aeruginosa.* Cultures for mycobacterium were positive, and DNA probes identified *Mycobacterium avium* in the sputum, blood, bone marrow, nasal tissue, and the skin. Cultures were negative for anaerobic bacteria, fungi, and viruses.

A biopsy received from the nasal cavity consisted of multiple tan to white fragments. The skin biopsy consisted of a dark ellipse without a grossly described lesion. Microscopically, the nasal tissue demonstrated chronic necrotizing granulomatous inflammation and many spherules, approximately 10 μm in diameter, with thick, refractile, eosinophilic cell walls; flocculent cytoplasm; and round nuclei with a large central nucleolus. The walls stained strongly for periodic acid–Schiff after diastase pretreatment (Figure 3), weakly with methenamine silver, and did not stain with mucicarmine. Biopsies from the skin demonstrated acute folliculitis with abscess formation, as well as morphologically and histochemically similar organisms.

What is your diagnosis?

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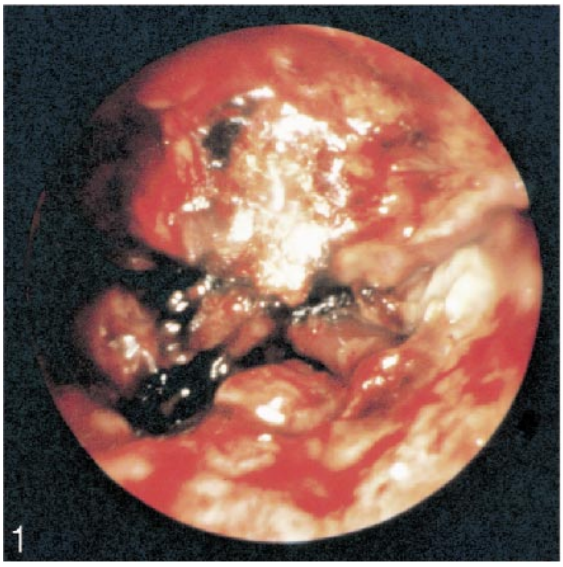

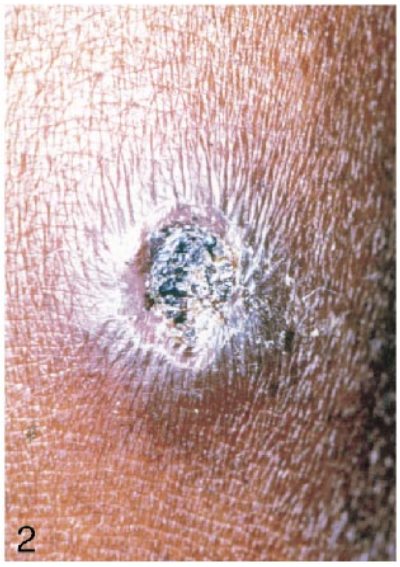

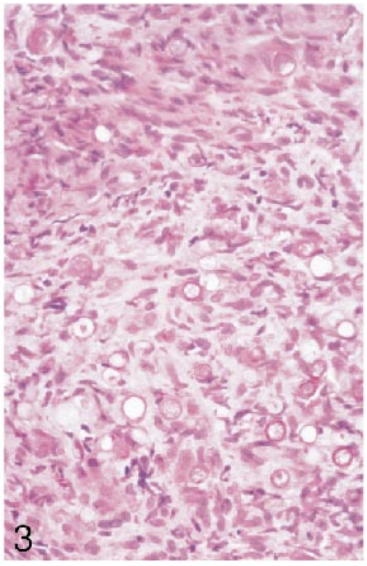

Pathologic Diagnosis: Rhinosporidiosis

The patient was admitted and given intravenous dapsone for rhinosporidia infection, as well as antimycobacterium agents, antiviral drugs, and antibiotics to treat the *Pseudomonas* infection. Upon further questioning, the patient reported swimming in cattle stock ponds. His skin lesions were felt to be a result of autoinoculation from the nasal cavity. He was discharged after 15 days and chose not to follow up with our institution.

COMMENT

Epidemiology and Etiology

Rhinosporidiosis is a fungal disease that usually affects the mucus membrane, but infection has been found in other regions of the body as well. The etiologic agent, *Rhinosporidium seeberi,* has not yet been isolated, and animal inoculation appears to be ineffective.[1] The disease was first described in 1892 and was thought to be a parasite until it was reclassified in 1923 as a fungus.[2] The majority of cases (88%) occur in southern India and Sri Lanka; however, a few cases have been reported in South and Central America. In the United States, only 40 to 50 cases have been diagnosed since it was discovered in a native Tennessee farmer in 1907.[1] Humans are not alone in contracting rhinosporidiosis; cows, horses, and mules have been diagnosed with the disease as well.[3] Based on the cases reported to date, males contract the disease 70% to 80% of the time, and the age of patients ranges from 3 to 90 years. The disease has been correlated with working or playing in stagnant fresh water; however, populations living in arid regions where dust storms are common have a higher incidence of ocular infections.[4] Trauma seems to be necessary to introduce the fungus, and disseminated infections are rare.[5] Most of the cases reported in the United States have occurred in Texas.

The primary site of infection tends to be the nares (70%); ocular lesions account for 15% of cases and other mucocutaneous sites account for 8%. Rarely, infections occur in the skin.[1]

Pathogenesis and Pathology

Nasal lesions often present as a sessile or pedunculated polyp with white projections, which give the polyp a ''strawberry'' appearance. These lesions are felt to be sporangia, and the spherules can be appreciated macroscopically. The polyps can become quite large and have been reported to hang out of the nose and extend to the lips.These lesions commonly spread into the sinuses or serve as reservoirs for autoinoculation of other sites. They present histologically as a chronic granulomatous reaction with increased vascularity. The epithelium may become invaginated and form pseudocysts. Mature sporangia often lie just below the surface, but all stages of development will be present. Sporangia stain well with methenamine silver, periodic acid–Schiff, and Gridley, but the trophocytes may stain weakly. Staining with mucicarmine can be positive for selected elements.

Ocular infections account for almost all cases in dry dusty climates. Commonly, the lesion lies between the lid and the eyeball. Previous sites of injury have also become infected, and may be mistaken clinically for hemangiomas.[1]

Infections of the skin are uncommon and usually represent concomitant mucocutaneous disease. Lesions begin as tiny papules that develop into friable warty growths, which often ulcerate.[3]

Laboratory Identification

The fungus cannot be detected by serology,[6] and molecular approaches have been advocated.[7] A single study demonstrated rhinosporidiosis cultured on potato dextrose agar; however, these results have not been reproduced.[8] Diagnosis is made by histologic examination of the infected tissue.

Prognosis and Therapy

Rhinosporidiosis is a chronic disease that can exist in the body for decades without causing major debility. Reoccurrence is a problem, however, as most patients require multiple surgical excisions for growth removal.[9] There is no apparent immunity developed against this disease,[1] and treatment consists primary of surgical debridement. Local injections of amphotericin B have been tried to limit spread after surgical excision, but no studies exist to prove this drug inhibits growth. Dapsone has been shown to lessen the rate of reoccurrence from 93% to 28% over a 3-year period, but this treatment is not considered curative.[1]

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