Disseminated Rhinosporidiosis Masquerading as Sarcoma

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Abstract
Rhinosporidiosis, though reported from several countries, has the highest incidence in India (notably South India) and Sri Lanka, with most cases presenting as nasal polyps. Nasopharynx and palpebral conjunctiva account for 85% of the sites. Other mucosal sites affected are oropharynx, larynx, rectum and external genitalia. Disseminated rhinosporidiosis is usually associated with mucosal lesions. Rhinosporidiosis affecting skin, subcutaneous tissue and bone are uncommon. Herein we report a rare case of disseminated rhinosporidiosis mimicking a soft tissue sarcoma.

Introduction
Rhinosporidiosis is a granulomatous disease caused by the protistan Rhinosporidium seeberi (R. seeberi). Rhinosporidiosis has been known for over a century since its first description in Argentina. It occurs universally, although it is endemic in South Asia, notably Southern India and Sri Lanka. The disease is presumed to be transmitted through infected soil and water. Stagnant water is a potential source of infection as most infected patients are from rural background, and have the habit of bathing in ponds. It is a chronic disease, with frequent recurrence after surgery, and occasional dissemination from the initial focus which is commonly the upper respiratory tract. The case presented here is to depict the unusual presentation of rhinosporidiosis as a muscle mass, mimicking a tumor.

Case Report
A 52-year-old man with swellings over the left calf and right elbow presented to our outpatient clinic. He was an agricultural worker in rural Tamil Nadu, South India. He complained that the painless masses were slowly growing over past few months. At the clinic, he appeared comfortable; his vital signs were normal. There was a mass in his left calf measuring 10 cm x 7 cm, firm and indurated. He also had a dumb-bell shaped swelling over his right elbow measuring 8 cm x 5 cm; the swelling was firm, fixed and non-tender with no signs of inflammation (Figure 1). Rest of the examination was unremarkable. Routine laboratory values revealed normocytic anemia. A MRI of the left leg (Figure 2) showed a lobulated mass in the subcutaneous fat plane of the calf with moderate heterogenous enhancement (coronal T1W1). Irregular, relatively non enhancing areas were seen centrally within the moderately enhancing matrix without involvement of underlying bone. Based on the clinical and MRI findings, a clinical diagnosis of sarcoma involving the calf muscle was made.

He underwent complete excision of the mass over the left calf and right elbow. Intra-operatively, both the masses were found to be well-encapsulated and both were invading the muscle planes (Figure 3). Histopathology of the excised calf specimen showed rhinosporidial sporangia with spores in multiple stages of development, invading skeletal muscle fibres (Figure 4). Biopsy specimen from right elbow mass also revealed multiple rhinosporidial spores with muscle invasion.

On further examination, multiple polypoidal masses were noted in the nasal cavity (Figure 5). On enquiry, he described his habit of bathing in a pond for several years; additionally he had symptoms of persistent nasal obstruction.

Hence, he was diagnosed as disseminated rhinosporidiosis involving two non-contiguous intramuscular sites.

The patient refused to undergo excision of the intra-nasal lesions. Therapy was started with dapsone 100 mg/day. He was advised not to bathe in stagnant water any more. At 5 month follow-up, he remains asymptomatic without any evidence of recurrence.

Discussion
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usually associated with mucosal sites affected are oropharynx, nasopharynx and palpebral conjunctiva.

R. seeberi was initially classified as a fungus, but now, using fluorescence in situ hybridization and R. seeberi-specific PCR, it is identified as a novel class of aquatic protistan parasite infecting fish and amphibians. Since R. seeberi is difficult to culture, details of its life cycle are unclear. Mature spores give rise to electron dense units which are considered infective. Rhinosporidiosis is an infective disease in the sense that the tissue lesions are always associated with the presence of the pathogen. No transmission has ever been documented of cross-infection between members of the same family or between animals and humans and thus there is no evidence that it is contagious.

Definite mode of transmission of the pathogen and natural reservoirs are not yet known; the disease is presumed to be transmitted through infected soil and water. Stagnant water is suggested as a potential source of infection as well as in asymptomatic persons in Sri Lanka; some have been reported from South America and Africa. R. seeberi (R. seeberi) affecting the mucous membranes. First described in 1900 by Guillermo Seeber in Argentina, it generally presents as swollen, pink or red polyps in the nasal cavity or the ocular conjunctivae. Most of the reported cases are from Southern India or Sri Lanka; some have been reported from South America and Africa.1

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The definitive diagnosis of rhinosporidiosis is by histopathology of biopsied or resected tissues, with the identification of the pathogen in its different stages.3 Coccioides spherules has very similar morphology, but its sporangia and endospores are smaller than R. seeberi spherules, and inner sporangial wall of R. seeberi stains with mucicarmine. In her study, Anuradha et al found that, in all the 17 cases that were initially evaluated on fine needle aspiration cytology (FNAC) or with combined aspiration and scrape cytology, rhinosporidiosis was correctly diagnosed, which later on was confirmed by histopathology. This suggests that FNAC could be useful screening tool through histopathology is definitive.6 Anti-rhinosporidial antibody response occurs in rhinosporidial patients, as well as in asymptomatic persons who were exposed to R. seeberi in the environment. Anti-R. seeberi antibody does not appear to be protective in rhinosporidiosis since appreciable titres were present in patients with recurrent, single, multiple or disseminated lesions of long duration.7

Excision is the definitive treatment for rhinosporidiosis, however recurrence is common due to traumatic inoculation. Antimicrobial therapy for this infection is sub-optimal. The only drug which appears to have some anti-rhinosporidial effect is dapsone given at a dose of 100mg/day for a period of 6 months in recurrent and disseminated rhinosporidiosis. Dapsone appears to arrest the maturation of sporangia and promotes fibrosis in stroma.8 Our patient was advised not to bathe in stagnant water. Call for attention to sanitation measures and better therapeutic intervention are urgently needed for this neglected, disfiguring infection.

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References