



## Nasal Rhinosporidiosis-A Clinicopathological Case Report

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### Abstract

*Rhinosporidiosis is a disease caused by Rhinosporidium seeberi which primarily affects the mucosa of the nose, conjunctiva and urethra. While it is endemic in some Asian regions, isolated cases are reported in other parts of the world also.. Its manifests as a polypoidal mass growing inside the affected cavity and the only treatment is surgical excision. Rhinosporidiosis is a condition which both clinicians and pathologists should keep in mind when managing patients with nasal masses even from non endemic areas. It is critical in such cases to follow the clinical course to ensure against the recurrence of disease. This study describes the clinical features, histological diagnosis, and treatment of rhinosporidiosis of the nose in a case in Jaipur, India.*

**Keywords;** *Rhinosporidiosis, histological diagnosis, Rhinosporidium seeberi.*

### Introduction

Rhinosporidiosis is a rare chronic granulomatous disease endemic in some parts of Asia, such as India and Sri Lanka, but infections have been reported to have occurred in America, Europe and Africa. It is found in the tropics due to increased migration of those who have acquired rhinosporidiosis in their native Asian countries to the west<sup>[1]</sup>. Rhinosporidiosis is a chronic localized infection of the mucus membranes and the lesions present clinically as polypoidal, soft masses of the nose, throat, ear, and even the genitalia. The presumed mode of infection from the natural aquatic habitat of *Rhinosporidium seeberi* is through the traumatized epithelium most commonly in nasal sites.<sup>[2]</sup>

The etiological agent is *Rhinosporidium seeberi*, the taxonomy of which has been debated in the

last few decades since the microorganism is intractable to isolation and microbiological culture<sup>[3]</sup>. In the 1890s an apparent sporozoan parasite was described in nasal polyps and was named *Coccidium seeberia* after the protozoal subdivision *Coccidia*. Later in the early 1900s the life cycle of the organism was described and it was argued to be a fungus with a proposed name *Rhinosporidium seeberi*. Since then, the microbe has been considered a fungus by most pathologists and microbiologists, although its taxonomy has been debated. Through phylogenetic analysis of *Rhinosporidium seeberi* 18S rRNA gene, this group of pathogens was originally identified by Ragan et al. as in the DRIP clade (acronym derived from *Dermocystidium*, rosette agent, *Ichthyophonus* and *Psorospermium*)<sup>[4]</sup>. Herr et al. replaced it with the term *Mesomycetozoa*

(between fungi and animals)<sup>[5]</sup>. The phylogenetic distribution of this novel group of parasites suggests that the features they possibly share with early diverging animals and fungi may offer clues on the appearance of this ancestor. The phylogenetic hunt reassured that *Rhinosporidium seeberi* produces endosporulating cells in their infected host and the presence of chitin synthase genes reduced the divergence of its existence.

The infectious agent forms round and thick-walled sporangia in the submucosa of the affected site, varying from 10 – 200 μm in size, which are visible as white dots in the mucosa containing smaller ‘daughter cells’ (called ‘sporangiospores’). It can be visualized with fungal stains such as Gomori methenamine silver (GMS) and periodic acid-Schiff (PAS), as well as with standard haematoxylin and eosin (H&E) staining. The only curative approach is the surgical excision combined with electrocoagulation. The failure to propagate *Rhinosporidium seeberi* in vitro has prevented the determination of its in vitro sensitivity to drugs that might have clinical application. Recurrence, dissemination in anatomically close sites and local secondary bacterial infections are the most frequent complications<sup>[1]</sup>.

We present here a case of rhinosporidiosis that presented in our tertiary care hospital in Jaipur, emphasizing the clinical presentation, diagnosis and management for the prevention of recurrence to the best of our knowledge

### Case Presentation

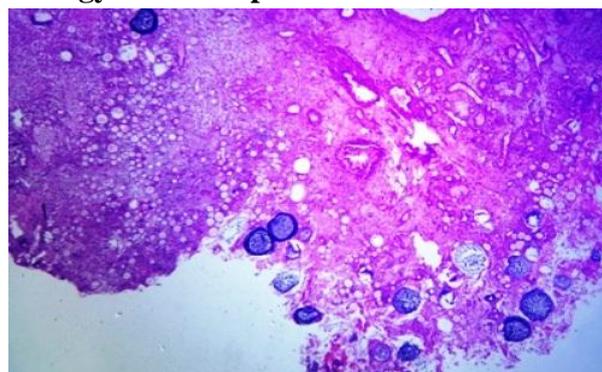
A 35-year-old man, a native of Jaipur, presented to our tertiary care hospital with a history of foreign body sensation and a small painless mass in the right nostril, associated with occasional epistaxis and nasal discharge since 6 months. Although there was no history of trauma, there was a history of animal handling and contact with contaminated water. He was a milkman by profession, residing in a suburban village near to the hospital. Physical examination showed a polyp, pinkish red in colour, nearly 10 mm

diameter. An apparent diagnosis of rhinosporidiosis was made. The entire mass was resected and sent for histopathological and microbiological study.

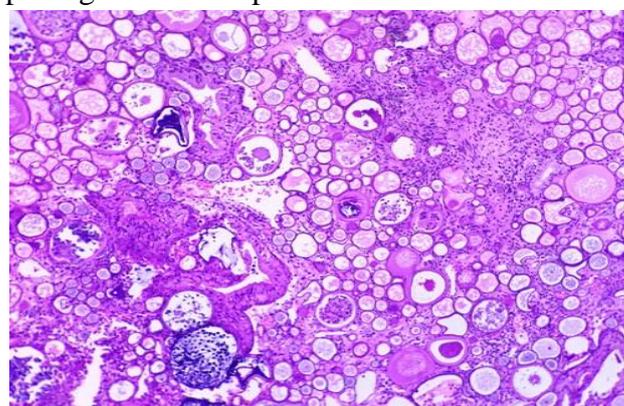
The diagnosis was confirmed on histological examination. Macroscopically, the mass was pink and fleshy, studded with scattered gray-white spots on the surface. The mass section was stained with a hematoxylin and eosin stain and studies showed multiple giant cells and lymphocytes around the mature sporangium. Microscopically, the lesion had the characteristic features of rhinosporidiosis, i.e., hyperplastic squamous epithelium, edematous fibro-connective tissue containing many thick-walled globular cysts (sporangia), which in turn contained numerous endospores. He made an uneventful recovery after the excisional surgery.(Image 1-4)

### Microscopic Examination

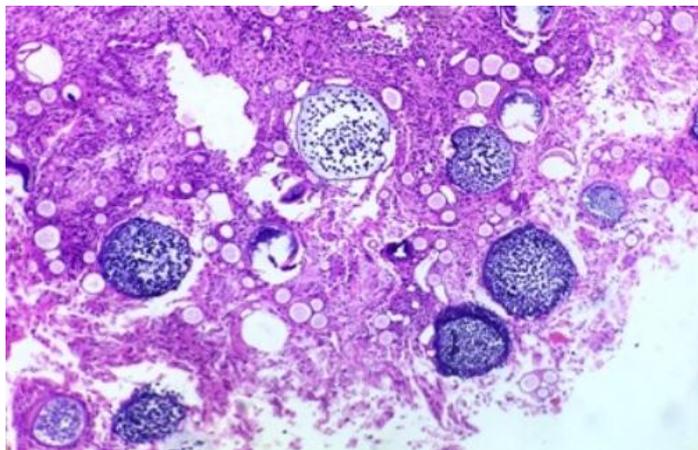
#### Histology of Rhinosporidiosis



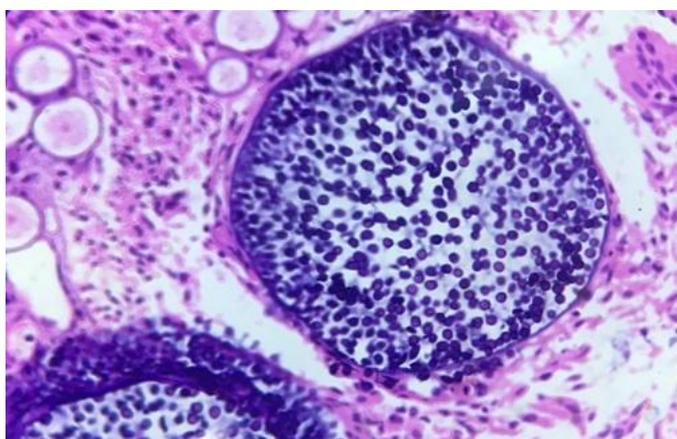
**Image 1** (10x) Histopathology of the resected specimen shows nasal subepithelium with sporangia of *Rhinosporidium*.



**Image 2** (40X) The thick walls of immature *R. seeberi* trophocytes stain with PAS (pink), and the spherical organisms are surrounded by inflammatory cells.



**Image 3** (40x) Various stages of sporangia seen: young, collapsed and empty forms and bigger mature forms towards the surface



**Image 4** (40x) Detailed image shows hundreds of spores within the mature forms

### Discussion

This study documents and reports a case of nasal rhinosporidiosis in Jaipur. The disease has been reported from about 70 countries with diverse geographical features<sup>[2,6]</sup>. Infrequently, isolated cases are reported in other parts of the world, mainly due to migration<sup>[1,7]</sup>. In contrast with more recent fungal infections, some aspects of the taxonomy, morphology, ontogenesis and epidemiology of those caused by *Rhinosporidium seeberi* remain controversial and have not been resolved. Though now related to a group of fish parasites referred to as the DRIP clade, most pathologists and microbiologists initially considered it a fungus on the basis of its property to be stained by fungal stains such as GMS and PAS<sup>[1]</sup>. A study done by Silva et al using phylogenetic analysis of the complete internal

transcribed spacer sequences raises the possibility that the genus *Rhinosporidium* may possess multiple host-specific strains and indicates that *Rhinosporidium seeberi* recovered from humans could have diverged according to its geographical location<sup>[8]</sup>. These authors indicated that a combination of host specificities and resistance of *Rhinosporidium* to grow in culture may account for the failure to produce experimental rhinosporidiosis. Ajello and Mendoza effectively proposed its class Mesomycetozoa<sup>[9]</sup>. Recent studies done using fluorescent in-situ-hybridization techniques provide evidence that its natural habitat are water reservoirs and perhaps soil contaminated by waste<sup>[10]</sup>. In addition, other aquatic micro-organisms might be relevant to a possible synergistic action in the establishment of natural rhinosporidiosis. There are examples of such synergism between bacteria and parasites, e.g., lactobacilli with *Trichomonas* and *Wolbachia* with filarial nematodes. The class Mesomycetozoa has two orders, that is, the Dermocystida and the Ichthyophonida. In the order Dermocystida is the family Rhinosporideaceae which includes *Rhinosporidium seeberi* *Dermocystidium* spp. and the rosette agent. In the order Ichthyophonida, the class Ichthyophonae has members with phylogenetic features in common with the genus *Ichthyophonus* and *Psorospermium*<sup>[9]</sup>.

The route of transmission of *Rhinosporidium* remains unclear even though the presumed mode of infection from the natural aquatic habitat of *Rhinosporidium seeberi* is through the traumatized epithelium ('transepithelial infection'), most commonly in nasal sites. Various modes of spread have been documented by several workers including; (i) auto-inoculation through spillage of endospores from polyps after trauma or surgery, (ii) haematogenous dissemination to distant sites, (iii) lymphatic routes, and (iv) sexual<sup>[11]</sup>.

The disease is prevalent in rural settings, particularly among individuals working or in contact with contaminated soil, stagnant water (ponds, or lakes) or sand. In our case series, the patient did give a history of contact with

contaminated pond water. Patient belonged to a rural area having contact with feces of infected livestock and even worked in contaminated agricultural fields. A curious feature in the incidence of the disease is that while several hundred people bathe in the stagnant waters, only a few develop progressive disease. This might indicate the existence of predisposing, though obscure, factors in the host. The possibility that nonspecific immune reactivity in the host, blood group and HLA types has been suggested by various investigations as possibly important in the pathogenesis of *Rhinosporidium seeberii* to establish an initial focus of infection<sup>[2]</sup>.

As the disease has a slow course, lesions may be present for many years before the patients become symptomatic which was true in our case. Rhinosporidiosis manifests as tumor-like masses, usually of the nasal mucosa or ocular, conjunctivae of humans and animals. Patients with nasal involvement often have masses leading to nasal obstruction or bleeding due to polyp formation and it can spread to the nasopharynx, oropharynx, and the maxillary antrum, as was evident in one of our cases. The diagnosis is established by observing the characteristic appearance of the organism in tissue biopsies and CT scans. The lesion is friable, a vascular pedunculated or sessile polyp, with a surface studded with tiny white dots due to spores beneath the epithelium, giving a 'strawberry-like' appearance which was evident in our case. This made the clinical diagnosis relatively easy to establish. Apart from this appearance, lesions have been associated with other areas in the head and neck region and urethral, vaginal and rectum<sup>[12]</sup>. Systemic disease is rare but can include multiple mucocutaneous, hepatic, renal, pulmonary, splenic or bone lesions, associated with fever, wasting, and even death<sup>[13]</sup>. The disease is more common in younger age groups as has been observed by various authors. An uncommon pathogen, typically restricted to tropical areas and seems to occur more in the younger age group, more so in men, as this group

is frequently occupationally active (agriculturists, sand workers, divers etc). Less outdoor activity and less chance of contact with animals could explain fewer incidences among women<sup>[14]</sup>. This finding was evident in our case as the case was in a young male. In addition, Indian social, cultural habits and the custom of bathing in open ponds expose individuals to several innocuous water-borne organisms. The epidemiology of rhinosporidiosis still remains unclear and the phylogenetic relationship of its life cycle creates difficulties in understanding the actual incidence of infection and the populations of patients at risk. Many investigations are therefore needed to understand whether rhinosporidiosis is acquired in particular communities or if unrecognized factors exist that may explain the emerging epidemiology of this infection.

With no significant travel history to the known endemic states of India for rhinosporidiosis, nor having any contact with infected patients as per the history given by them, we presumed that the patients in our cases acquired the infection locally. Spontaneous regression of rhinosporidial growths has been noted in animals and in humans but is rare. Therefore, medical and/or surgical intervention is necessary.

Wide local surgical excision of the lesion is the treatment of choice to reduce the risk of recurrence, though this may be associated with significant morbidity due to hemorrhage and nasal septal perforation. So, limited surgical excision and adjuvant medical therapies, including antifungals such as griseofluvin and amphotericin B, trimethoprim-sulphadiazine, and sodium stibogluconate have been tried with varied success<sup>[6,15 - 17]</sup>. All drugs were endospore-static rather than endosporicidal. Data on antimicrobial drug resistance in *Rhinosporidium seeberi* is lacking. The strains obtained from human and animal rhinosporidiosis have shown genetic variations which might explain the variation of responses to some drugs<sup>[17]</sup>. The only drug appearing to have clinical promise is Dapsone<sup>[18]</sup>. It arrests the maturation of sporangia and promotes fibrosis in

the stroma, when used as an adjunct to surgery<sup>[2]</sup>. It could therefore be expected that presurgical Dapsone would minimize both the hemorrhage by its promotion of fibrosis, as well as preventing the colonization and infection of new sites after the release of endospores from the surgically traumatized polyps<sup>[17]</sup>. Laser and endoscopic excision promises to be the mainstream treatment of nasal/nasopharyngeal rhinosporidiosis in the future<sup>[6]</sup>. Our patient had complete excision with wide surgical margins and cautery of the base of the lesion and was treated subsequently with Dapsone.

### Conclusion

In conclusion, in a non-endemic area like Jaipur, India, rhinosporidiosis is uncommon, may pose a diagnostic challenge. However, with a significant proportion of the migrant population from the endemic states, it is likely that it will be observed more frequently in future. It is thus prudent for both clinicians and pathologists to keep this condition in mind when managing patients with nasal masses even from non-endemic areas. Moreover, it will be very crucial to follow in the next few years the clinical course of these patients to exclude the possibility of recurrence of the lesion, which usually occurs after an extended time period, to evaluate the best treatment for this infection. Nevertheless, rhinosporidiosis continues to be an enigma and a large number of further studies from endemic and non-endemic areas are needed. To the best of our knowledge, our case is one of the few nasal and sino-nasopharyngeal rhinosporidiosis reported from Jaipur.

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