# Management of Disseminated Rhinosporidiosis: Experience From a Single Tertiary Institution

Original Investigation

Kalaiarasi Raja[1], Saranya Thangavel[2], Akshat Kushwaha[1], Bheemanathi Hanuman Srinivas[3], Rakhee Kar[3], Arun Alexander[1], Lokesh Kumar Penubarthi[1], Sunil Kumar Saxena[1]

[1]Department of Otorhinolaryngology, Jawaharlal Institute of Post Graduate Medical Education and Research (JIPMER), Pondicherry, India
[2]Department of Otorhinolaryngology, Pondicherry Institute of Medical Sciences, Pondicherry, India
[3]Department of Pathology, Jawaharlal Institute of Post Graduate Medical Education and Research (JIPMER), Pondicherry, India

## Abstract

**Objective:** This study aims to present a series of patients with disseminated rhinosporidiosis with diagnostic and therapeutic features.

**Methods:** A retrospective study was conducted in a tertiary health care centre in South India from 2007 to 2020 with disseminated rhinosporidiosis. Twelve patients with multiple sites of involvement like the nose, nasopharynx, oropharynx, larynx, lacrimal sac and skin were included in the study. All patients underwent surgical excision, followed by peroral dapsone for one year.

**Results:** The age group was around 30-55 years, with male predominance (11:1). Pond bathing history was present in 50% (n=6). The most common site of lesion was the nose (100%), oropharynx (83.3%), skin (75%), larynx (50%) and less commonly, nasopharynx (41.6%) and lacrimal sac (25%). One patient underwent surgery four times (8.3%), followed by thrice and twice by five (41.6%) and six (50%) patients, respectively. On two years of follow-up, two patients (16.6%) had a recurrence in the nose and larynx whereas eight patients (66.6%) had no recurrence and two patients (16.6%) were lost to follow-up.

**Conclusion:** This original article highlights the rare occurrence of disseminated rhinosporidiosis, the possibility of which should be kept in mind, mainly when two or more sites are involved. The most significant number of disseminated rhinosporidiosis cases in the literature is reported here. Dissemination with the cutaneous and multisite disease is rarely reported and poses difficulty in management. Early diagnosis and intervention prevent the dissemination of spores into various parts of the body.

**Keywords:** Rhinosporidiosis, disseminated infections, cutaneous manifestations, nasopharyngeal diseases, disease management, tertiary care centers, otolaryngology

***ORCID IDs of the authors:***
*K.R. 0000-0002-0378-4141;*
*S.T. 0000-0001-6954-1364;*
*A.K. 0000-0003-1693-9330;*
*B.H.S. 0000-0002-9619-6719;*
*R.K. 0000-0001-6041-1512;*
*A.A. 0000-0003-1026-4678;*
*L.K.P. 0000-0003-1007-7776;*
*S.K.S. 0000-0003-1119-6072.*

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**Corresponding Author:**
Saranya Thangavel;
softsaran.nrp@gmail.com

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## Introduction

Rhinosporidiosis is one of the differential diagnoses for nasal mass with epistaxis. It is a chronic granulomatous disease that is endemic in South India. Malbran first identified the causative organism, but Guellermo Seeber described its structure and named it *Rhinosporidium seeberi* (1).

It mainly involves mucosal surfaces of the nose, nasopharynx, and oropharynx (70–75%), followed by the eye (15%), skin, or disseminated areas. The floor of the nose and inferior turbinate are the most common sites of involvement; the lesions may appear elsewhere too. Three or more sites involved in the disease is labeled as disseminated rhinosporidiosis. The organism enters the body via traumatized epithelium followed by the formation of microcysts and its local replication. This is associated with localized immune response and host cell hyperplasia. No host immunity against the organism has been noted. Dissemination to other sites may be due to autoinoculation, hematogeneous or lymphatic spread (2). Satellite lesions are seen adjacent to the lesions because of autoinoculation. The diagnosis is based on strong clinical suspicion whereas histopathology confirms the diagnosis. This retrospective study highlighted the largest number of disseminated rhinosporidiosis cases reported in the literature. Additionally, it described cases of dissemination involving cutaneous and multisite disease, and detailed how we managed patients with disseminated rhinosporidiosis.

## Methods

From retrospective data collection, 127 patients had isolated rhinosporidial lesions. Being a tertiary hospital, disseminated cases were referred to our center. This was a retrospective study of twelve patients treated for disseminated rhinosporidiosis at our institution from 2007 to 2020. The age groups of patients in the study were 30–55 years and predominantly males (male: female 11:1). Informed consent and ethical committee approval were obtained. The patients were presented with varied symptoms (Table 1). On examination, almost all patients (n=100%) noted a pink polypoidal lesion in the nasal cavity and oropharynx (Figure 1). Diagnostic nasal endoscopy was done in all patients showing a pinkish polypoidal, friable mass with yellow spots that bled on touch. One patient (n=8.3%) had left medial canthal swelling associated with epiphora (Figure 2). The visual acuity and extraocular movements were normal. There was a regurgitation of fluid from the puncta after giving pressure over the swelling, indicating lacrimal sac disease. Around six patients (n=50%) presented with either hoarseness or breathing difficulty. The larynx showed polypoidal lesions on direct laryngoscopy involving true cords at the anterior commissure without compromising the airway (Figure 3). Cutaneous lesions (n=75%) showed warty papules or nodules with crusting, bleeding, and whitish spots on the surface (Figure 4). One of the patients presented with nasal obstruction, multiple cutaneous papules over the face, chest, abdomen, and back and calf swelling (Figure 5). Blood investigations were normal, and serology reports (HIV, HbsAg and HCV) were negative. All patients underwent surgical excision and electro-desiccation of the base of the lesion under general anesthesia. The patients with airway involvement were managed using spontaneous anesthetic techniques and surgical procedures. In patients with laryngo-tracheal lesions, excision with microlaryngeal surgery (MLS) was performed. Histopathological examination showed numerous sporocysts in various stages of maturation with surrounding vessels, suggestive of *Rhinosporidium seeberi* (Figure 6). Peroral dapsone (100 mg/day) was given for one year for all disseminated rhinosporidiosis patients after surgery. All patients were followed up for two years.

**Table 1.** Patient symptoms

| Symptoms | % of patients (n=12) |
|---|---|
| Nasal obstruction | 100 (12) |
| Snoring and mouth breathing | 91.6 (11) |
| Epistaxis | 100 (12) |
| Hoarseness or breathing difficulty | 50 (6) |
| Watering of eyes | 25 (3) |
| Skin lesions | 75 (9) |
| Eye swelling | 8.3 (1) |
| Calf swelling | 8.3 (1) |

## Results

This is a retrospective study of disseminated rhinosporidiosis from 2007 to 2020. Twelve cases were diagnosed with disseminated rhinosporidiosis over this period (Table 2). The findings and patient characteristics are summarized in Table 2. There were eleven male patients and one female patient. The history of taking baths in ponds was present in 50%. One patient underwent surgery four times (8.3%), followed by thrice and twice by five (41.6%) and six (50%) patients, respectively. On two years of follow-ups, two patients (16.6%) had a recurrence in the nose and larynx whereas eight patients (66.6%) had no recurrence and two patients (16.6%) were lost to follow-up.

## Discussion

Rhinosporidiosis was first described in Argentina (3). The causative organism, *Rhinosporidium seeberi*, is a cryptic microbe. Still, it belongs to the human pathogen, Mesomycetozoa, which includes Dermocystidium, Rosette agent, Ichthyophonus and Psorosperminum clade parasite, depending upon its phylogenetic analysis of the *18sRNA* gene (4). *Cyanobacterium Microcystis aeruginosa* had been reported as the causative agent for rhinosporidiosis (5).


**Figure 1.** Shows a pink polypoidal lesion in the nose and oropharynx

**Table 2.** Prentation and outcome of 12 cases with their presentation and outcome

| S. no | Age/sex | Sites of lesion | Pond bathing | Number of procedures | Outcome |
|---|---|---|---|---|---|
| 1 | 30/M | Nose, nasopharynx, oropharynx, larynx, skin | No | 3 | No recurrence |
| 2 | 43/M | Nose, nasopharynx, oropharynx, larynx | No | 4 | No recurrence |
| 3 | 64/M | Nose, oropharynx, lacrimal sac, skin | Yes | 2 | No recurrence |
| 4 | 45/F | Nose, oropharynx, larynx, trachea | Yes | 3 | Recurrence in larynx |
| 5 | 37/M | Nose, oropharynx, skin | Yes | 3 | Lost to follow-up |
| 6 | 47/M | Nose, oropharynx, lacrimal sac, skin | No | 3 | No recurrence |
| 7 | 39/M | Nose, nasopharynx, oropharynx, larynx, skin | No | 3 | No recurrence |
| 8 | 34/M | Nose, oropharynx, larynx, skin | No | 2 | No recurrence |
| 9 | 46/M | Nose, nasopharynx, oropharynx, skin | No | 2 | No recurrence |
| 10 | 39/M | Nose, oropharynx, larynx | Yes | 2 | Recurrence in nose |
| 11 | 53/M | Nose, lacrimal sac, skin | Yes | 2 | Lost to follow-up |
| 12 | 30/M | Nose, nasopharynx, skin, calf swelling | Yes | 2 | No recurrence |

M: Male, F: Female

Rhinosporidiosis is endemic in South India, Sri Lanka and Africa. The endemicity is high temperature and humidity, favouring the development of spores. It spreads through contaminated water and soil and enters the body via minor abrasions. Our patients had a history of pond bathing contaminated with spores (6). It most commonly affects men more than women. It enters the body as trophozoite (6–8 μm), develops into immature and mature sporangia, and is released as free electron-dense bodies.

Manifestations of the disease can be nasal, ocular, cutaneous, and disseminated (1,7). Autoinoculation, hematogeneous, and lymphatic spread are the routes of spread of spores. The nose and pharynx is the most common site of inoculation via minor abrasions (trans epithelial infection) over the mucosal surfaces (8). Kirkpatrick (9) published the first case of lacrimal sac rhinosporidiosis in 1912. Ocular rhinosporidiosis occurs because of the spread of infection through the nasolacrimal duct. Mishra et al. (3) reported a lacrimal sac rhinosporidiosis who underwent dacryocystorhinostomy followed by peroral dapsone 100 mg once a day for one year.

Out of 127 patients with rhinosporidiosis, 12 patients had disseminated disease at the time of presentation. Studies discussing disseminated cases were quite low. Our disseminated cases were mostly from endemic areas. The chance of recurrence increases proportionately with inadequate removal, bleeding, surgical techniques, and injury to nearby tissues. The dissemination could be because of surgical techniques used in the previous surgeries and endemic factors. Recurrence rates following surgical and medical therapy are quoted in the literature as 5–63%. This indicates the risk of dissemination of spores into the submucosa during surgery. The host factors and endemicity also contribute to dissemination and recurrence.

The clinical symptoms include nasal obstruction and epistaxis (nasal); epiphora and swelling (ocular); snoring, mouth breathing, and dysphagia (pharyngeal); stridor,

hemoptysis, and voice change (laryngeal). Cutaneous rhinosporidiosis results in satellite lesions, generalized cutaneous (hematogeneous) or primary cutaneous lesions (autoinoculation). All 11 patients (n=100%) had a nasal obstruction. Daharwal et al. (10) reported a rare case of laryngeal rhinosporidiosis who underwent MLS excision. Usage of $CO_2$ or KTP laser aids in the complete excision of the lesion by providing proper visualization with better clearance margins, causing minimal trauma, reducing intra-operative bleeding, decreasing the chance of recurrence due to less contamination with spores, eliminating direct contact with the lesion, and ensuring good postoperative voice quality (11).

Ali et al. (12) reported a case with nasopharyngeal rhinosporidiosis extending to the oropharynx, whose main complaint was something stuck in the throat like a foreign body. The patient underwent an excisional biopsy and was disease-free after surgery (12,13). Clinical examination reveals a pinkish polypoidal mass studded with yellow spots, with typical strawberry-like regions that are friable and bleeds on touch. Nasopharyngeal polyps often have a variegated appearance and are multi-lobed (12,13). Prasad et al. (14) reported a case of disseminated cutaneous rhinosporidiosis with nasal and pharyngeal lesions who underwent surgical excision and became disease free after one year of peroral dapsone use.

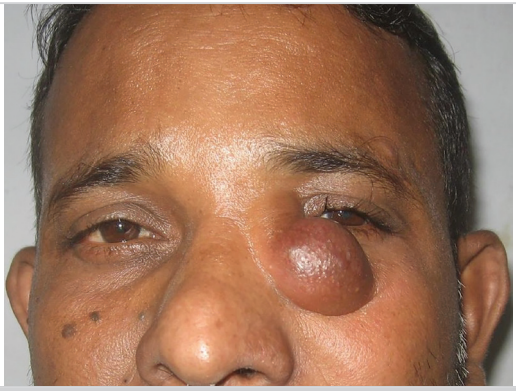

**Figure 2.** Shows left medial canthal swelling

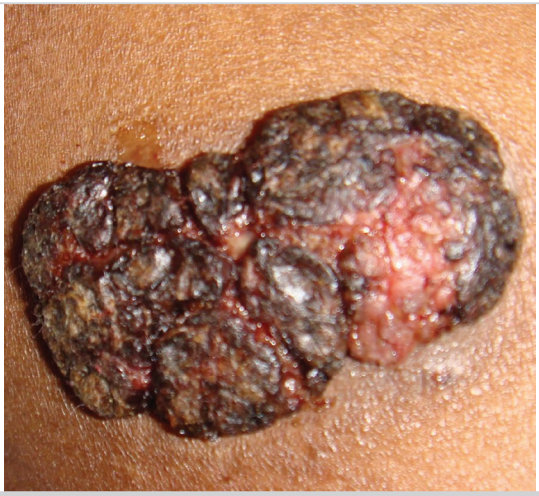

**Figure 3.** Shows cutaneous lesions

Computed tomography (CT) and magnetic resonance imaging have a limited role in the diagnosis but help in the preoperative extent of the disease and surgical excision planning. CT dacrocystography helps in identifying lacrimal sac involvement (15).

Histopathological examination confirms the diagnosis of rhinosporidiosis (16). It has characteristic features of numerous sporocysts at various stages of development and the stromal and cellular reaction of the host as well. The absence of Splendore–Hoeppli (antibody-mediated) eosinophilic deposit around rhinosporidial bodies differentiates it from other mycelial infections because these patients have high antibody titres (16).

The differential diagnoses include coccidiomycosis, warts and verrucous tuberculosis, pyogenic granuloma, hemangioma, condyloma acuminate, and lacrimal sac tumour or mucocele (1,13). The standard treatment is surgical excision of the lesion and electrocautery at the base. The laser can also be used to excise lesions with less chance of recurrence. Multiple site involvement requires single or multiple surgeries. The site involved first should be operated first because the epicentre will be at that site. Multiple site involvement requires surgery addressing the inferior site to the superior site to prevent contamination with spores to other sites and for better visualization without bleeding from the superior site. Localised disease usually will be cleared by surgery. But disseminated cases presents with recurrence more often and our patients also underwent multiple surgeries for disease clearance. Powered

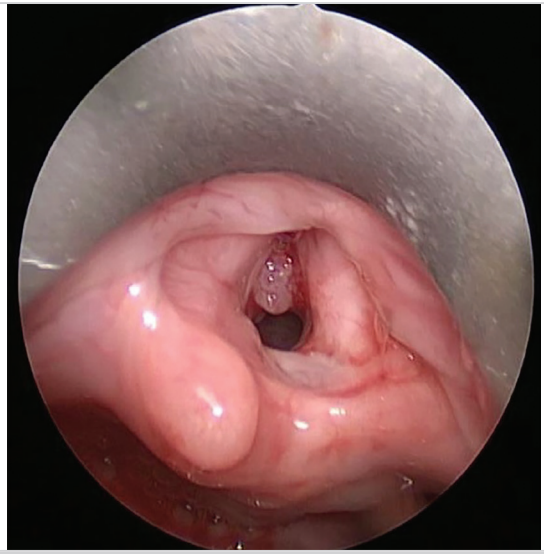

**Figure 4.** Shows a lesion at the anterior commissure on direct laryngoscopy

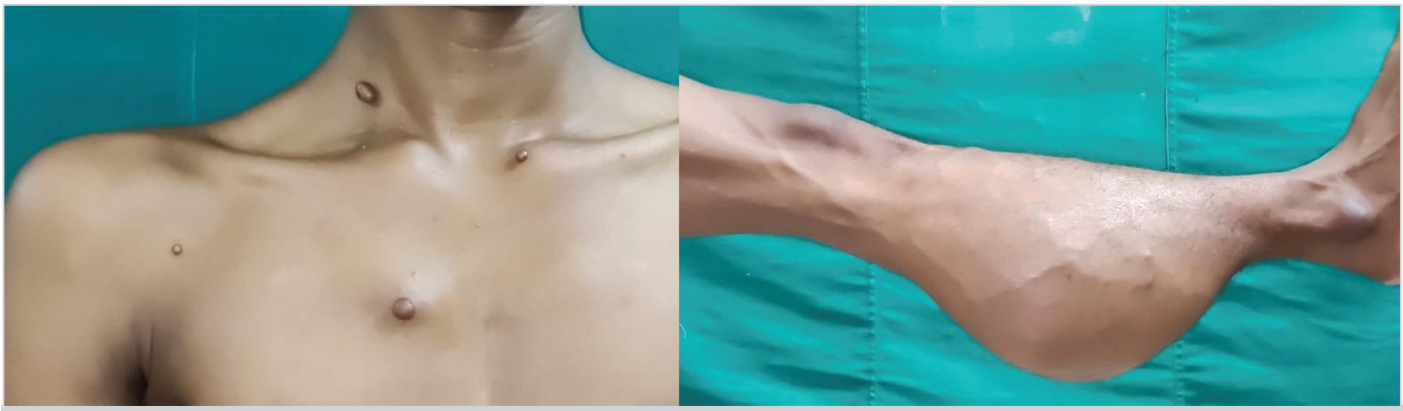

**Figure 5.** Shows multiple cutaneous lesions and calf swelling

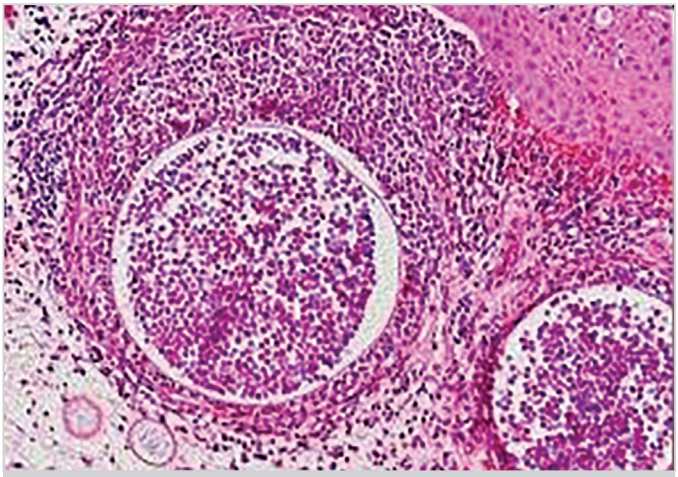

**Figure 6.** H&E stain of specimen (x400 magnification) showing hyperplastic epithelium with numerous sporangia in different stages of development and there is a surrounding dense, mixed inflammatory infiltrates

H&E: Hematoxylin and eosin

instruments like coblation and harmonic scalpel are useful in preventing dissemination of spores during surgery and thereby recurrence. These instrument help in the complete removal and keep the surrounding tissues intact without any injury (17).

Spontaneous regression of the lesions is rare. Since the organism could not be propagated *in vitro*, the sensitivity of testing of the drugs is not possible (2). Anti-fungal drugs like amphotericin B, ketoconazole, and dapsone and antibiotics like ciprofloxacin were tried, but dapsone is proven more effective. Cycloserine, an anti-tubercular drug can also be used. Dapsone (diaminodiphenyl sulfone) appears to arrest the maturation of spores and induce stromal fibrosis by accelerating degenerative responses (18). Our patients also responded well to peroral dapsone, which was given for one year. Eight patients (66.6%) had no recurrences on two years follow-up, whereas two patients (16.6%) had a recurrence in the nose and larynx. None of the patients had been tried only with medical therapy. All of our patients either localised or disseminated underwent surgery. But disseminated cases only were given peroral dapsone for one year. Localised cases were kept under regular follow-up and recurrence rates were less. Long-course usage of dapsone may be helpful in disseminated rhinosporidiosis. But prolonged usage of dapsone results in methemoglobinemia and hemolytic anaemia. These side effects are most commonly seen in patients with glucose-6-phosphate dehydrogenase deficiency (18). But none of our patients ever experienced these complications. Multi-drug therapy can be used in disseminated rhinosporidiosis. This will help in reducing the size of visceral and subcutaneous lesions and the disappearance of friable lesions. The disease's recurrence rate is high because of incomplete removal, reinfection and lack of oral medications. Disseminated cases requires multiple revision surgeries and regular follow-up (17,18).

## Conclusion

This original article described how the disseminated rhinosporidiosis cases were managed successfully without affecting the quality of life-avoiding a bath in stagnant water or ponds, proper hygiene, and early diagnosis and treatment to help prevent dissemination or autoinoculation of spores.

**Informed Consent:** Obtained from the patients for participation.

### Authorship Contributions

Surgical and Medical Practices: K.R., S.T., A.A., L.K.P., S.K.S., Concept: K.R., Design: K.R., S.T., A.A., L.K.P., S.K.S., Data Collection and/or Processing: K.R., S.T., A.K., B.H.S., R.K., Analysis and/or Interpretation: S.T., A.A., L.K.P., S.K.S., Literature Search: S.T., Writing: S.T., A.K., R.K.

**Conflict of Interest:** The authors have no conflict of interest to declare.

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**Main Points**

- Disseminated rhinosporidiosis is a mucocutaneous chronic granulomatous disease.
- The causative organism is an aquatic protistan parasite, *Rhinosporidium seeberi*, that belongs to the class Mesomycetozoa.
- Stagnant water contaminated with spores is the source of infection.
- Dissemination occurs through autoinoculation, hematogenous or lymphatic spread.
- This organism could not be grown in culture media. So, antimicrobial or anti-fungal therapy is ineffective.
- Dissemination with the cutaneous and multisite involvement is rarely reported and poses difficulty in management because it requires multiple surgeries, long term medical therapy and regular follow-up.

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