

# Rhinosporidiosis: Analysis Of Cases Presenting To A Tertiary Care Hospital In Nepal

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## Abstract

**BACKGROUND:** Described first by Seeberi in 1900, Rhinosporidiosis is caused by *Rhinosporidium seeberi*. It involves nasal mucosa, and is seen in both humans and animals. It is a waterborne disease endemic to the Indian subcontinent. It is also common in Nepal in the Terai belt, but a review of the literature did not reveal any study with respect to the clinicopathological profile of Rhinosporidiosis. **OBJECTIVES:** To study the clinicopathological profile of Rhinosporidiosis in eastern Nepal. **METHODOLOGY:** The study included 84 cases of Rhinosporidiosis over a period of 4 years. All were diagnosed on a clinical basis. All subjects were treated surgically by wide excision and electrocautery, and the specimens were sent for histopathological examination. Dapsone was given to all patients with recurrence. Subjects were followed up for complications, outcome and recurrence. **OBSERVATION:** The study was carried out over a period of 4 years with patients being between 7 and 54 years of age. The disease has a male preponderance with an average duration of symptoms of 4 years. All patients had a history of bathing in rivers or ponds that were also used by their cattle. 19 cases were recurrent. **CONCLUSION:** The study reflects the endemicity of this disease in eastern Nepal and related border areas adjoining India. It is rare in the hilly region of Nepal. High incidence is particularly noticed in those bathing in ponds accessed by cattle, and raising public awareness regarding this disease would go a long way in decreasing its incidence.

## INTRODUCTION

Rhinosporidiosis has been known for over a hundred years since its first description in Argentina. It is a chronic disease, with frequent recurrence after surgery. It is endemic to south Asia, notably southern India and Sri Lanka<sup>1</sup>. Sporadic cases are reported from North, Central and South America, South and East Africa, Japan, the Philippine Islands, Malay States, Canada, Uganda and Iran. Increased migration of persons who acquired Rhinosporidiosis in their native Asian countries has resulted in the increasing incidence of this disease in non-endemic areas. Rhinosporidiosis is an infective disease as tissue lesions are always associated with the presence of the pathogen. No evidence has ever been documented of cross-infection between members of the same family or between animals and humans. The great majority of cases are sporadic, but predisposing factors in the host may be a cause of its endemicity. There is some data in India showing a higher incidence of Rhinosporidiosis in subjects with blood group O<sup>2</sup>. The probable mode of infection from the natural aquatic habitat of *Rhinosporidium seeberi* is through traumatized epithelium, most commonly in the nasal cavity<sup>3</sup>. Autoinoculation is known to cause satellite lesions adjacent to granulomas especially in the

upper respiratory sites<sup>4</sup>. Spillage of endospores from polyps after trauma or surgery is thought to be followed by autoinoculation through the adjacent epithelium. There is evidence for hematogenous spread of rhinosporidiosis to anatomically distant sites, as evidenced by the development of subcutaneous granulomas in the limbs without breach of the overlying skin<sup>5</sup>. The mode of regional spread, however, is controversial. Ashworth suggested the possibility of lymphatic spread<sup>6</sup>. Dissemination to the limbs, trunk and viscera has been described in a few cases, with a rare fatality especially when the brain was involved. In rhinosporidial lesions of the limbs, a notable feature is destruction of underlying bone. Rare cases of spontaneous regression have also been recorded.

Characteristically, rhinosporidial lesions in the nasal cavity are polypoidal, granular, and red in colour due to pronounced vascularity. The surface contains yellowish pinhead-sized spots representing underlying mature sporangia. A covering of mucoid secretions is not uncommon. Nasopharyngeal lesions are often multi-lobed with variegated appearance, typical strawberry-like regions and areas that have relatively less vascular lobes with

smooth surfaces. Lesions on the face and trunk are either pedunculated, or sessile on broad bases simulating verrucous warts. Rhinosporidial granulomas clinically present as ulcerated growths mimicking malignant lesions such as sarcomas and carcinomas.

A majority of cases occur in upper respiratory sites, notably the anterior nasal cavity, the inferior turbinate, septum and floor. Rhinosporidial lesions also occur in the soft palate, nasopharynx and larynx, and rarely the buccal mucosa. The disease, while being of special interest to an otorhinolaryngologist, is of interest to dermatologists and ophthalmologists as well, through the occurrence of granulomas in the skin, subcutaneous tissues and eyes. About 15% of cases of rhinosporidiosis affect the bulbar and palpebral conjunctiva. Rhinosporidiosis of the lacrimal sac and nasolacrimal duct has also been documented<sup>7</sup>.

The definitive diagnosis of rhinosporidiosis is by histopathology, with the identification of the pathogen in its diverse stages. Although all the developmental stages of the pathogen has definitive characteristics which allow histopathological diagnosis, some notable features include the wide variation within a single specimen, in lesions of different sites, in respect to both endospores and sporangia, as well as the cellular response of the host.

The etiologic agent *Rhinosporidium seeberi*, initially described as a protozoan by Seeberi, started to be classified as a fungus by Ashworth. However, several enigmas still persist even today regarding *R. seeberi*, due to the failure of attempts at sustained in vitro culture, and the failure to establish rhinosporidiosis in experimental animals. Though molecular biological analysis of the organism's ribosomal DNA suggests it to be a cyanobacterium, like *Microcystitis aeruginosa*<sup>8</sup>, some consider it a mesomycetozoon<sup>9</sup>.

Associated diseases are incidental, but schistomiasis, syphilis, leprosy and malignancy are reported. Rhinosporidiosis is rarely fatal. Hemorrhage and generalized dissemination may, however, lead to death<sup>10</sup>.

Several trials have shown that the most successful treatment, with the lowest incidence of recurrence, is surgical extirpation and cauterization of the base of the lesions. Unfortunately, reports regarding recurrence rates are very few. Satyanarayana et al. reported a recurrence rate of 11% after surgical treatment; Khan et al. followed with 22 patients of whom 18 were treated by cutting diathermy. The latter patients had no recurrences, whereas of the four cases

in which the lesions were removed with forceps and snares, all had recurrent disease. Conservative treatment as a single modality is not widely used, but in one clinical trial by Nair et al. in 1979, applying diaminophenylsulfone to 32 patients with nasopharyngeal rhinosporidiosis, 71.4% patients did not show recurrence in a three-year period and none of them needed additional surgery during that period.

## **AIMS AND OBJECTIVES**

To study the clinicopathological profile of Rhinosporidiosis in Eastern Nepal

## **MATERIALS AND METHODS**

It was a prospective study carried out from 2006-2010. This series included 84 subjects with a clinical and histopathological diagnosis of rhinosporidiosis treated at the B P Koirala Institute of Medical Sciences, Dharan, Nepal.

A detailed history, including age, sex, duration of symptoms, personal habits, and area of residence was recorded.

Particular focus was placed on the bathing habits and work profile of the subject. Clinical examination and diagnostic nasal endoscopy were done to localize the site and extent of lesions in each case. All cases were provisionally diagnosed on a clinical basis, and preoperative biopsy was not advised in any of the case. History of other medical or surgical illness was also recorded. All routine hematological investigations, including blood group, of each patient were also done. Clinical diagnosis of rhinosporidiosis was based on the following:

All subjects were treated endoscopically by wide surgical excision and electrocautery of the base of lesion under general anesthesia. Extensive posterior nasal attachment was also excised endoscopically. Intraoperatively, the extent and attachment of lesions was noted for every case. Average blood loss was also noted and a specimen was sent for histopathological examination. In extensive and recurrent cases, Dapsone 100mg daily for 3 months was given. Origin from more than one site was considered extensive. Subjects were followed up endoscopically at 2 weeks, monthly for the next 6 months, every 2 months up to 1 year and thereafter every 3 months for a year for signs of recurrence and outcome.

## **OBSERVATION & RESULTS**

A total of 84 cases were treated over a period of 3 years (January 2007- January 2010). There were 53 males (63%) and 31 females (37%) in the study. Patients ranged from 7 years to 54 years of age, being predominantly in the 2<sup>nd</sup> and

3<sup>rd</sup> decade of life (Table 1).

**Figure 1**

TABLE 1: Age and sex distribution of Rhinosporidiosis

AGE (in years)	MALE	FEMALE	TOTAL No ( %)
0-10	4	2	06 (7.1%)
11-20	13	8	21(25%)
21-30	21	15	36(42.8%)
31-40	8	3	11(13%)
41-50	5	3	08(9.5%)
51-60	2	0	02(2.3%)

All patients were of Indo-Aryan race belonging to the border area adjoining India (Table 2).

**Figure 2**

TABLE 2: Geographic distribution of Rhinosporidiosis

Area	Number of cases (%)
Saptari District	36 (42.8%)
Siraha District	31 (37.0%)
Dhanusha District	17 (20.2%)
Total	84 (100%)

The average duration of symptoms was 4 years (range 1 month to 10 years). All patients gave a definitive history of bathing in ponds or rivers and also had contact with cattle or were cattle grazers. Nasal obstruction (54%) was the most common symptom followed by epistaxis (40%) (Table 3).

**Figure 3**

TABLE 3: Symptomatology of subjects

SYMPTOM	INCIDENCE
Nasal Obstruction	54%
Epistaxis	40%
Dysphagia and foreign body sensation	06%

Lateral wall of nose (39%) (Figure 1), was the most common site of origin and nasopharynx (4%) (Figure 2), was the least common site of origin (Table 4).

**Figure 4**

FIGURE 1: RHINOSPORODIOSIS ARISING FROM RIGHT LATERAL NASAL WALL.



**Figure 5**

FIGURE 2- Rhinosporidiosis of Nasopharynx presenting orally.



**Figure 6**

TABLE 4: Sites of involvement

SITE	INCIDENCE
Lateral nasal wall	39%
Nasal septum	33%
Floor of nasal cavity	24%

Rhinosporidiosis involved both nasal cavities in 4% of the cases and multiple sites were involved in 23% of the cases.

14 patients had a previous history of surgery, all presenting with multiple attachments when reviewed at our center. The previous extent of disease could not be ascertained in any of these 14 cases. 43% of the subjects had O positive blood type whereas subjects with blood group A positive, B positive and AB positive made up 25%, 18% and 14% of our subjects, respectively (Table 5).

### Figure 7

TABLE 5: Blood group distribution of patients

BLOOD GROUP	Number (%)
O positive	36 (43%)
A positive	21 (25%)
B positive	15 (18%)
AB positive	12 (14%)

Endoscope-assisted wide excision was done in all cases followed by electrocautery of the base. 83 patients had uneventful recoveries while in 1 case with extensive attachment, severe intraoperative hemorrhage was encountered which could only be controlled with external carotid artery ligation. All 84 specimens were reported histologically as rhinosporidiosis. Recurrence was seen in 19 cases (23%). 15 cases with multiple attachment and 4 cases with single attachment showed recurrence, and the average period of recurrence was within 1 year. All cases involving recurrence had extensive disease with multiple attachments mostly in the lateral wall of nose. Recurrence was treated with revision endoscope-assisted excision and Dapsone therapy. Subjects were treated with 100mg Dapsone daily for three months. None reported with any recurrence on subsequent follow-ups.

## DISCUSSION

Rhinosporidiosis is an illness of low incidence, limited to the endemic zones in tropical and subtropical regions of India and Sri Lanka. Literature review revealed little about the possible habitat and mechanism of transmission of this pathogen. The most accepted hypotheses are still possibilities, pointing to a relation between contagion and patients who had contact with backwater. Some cases are described in animals such as horses, fish and cattle in general. However, animal-to-man transmission has not been proven. In our series, all subjects had a history of exposure to backwater and/or contact with animals. It is suggested that the contact of the nasal mucosa with contaminated water inoculated through digital microtraumas is a possible method of transmission that explains the predominance of lesions in the nasal cavities and eyes. The populations of low economic status are more frequently affected, with the majority of

patients coming from agricultural zones, and those taking baths in ponds.

Cases may occur between the ages of 3 to 73 years with a maximum incidence between the ages of 20 and 30 years<sup>11</sup>. One author reported a maximum incidence between 11-20 years. Rhinosporidiosis is more common in males than females (3:1). Although the sex distribution varies in different reports<sup>12</sup>, this may be due to the fact that males more typically have jobs related to land and backwater. We did observe a male predominance in relation to the sexes (1.7:1). Most of the cases observed in our series were between 10 and 40 years of age. The literature suggests susceptibility among races is not known. Our evidence suggests affection in the Indo-Aryan race; we however believe that this predominance is due to the population of this race in the eastern agricultural zone of Nepal. In order to determine the relation of any possible host factor, the blood group related data of all the subjects was analyzed. The distribution of O<sup>+</sup>, A<sup>+</sup>, B<sup>+</sup> and AB<sup>+</sup> was found to be 43%, 25%, 18% and 14%, respectively. There were no subjects with a rhesus negative group, and nearly half of the patients were of the O<sup>+</sup> blood group, leading us to the conclusion that Rhinosporidiosis is more common in subjects with rhesus positivity, even more so in group O. Further research regarding the same would possibly bring forth interesting results.

Soft and friable nasal polyps and mucosa can be found upon examination. Grey or yellow spots, which represent the bulging sporangia through the attenuated epithelium, give a characteristic strawberry appearance. We observed a similar pattern in appearance of the lesions in our series and found it to be an important diagnostic criterion as far as rhinosporidiosis is concerned.

The most frequent site is the nasal cavity although it is found in other localizations such as the conjunctivae, oral cavity, lacrimal sac, urethra, paranasal sinuses, larynx, skin, and bone. It can affect more than one site<sup>13</sup>. The most common nasal sites are, in decreasing order, nasal septal mucosa, inferior turbinate and nasal floor. Our findings suggest the lateral nasal wall (39%) as the most common site of involvement. Other sites in decreasing order were the nasal septum (33%), the floor of the nasal cavity (24%) and the nasopharynx (4%). 4% cases had bilateral involvement, and 23% cases had multiple sites of attachment in the nasal cavity. Our series included 14 subjects with previous history

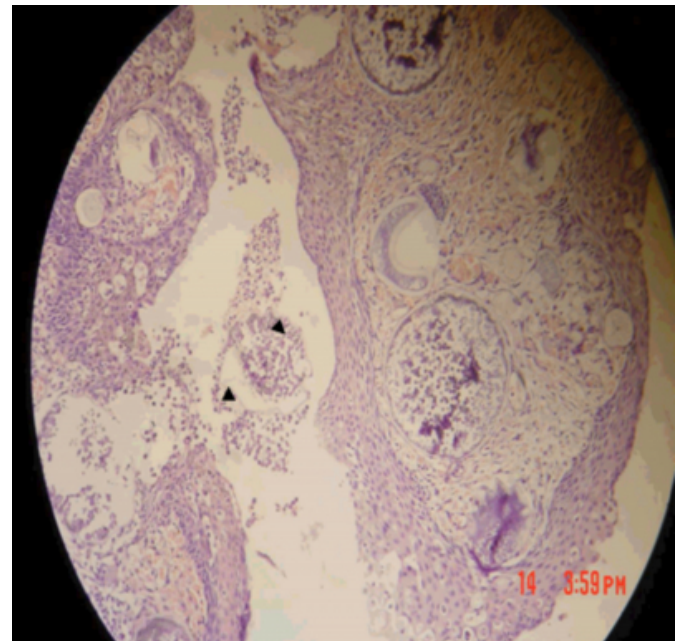
of surgery presenting to our center with recurrences. Out of the 14, 1 subject (7%) had bilateral involvement and 11 (79%) had multiple attachments, which led us to conclude that recurrent disease was mostly extensive and difficult to treat.

The progression of symptoms is slow and the presenting complaints are nasal blockage, epistaxis, nasal discharge, nasal mass, and sensation of a foreign body. Clinically it presents as a polypoid mass which is friable, painless, sessile or peduncle and finely lobated with small yellow points over the mass (strawberry aspect), corresponding to mature sporangia. It bleeds on touch. Our findings revealed a similar pattern with nasal obstruction (54%) being the most common presenting symptom followed by epistaxis (40%), dysphagia and foreign body sensation in throat (6%) being the least common. Rhinoscleroma, among others, is an important differential diagnostic condition to be considered. The diagnosis is based on clinical history, with important epidemiologic approach, detailed otorhinolaryngological examination and histopathological confirmation.

The histological diagnosis is on the basis of the presence of the sporangia in different periods of maturation. Stroma shows a chronic inflammatory reaction with predominance of plasmatic cells, lymphocytes and scarce neutrophils. They differ from the allergic polyps in the lesser amount of eosinophils. Some of these sporangia can be ragged, and spores are observed in the exudates of stroma. These ragged sporangia can provoke granulomatous reactions (Figure 3).

**Figure 8**

**FIGURE 3: HISTOPATHOLOGICAL SLIDE SHOWING SPORANGIA (Arrow Heads) IN A CASE OF RHINOSPORODIOSIS**



Treatment of the disease is far from satisfactory, as recurrence is the rule rather than the exception. Our study showed an overall recurrence rate of 22.6% and about 4% had multiple recurrences. Bleeding during surgery is profuse and can be life threatening. To date, the treatment of choice is wide local excision and cauterization of the base. The advent of the endoscope has facilitated the removal of small granulomas, but bleeding from large granulomas is cumbersome and poses difficulties during endoscopic surgery. The advent of KTP and Nd-YAG laser has also helped in reducing blood loss during surgery as well as recurrences<sup>14</sup>. Hemorrhage is the most common complication. It would be pertinent to mention here that in our study, endoscope assisted excision was done for all cases; the average blood loss was about 100ml except in 1 case where severe hemorrhage was encountered which ceased after external carotid artery ligation.

Medical treatment is described in the literature; however it is without useful results when used as the only modality of treatment. Diaminodiphenyl sulphone (Dapsone) has been used by some authors as an adjuvant to the surgical treatment to reduce the recurrence of disease. It appears to arrest the maturation of the sporangia and promotes fibrosis in the stroma when used as an adjunct to surgery. We used Dapsone in the dose of 100mg orally per day for 3 months for all 19 patients who were found to have recurrence on



follow up. Most of the recurrences were within 6 months (73%) and the rest were within a year. All patients were screened with monthly hemogram and liver function tests for any side effects of Dapsone. None of the patients reported any side effects of Dapsone. Post-Dapsone therapy, 16 patients have been followed up for more than a year but have not reported any recurrence, 2 patients were lost to follow up after 4 months and 1 after 6 weeks.

Literature review suggests a residual or recurrence rate between 10 and 70%. However, most of the reports show an incidence on average of 10%, related to incomplete excision of the mass. In the present study we observed recurrence in 22% of cases. Extensive disease was an independent risk factor for recurrence in our study. A follow up of at least 6 months should be the protocol to rule out recurrences.

## **CONCLUSION**

Rhinosporidiosis is disease of dubious etiology affecting mainly farmers and is common in tropical regions of the world. Wide local excision with cauterization of base is the main modality of treatment, with Dapsone as an adjuvant therapy in cases of recurrence and extensive diseases. Recurrence is the rule rather than the exception. Education of the population at risk should involve appropriate public health strategies and avoidance of the unhealthy practice of bathing in ponds and rivers open to animals.

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