Review Article

# Rhinosporidiosis: Bizarre Clinical presentation and Mode of Dissemination

**Goswami R**

Dr Reema Goswami, Assistant Professor, Department of E.N.T, Bundelkhand Medical College, Sagar, Madhya Pradesh, India

**Address for Correspondence**: Dr Reema Goswami, Email: reemadevgoswami@ gmail.com.

## Abstract

Rhinosporidiosis is a benign chronic inflammatory infection which manifests as a granulomatous polyp predominantly in the nose and nasopharynx. The extranasal sites of involvement include the lacrimal sac, tongue, palate, larynx, external genitalia, skin and bones. Few cases of disseminated rhinosporidiosis have been reported. Literature regarding various bizarre presentations and the mode of dissemination from primary site to non-mucosal sites has been reviewed.

**Keywords:** Rhinosporidiosis, Granulomatous polyp, Nose, Extranasal sites, Cutaneous lesions, Osteolytic lesions.

## Introduction

Rhinosporidiosis is a chronic granulomatous disease which manifests as a strawberry like polyp predominantly in the nose and nasopharynx. This appearance results from sporangia which are visible as gray or yellow spots in the vascular polypoidal mass which bleeds on touch.

Rhinosporidiosis occurs predominantly in tropical and subtropical regions although a few sporadic cases have been reported almost universally. India and Sri lanka have the highest prevalence but Argentina, Brazil, Texas and Uganda are also regarded as endemic focus. In India the highly endemic belt includes Chhattisgarh, Orissa and costal rice producing states of Tamilnadu, Kerala and Karnataka.[1, 2]

In this review various non-mucosal presentations of Rhinosporidosis will be discussed. The mode of transmission from the primary site to non-mucosal sites and factors that make visceral involvement noncondusive to growth are analyzed.

## Etiological Agent

On reviewing the historical aspects, Malbrun of Buenos Aires in 1892 first described what he called infection with sporozoal parasite from a specimen of nasal polypus. He however did not publish his findings.

In 1900 Guillermo Seeber of Argentina described the etiological agent in a nasal polyp. Wernicke in 1903 named the organism Rhinosporidium seeberi.[3] Ashworth in 1923 gave a detailed account of the pathogen's life cycle and concluded that Rhinospriodium seeberi was a lower aquatic fungus belonging to the class phycomycetes.[4] The earliest stage of the organism was termed as trophi by Ashworth was a round cell of 6-7 micron and is the infective stage of the fungus. Infective spores after implantation in the mucosa or subcutaneous lead a parasitic life. It increases by asexual multiplication and develops into a sporangium, containing as many as 16000 spores. The mature sporangia bursts at a weak point known as pore and spores are released in the tissue to commence the life cycle again.

Rhinosporidiosis seeberi has never been successfully propagated in vitro. A recent breakthrough in to its phylogentic affinities using 18S rRNA gene analysis has revealed it is closer to a fish parasite in the kingdom protoctista and is probably an aquatic protistan parasite.[5]

## Source of infection

History of pond bath is recorded by most authors reporting series of Rhinosporidiosis cases.[6] It is possible that spores escape with nasal discharge into the ponds when infected animals or persons bathe in them. These spores may infect some vulnerable persons who swim in water. Swimming ensures prolonged contact with spores against nasal mucosa and helps in inoculation of spores into these sites. The spores protected by their chitinous wall remaining in the stagnant pond water, unlike river water where even if contaminated has least chance of spreading of infection, as water flows continuously.

Manuscript received: 24th Dec 2013
Reviewed: 29th Dec 2013
Author Corrected: 15th Jan 2014
Accepted for Publication: 29th Jan 2014

Karunaratne in 1964 pointed out that the disease does not seem to have any relation with paddy crop but to the soil and muddy water to which the cultivators are brought to intimate contact during paddy cultivation. It is a common practice for people to use this water for washing and bathing.[7]

In a detailed epidemiological investigation by Z Vokovic into the first outbreak of Rhinosporidiosis in Europe in 1992-1993, 17 patients suffering had a preceding history of swimming in silver lake.[8]

The disease is not contagious as prolonged contact with an infected person does not seem to cause disease in any other family member.

Though some authors have reported more than one member of the family suffering from the same disease, it may be due to similar life style viz pond bath and paddy cultivation.[7,9]

In the ICMR epidemiological survey in 1971-72, Gupta and Billore recorded history of contact with domestic animals in 19.04 % cases only.

Direct animal to man transmission may be ruled out as all attempts of animal inoculation with spores have been unsuccessful. Spores may escape into stagnant water and infect man.[6]

It appears that spores do not enter the intact epithelium and a breach in the continuity of the epithelium is necessary to facilitate infection. This is the reason why all individuals exposed to the same environment are not infected. Some authors have reported nose picking and history of trauma in their case series.[7]

Habits like nose picking and scratching may go unnoticed by patients, though may cause microabrasions to facilitate inoculation of the parasite.

## Bizarre presentations of Rhinosporidiosis

Rhinosporidiosis causes infection of the nose and nasopharynx in 70% cases and ocular infection in 15% cases. [2, 3] Beattie 1907 first reported a bizarre presentation of rhinosporidiosis as an aural polyp.[4]

### Cutaneous lesions

Cutaneous lesions are classified into 3 types:

1. Satellite lesion with primary in the nasal cavity and involvement of adjacent skin.
2. Generalized cutaneous lesions with or without nasal involvement.
3. Independent lesions.

Forsyth (1923 and 1924) placed on record the first case of skin infection of rhinosporidiosis.[10] Allen and Dave (1936) recorded 2 cases of satellite lesions of skin and 2 cases of generalized cutaneous lesions with nasal involvement[11] Dhayagude (1941) reported a rare and unusual case of generalized rhinosporidial granulomata in a 40 year old male.

Subcutaneous swellings were noted on the face, abdomen, thorax and back.[12.] No primary lesion was discovered in the nose and nasopharynx.

Rajam et al. reported a case report with 33 subcutaneous and cutaneous lesions of rhinosporidiosis.[13]

### Osteolytic Lesions

Nguyen van Ai et al recorded a case of rhinosporidiosis in which there was destruction of the bone at the wrist and which was replaced by rhinosporidial growth.[14] In the left foot there was complete osteolysis of distal epiphysis of 3rd metatarsal.

Various others like Chatterjee PK, Dastidar et al. and Arvindan have reported cases of rhinosporidiosis of the bone.

The curetted material from the lesion showed granulation tissue and multiple sprorangia of Rhinosporidium seebri[15], [16,]

### Rhinosporidiosis of larynx and bronchi

Larynx, trachea and bronchi are rare sites of involvement and usually there is co existent infection of nose and nasopharynx though cases have been reported where the lower respiratory site is the sole site of infection.

Allen and Dave in 1936 reported a case of laryngeal rhinosporidiosis and were removed by laryngofissure approach.[11] Thomas et al. reported the first case of rhinosporidiosis in the bronchus.[17]

The Patient complained of cough with mucoid sputum and hemoptysis. Subramanayam and Ramana Rao believed that in every case of nasal and nasopharyngeal rhinosporidiosis, when symptoms of cough and dyspnea are out of proportions of nasal obstruction endoscopic examination of larynx, trachea and bronchi should be done.[18]

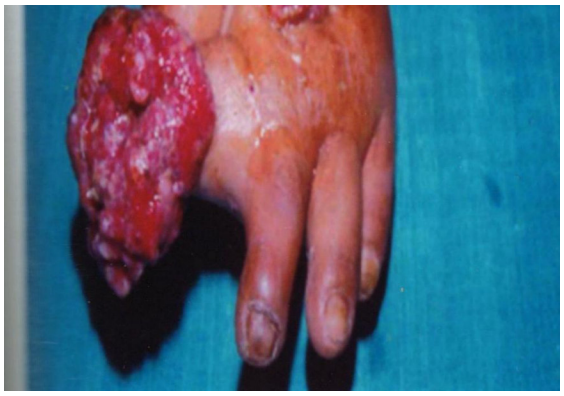

Fig 1: .Rhinosporidial mass from great toe, 2nd toe and dorsum of foot

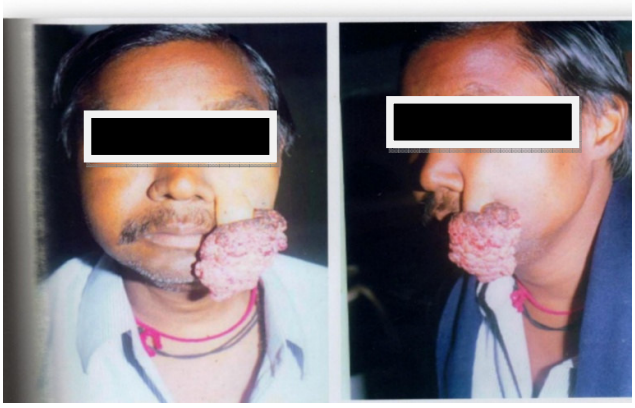

Fig 2: Cauliflower like rhinosporidial growth arising from skin of of cheek

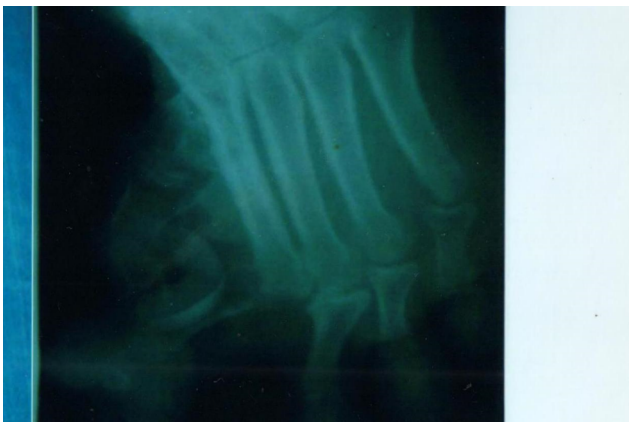

Fig 3: X-Ray of foot showing destruction and erosion of 1st metatarsal with displacement of proximal phalanx and soft tissue swelling

### Rhinosporidiosis of parotid gland.

The first case of Rhinosporidosis of the parotid duct was reported by Mahadevan in 1952 in a 20 year male. There was a painful cystic swelling over the parotid region and blood stained discharge in the mouth on pressing the swelling. Sialogram did not show sialectasis.[19] Other authors have also reported similar cases, diagnosis of Rhinosporidosis being confirmed by histopathological examination of excised mass.[20]

### Rhinosporidosis of external genitalia

Dhayagude in 1941 reported a case of uretheral rhinosporidiosis protruding from the external meatus while micturation, the regional lymph nodes were not enlarged.[12] Borezene et al.in 1951 reported the first case of rhinosporidiosis of the vulva in an Argentinean women.Karunaratne reported a case of vaginal Rhinosporidiosis who presented with sangious discharge which contained spores.[7]

### Systemic dissemination of Rhinosporidiosis

Rajam et al. in 1955 reported the first fatal of systemic dissemination of rhinosporidiosis in a 40year old male who presented with multiple cutaneous lesions and recurrent rhinosporidial infection in right nostril. According to Rajam the chain of events that lead to death was low grade rhinosporidial septicemia, hypochromic anemia, cirrhosis of liver recurrent ascites leading to death.[13] Ho and Tay 1986 reported finding of sporangia in the blood vessels.[21]

### Mode of dissemination

The preponderance of the lesion in the mucosal lining of nose and nasopharynx suggest direct implantation of spores on the mucosal epithelium but a breach in the epithelium seems to be a necessary condition for inoculation of the parasite.[7] This explains why all members of a particular family are not infected though all of them are exposed to the same environment.

Infection of contiguous sites may take place by permeation of spores along subepithelial connective tissue. Autoinoculation by fingers was regarded as a mode of dissemination to the skin by authors like Forsyth [10] but if this is the mode of dissemination the number of cases with cutaneous involvement should be more in children as nose pricking is a common habit in them.

The hypothesis of autoinoculation could be valid only for ulcerative lesion. Hence it is more likely the mode of dissemination to distant sites is by haematogenous route.

Dhayagude who reported a case of generalized cutaneous rhinosporidiosis also suggested a haematogenous route of dissemination, though he failed to demonstrate spores in blood. He reasoned out that culture examination becomes necessary to demonstrate mocroorganisms in all speticemic condition unless the infection is massive but there is no suitable method of culture of R. Seeberi.[12]

Chatterjee and Dastidar (1977) who reported rhinosporidiosisof the bones also suggested haemotogenous route of dissemination.[16] Cases with cutaneous & bone involvement have history of recurrent nasal infection with history of repeated surgeries for the same which may cause seeding of circulation with R.seeberi. Rajam et al. demonstrated R.seeberi in peripheral venous blood, urine and ascitic fluid is a fatal case of disseminated Rhinosporidiosis. Autopsy studies showed scattered spores in the liver, spleen, kidney, heart and brain but the being was spared.[13]

An unexplained feature of rhinosporidiosis is the extreme rarity of the involvement of the lung particularly in view of the frequency with which it occurs in upper respiratory passage. Rajam et al. suggested that the parasite does not thrive in vascular aerated lung tissue13.

Thus the factors that seem to favour localization of spores are low temperature low oxygen tension and loss of integrity of surface epithelium. The lung seems to be the least favored site with high oxygen tension.

## Treatment

Excision by diathermy and electro cautery of the base has been recommended by most authors. Good results obtained by diathermy were explained on the basis that as destruction of tissue is deep with diathermy, it avoids implantation of spores[22] The use of KTP laser is effective in localized small growth of Rhinosporidiosis.[23]

Therapy with Dapsone has been apparently successful on clinical trials of Dapsone on nasal and nasopharyngeal rhinosporidiosis by Nair (1979).[24] Woodard and Hudson 1984 suggested failure of the therapy is attributed to impenetrability of sporangial wall.[25] Herr et al. reported that patients with rhinosporidiosis possess anti R. seeberi IgG. to inner wall antigen expressed during mature sporangial stage The findings suggest that mapping of antigenic proteins may lead to important antigens with potential as vaccine candidates.[26]

## Conclusion

The haematogenous route of dissemination from the primary site to distant site is the only possible explanation for occurrence of disease in subcutaneous tissue, bone and other sites. Non-involvement of other tissue like lung and brain may be due to noncondusive biological environment for growth like high oxygen tension.

The distinct characteristics of nasal rhinosporidiosis make its diagnosis simple but extranasal involvement may be missed out because of lack of familiarity with such manifestations. In endemic areas the possibility of rhinosporidial infection should be borne in mind for patients presenting in bizarre ways.

**Funding**: Nil
**Conflict of interest**: Nil
**Permission from IRB**: Yes

## References

1. Darbari BS, Gupta RL, Shukla IM and Arora MM. Rhinosporidiosis in Raipur. A Clinico-pathological study of 348 cases. Indian Journal of Pathology and Bacteriology. 1972; 15(4): 103-107.

2. Iqbal Md, Dani SK. Clinicoepidomological profile of rhinosporidiosis. Indian Journal of Otorhinolargyngology and Head and Neck surgery. June1993; 45(2):69

3. Mukherjee B, Mohan A,Sumathi V, Biswas J. Infestation of lacrimal sac by Rhinosporidium seeberi:A clinopathological report. Indian Journal of Opthalmol 2013; 61:588-90.

4. ArseculeratneSN. Recent advances in Rhinosporidiosis and Rhinosporidium seeberi. Indian Journal of Medical Microbiology.2002; 20 (3):119-131.

5. HerrRA, AjelloL, TaylorJW, ArseculeratneSN. Phylogenetic analysis of Rhinosporidium seeberi's 18S small sununit ribosomal DNA group. This pathogen among members of protoctistan Mesomycetozoa clade.J of Clinical Microbial.1999; 37(9):2750-4.

6. GuptaRL, DarbariBS, BilloreOP. An epidemiological study of Rhinosporidiosis in and around Raipur. Indian Journal of Med Res1976;64-1293

7. KarunaratneWAE. The pathology of Rhinosporidiosis. The Journal of Pathology and Bacteriology.Jan1936; 42(1)193-202

8. VukovicZ, Bobic-RadovanovicA, LatkovicZ, RadovanovicZ. An epidemiological investigation of the first outbreak of Rhinosporidiosis in Europe. J of Tropical Med and Hygeine.1995; 98(5):333-7.

9. ChitravelV, SundaramBM, SubramanianS, KumaresanM, KunjithapathamS. Rhinosporidiosis in man: A case report. Mycopathologica1990;109:11-12

10. ForsythWL.Rhinosporidium Kinealyi. Lancet.1924;1: 951-2

11. Allen S, Dave ML. Treatment of Rhinosporidiosis in man based on study of cases. Indian Med Gaz.1936; 71:376-393.

12. Dhayagude RG. Unusual rhinosporidial infection in Man. Indian Med Gaz. 1941; 76:513-5.

13. Rajam RV, Viswanathan GC. Rhinosporidiosis: a study with a report of a fatal case with systemic dissemination. Indian J Surg 1955; 17:269-98.

14. Nguyen van UT, Nguyen van AI, Nguyen tri loc, Nguyen luv vein, Tran Ngog Ninh. A case of nasal, cutaneous and osseous rhinosporidiosis. PresseMed.1959; 67:2073-5.

15. Arivandan KP, Vishwanthan MK, Jose L.Rhinosporidium of bone-A case report. Indian Journal Pathol Microbiol.1989 ; 32(4):312-313

16. ChatterjeePK, KhatuaCR, ChatterjeeSN, DastidarN. Recurrent multiple rhinosporidiosis with osteolytic lesions in hand and foot. J. Laryngol Otol. 1977; 91(8):729-34.

17. ThomasT, GopinathN and BettsRH. Rhinosporidiosis of the Bronchus.Brit J,Surg 1956; 44:316.

18. Subramanyam CVS, and Ramana RaoAV. A fatal case of Tracheobronchial Rhinosporidosis.Brit J. Surg.1960; 47:411

19. MahadevanR. A rare case of parotid salivary cyst due to Rhinosporidosis.Indian J.Surg. 1952;14:271

20. Kamalchandra, RatnakarCR. Rhinosporidosis of parotid duct. Journal of Laryng. Otolology 1971; 85:1083

21. HoMS, TayBK. Disseminated Rhinosporidiosis. Ann Acad. Med Singapore. 1986; 15:80-3

22. Chandra Shekharyga SH, Suligavi SS, Chogule S, Dodddamani SS. Nasal Rhinosporidiosis Revisited. Clinical Rhinology:An International Journal .May-Aug 2010;3(2):735-75.

23. Kameshwaran M, Anand K, Murali S, Raghunandhan, Jacob J. KTP 532 in management of Rhinosporidiosis. Indian Journal of Otolaryngology and Head and Neck Surgery.Oct-Dec 2005; 54(4):298-300.

24. Nair KK. Clinical trial of diaminodiphenlyl sulphone in nasal and nasopharyngeal Rhinosporidosis. Laryngoscope.1979; 89: 291-5.

25. WoodardB, Hudson J. Rhinosporidosis ultra structural study of an infection in South Carolina.South Med.J. 1984; 77:1587-8.

26. Herr RA, MendozaL, ArseculeratneSN, AjelloL. Immunolocalization of endogenous antigenic material of Rhinosporidium seeberi only during mature sporangial development. FEMS.Immunol Med Microbiol. 1999;23: 205-12

.....

**How to cite this article?**

Goswami R. Rhinosporidiosis: Bizarre Clinical presentation and Mode of Dissemination. Int J Med Res Rev 2014;2(1):58-62, 10.17511/ijmrr.2014.i01.14

……