

Disseminated cutaneous rhinosporidiosis

¹TM Anoop, ²A Rajany, ³PS Deepa, ⁴P Sangamithra, ⁵R Jayaprakash

¹⁻³Resident; ⁴Associate Professor; ⁵Professor, Department of Medicine, Kottayam Medical College, Kerala, India

ABSTRACT Rhinosporidiosis is a chronic granulomatous disease of the mucocutaneous tissue, which clinically presents as polypoidal growths. Cutaneous lesions are infrequent and are generally associated with mucosal lesions. We present a case of disseminated cutaneous rhinosporidiosis in a 61-year-old male, a farmer from rural southern India who presented with generalised nodulo-ulcerative lesions all over the body but mainly on the face, thorax and extremities. The patient had a history of a surgical excision of a nasal polyp. No mucosal lesions were detected on anterior and posterior rhinoscopy. Fine needle aspiration cytology from the nodules stained by Papanicolaou and Gomori methenamine silver showed the presence of endospores suggestive of rhinosporidiosis. The patient was treated with dapsone 100 mg daily for six months. At a six-month follow-up the cutaneous lesions were unchanged. The majority of the cases of cutaneous rhinosporidiosis described in the literature have a primary lesion inside the nose. Here we report a rare case of disseminated cutaneous rhinosporidiosis without mucosal involvement.

Published online May 2008

Correspondence to TM Anoop,
Department of Medicine,
Kottayam Medical College, Kerala,
South India 686008

tel. +91 9447134973
e-mail dranooptm@yahoo.co.in

KEYWORDS Dapsone, disseminated cutaneous rhinosporidiosis

DECLARATION OF INTERESTS No conflict of interests declared.

CASE REPORT

A 61-year-old farmer from rural southern India presented with generalised nodulo-ulcerative lesions which had gradually increased in size and spread all over the body. The lesions had been present for three years, starting on the face and gradually spreading to the thorax and extremities. The patient had undergone surgical excision of a nasal polyp eight years earlier. Since he was a farmer, the patient had a history of contact with pond water. There was no history of atopy or hypersensitivity. He also had a history of bleeding from a cutaneous ulcerative lesion, which had been cauterised previously. On examination, multiple skin-coloured and erythematous nodules varying from 0.5–2 cm in size, most showing central ulceration with haemorrhagic crusting, were distributed over the face (Figure 1A), trunk and extremities (Figure 1B). Anterior and posterior rhinoscopy revealed no mass or polyps, systemic examination revealed no abnormality, and chest X-ray and ultrasonography of the abdomen showed no abnormality. A serology test for HIV and syphilis was negative.

Smears from fine needle aspiration cytology (FNAC) from the nodules, stained by the Papanicolaou (Figure 2C) and the Gomori methenamine silver methods (Figure 2D), showed the presence of endospores suggestive of rhinosporidiosis. Since surgical excision was not feasible for this patient with disseminated cutaneous rhinosporidiosis, he was advised to continue dapsone 100 mg daily for six months. At a six-month follow-up, the cutaneous lesions were unchanged. Although the patient did not show any improvement, he was reassured.

DISCUSSION

Rhinosporidiosis was first identified by Malbran in 1892. The pathogen was identified as a protozoan by Seeber in 1900 and as a phycomyces by Ashworth in 1923.¹ *Rhinosporidium seeberi* is the aetiological agent. Morphologically, it resembles both fungi and protozoa, and its taxonomy has been debated for decades. The micro-organism has not been isolated or grown in microbiological culture,² and its taxonomic position was only resolved in 1999 when Herr et al,³ using molecular biological techniques, definitively placed it in a new class, the Mesomycetozoea. This classification was independently confirmed in 2000.⁴

Rhinosporidiosis is a disease affecting primarily the mucosa of the nose, conjunctiva and urethra. Approximately 90% of all known cases occur in India, where the prevalence has been estimated at 1.4%,⁵ and in Sri Lanka. The disease has also been reported in America, Europe and Africa.⁶ Except in ocular rhinosporidiosis, which has a female preponderance, rhinosporidiosis in respiratory sites shows a higher incidence in males. Infection is usually caused by contact with stagnant ground water such as that in lakes and ponds. Clinically, the disease presents as polypoid, tumour-like, papillomatous or warty lesions which are hyperplastic, highly vascularised and friable. They may be sessile or pedunculated.

The common sites are the nose (the primary site of infection), the eye and its appendages, conjunctiva and the urethra. The larynx, trachea, skin and lungs are less



FIGURE 1 Disseminated cutaneous rhinosporidiosis in a 61-year-old male from rural southern India, who presented with generalised nodulo-ulcerative lesions all over the body but mainly on the face (A) with central ulceration and haemorrhagic crusting and (B) over an extremity.

frequently involved. The most common sites in the upper respiratory tract are the anterior nares and the inferior turbinates, the septum and the floor of the nasal cavity. Posteriorly, rhinosporidial polyps occur in the nasopharynx, larynx and soft palate. The buccal cavity is only rarely affected.

About 15% of cases of rhinosporidiosis are ocular in location, in the bulbar and palpebral conjunctiva. Osteolytic bone infiltration is another clinical presentation. Generalised rhinosporidiosis with skin and visceral involvement is extremely rare.¹⁰ Even though subcutaneous lesions are highly unusual, there are case reports of nasal rhinosporidiosis with cutaneous and subcutaneous dissemination.¹¹ Disseminated cutaneous lesions presenting as tumour-like swellings in a 48-year-old immunocompetent patient have also been reported.¹²

The presumed mode of infection from the natural aquatic habitat of the organism is through traumatised epithelium ('transepithelial infection'), most commonly

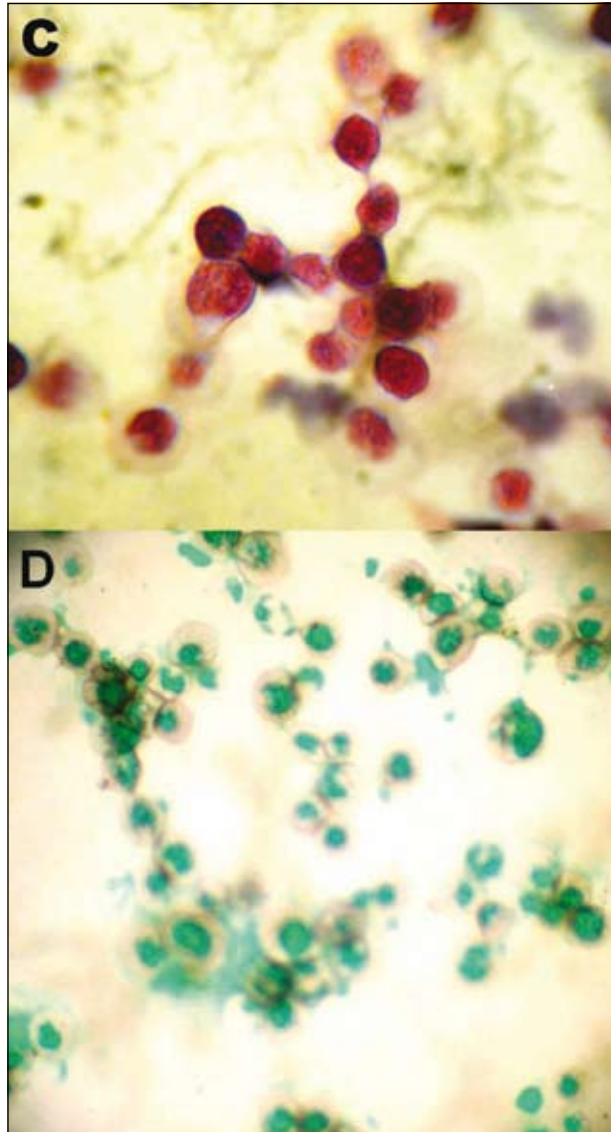


FIGURE 2 (C) Smear from a fine needle aspiration cytology of a nodule stained by the Papanicolaou method to show endospores, and (D) stained by Gomori methenamine silver staining to show multiple endospores; both images suggestive of rhinosporidiosis.

in nasal sites.⁷ Cutaneous lesions, although rare, can occur due to autoinoculation⁸ or haematogenous spread⁹ and are classified as:

1. Satellite lesions around the nasal polypoidal lesions
2. Disseminated lesions with visceral involvement
3. Primary cutaneous lesions without internal organ involvement.¹⁰

The absence of rhinosporidiosis in the sexual partners of patients with urethral rhinosporidiosis is cogent evidence that the disease is neither infectious nor contagious. Rhinosporidiosis has also been documented as occurring in several species of domesticated and wild animals, including cattle, buffaloes, dogs, cats, goats, horses, mules, several species of ducks, swans,¹³ geese and waterfowl.

A diagnosis of the disease can be made by simple aspiration cytology, the examination of aspirated material with Gomori methenamine silver and periodic acid–Schiff reaction, and the presence of the organism in different stages of maturation even in the absence of a histopathological study. However, a definitive diagnosis of rhinosporidiosis by histopathology on biopsied or resected tissues depends on the identification of the pathogen in its diverse stages of maturation. The biological agent has a mature stage that consists of large, thick-walled spherical structures called sporangia and smaller daughter cells called endospores that can be visualised with fungal stains as well as with standard haematoxylin and eosin staining. More refined morphological criteria are based on determining the diameter of the endospores (about 5 µm) and sporangia (about 1,000 µm).

The differential diagnosis is from coccidioidomycosis and adiaspiromycosis, which are easily distinguished by the different clinical presentation and the smaller size of the spherical coccidioid sporangia of less than 60 µm.¹⁴ The adiaconidia of adiaspiromycosis are much larger, have a thicker wall and do not contain endospores. In addition, the sporangia of the *Rhinosporidium seeberi* are distinguished from those of both coccidioidomycosis

and adiaspiromycosis by the zonation of the internal endospores and by the presence of distinctive spherical bodies contained within the mature endospores.

Although cases of spontaneous regression have been recorded, they are rare, and the mode of treatment remains surgical. Total excision of the polyp, preferably by electrocautery, is recommended. The only drug that has been found to have some anti-rhinosporidial effect is dapsone (4,4 diaminodiphenyl sulphone), which appears to arrest the maturation of the sporangia and to promote fibrosis in the stroma when used as an adjunct to surgery.¹⁵ Dapsone is usually given in an attempt to cure or prevent the recurrence of rhinosporidiosis.¹⁶

CONCLUSION

Rhinosporidiosis is a chronic granulomatous infection caused by *Rhinosporidium seeberi*. Mucosal involvement of the nose, nasopharynx and soft palate in the form of sessile or pedunculated vascular polyps is the most common presentation. Disseminated cutaneous lesions without mucosal involvement are extremely rare.¹⁷ The mode of spread in our patient could have been due to autoinoculation but is more likely to have been haematogenous spread.

REFERENCES

- 1 Ashworth JH. On *Rhinosporidium seeberi* with special reference to its sporulation and affinities. *Trans Royal Soc Edinb* 1923; 53:301–42.
- 2 Ahluwalia KB. New interpretations in rhinosporidiosis, enigmatic disease of the last nine decades. *J Submicrosc Cytol Pathol* 1992; 24:109–14.
- 3 Fredricks DN, Jolley JA, Lepp PW et al. *Rhinosporidium seeberi*: A human pathogen from a novel group of aquatic protistan parasites. *Emerg Infect Diseases* 2000; 6(3): 273–82.
- 4 Herr RA, Ajello L, Taylor JW et al. Phylogenetic analysis of *Rhinosporidium seeberi*'s 18S small-subunit ribosomal DNA groups this pathogen among members of the protoctistan Mesomycetozoa clade. *J Clin Microbiol* 1999; 37(9): 2750–4.
- 5 Moses JD, Shanmugham A. Epidemiological survey of rhinosporidiosis in man – a sample survey in a high school located in a hyperendemic area. *Indian Veterinary Journal* 1987; 64:34–8.
- 6 Loh KS, Chong SM, Pang YT et al. Rhinosporidiosis: differential diagnosis of a large nasal mass. *Otolaryngol Head Neck Surg* 2001; 124:121–2.
- 7 Karunaratne WAE. The pathology of rhinosporidiosis. *J Path Bact* 1934; XLII:193–202.
- 8 Karunaratne WAE. *Rhinosporidiosis in man*. London: The Athlone Press; 1964.
- 9 Rajam RV, Viswanathan GC. Rhinosporidiosis: A study with a report of a fatal case with systemic dissemination. *Ind J Surg* 1955; 17:269–98.
- 10 Kumari R, Laxmisha C, Thappa DM. Disseminated cutaneous rhinosporidiosis. *Dermatol Online J* 2005; 11(1):19.
- 11 Nayak S, Acharjya B, Devi B et al. Disseminated cutaneous rhinosporidiosis. *Indian J Dermatol Venereol Leprol* 2007; 73:185–7.
- 12 Tolat SN, Gokhale NR, Belgaumkar VA et al. Disseminated cutaneous rhinosporidiosis in an immunocompetent male. *Indian J Dermatol Venereol Leprol* 2007; 73:343–5.
- 13 Kennedy FA, Buggage RR, Ajello L. Rhinosporidiosis: A description of an unprecedented outbreak in captive swans (*Cygnus* spp.) and a proposal for revision of the ontogenetic nomenclature of *Rhinosporidium seeberi*. *J Med Vet Mycol* 1995; 37:157–65.
- 14 Longley BJ. Fungal diseases. In: Elder D, Elenitsas R, Jaworsky C, editors. *Lever's Histopathology of the skin*. 8th ed. Philadelphia: Lippincott Williams and Wilkins; 1997. p. 517–52.
- 15 Job A, Venkateswaran S, Mathan M et al. Medical therapy of rhinosporidiosis with dapsone. *J Laryngol Otol* 1993; 107:809–12.
- 16 Krishnan KN. Clinical trial of diamino-diphenyl sulphone in nasal and nasopharyngeal rhinosporidiosis. *Laryngoscope* 1979; 89:291–5.
- 17 Madhavan M, Ratnakar C, Mehdiratta KS. Rhinosporidial infection of the forehead. *J Postgrad Med* 1978; 24(4):235–6.