

Case Report

Disseminated Cutaneous Rhinosporidiosis: a Tumor like Lesion with Therapeutic Challenge

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ABSTRACT

Rhinosporidiosis is a granulomatous disorder caused by *Rhinosporidium seeberi*, an aquatic parasite. It spreads through contaminated water and soil. It frequently involves the nasal cavity and nasopharynx. Disseminated or systemic lesions are rare. A 56 year old man presented with multiple tumor like lesions all over the body in our cancer institute in April 2012. Fine needle aspiration cytology (FNAC) from the skin lesions revealed many sporangia and spores of *R. seeberi*. He had history of nasal mass being removed by excision and was immunocompetent. Histopathological examination and periodic acid Schiff (PAS) stain confirmed it to be disseminated rhinosporidiosis. His accessible lesions were excised with cautery and he was put on oral dapsons therapy. He did not show recurrence till six months of therapy. A common lesion with an uncommon presentation should not be missed which can increase the morbidity and even mortality in such a case.

Keywords: Rhinosporidiosis, Skin, India, Case Report

Introduction

Rhinosporidiosis is a chronic granulomatous disease caused by *Rhinosporidiosis seeberi*, an aquatic parasite of class Mesozoa. It is endemic in South East Asia, South America, and Africa, highest incidence being in Sri Lanka and India (prevalence rate of

1.4%) (1). It is transmitted by direct contact with spores through stagnant water and soil. Rhinosporidiosis frequently involves nasal cavity and nasopharynx (70%), presenting as a painless, friable and polypoidal growth (2). Disseminated systemic or cutaneous manifestation presenting as tumor-like swellings is uncommon. Cutaneous lesions may present as verrucous plaques,

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papules and nodules with whitish areas, crusting, and bleeding on the surface (3). Less than 30 cases have been reported in English literature till date. Disseminated rhinosporidiosis may mimic tumor lesions and pose diagnostic dilemma and threat to life if it is misdiagnosed or its treatment is delayed.

We here report a case of disseminated cutaneous rhinosporidiosis presenting as tumor like lesions all over the body.

Case Report

A 56-year-old man presented at the Surgery Out Patients Department (OPD) of our cancer institute in April 2012 with multiple nodular lesions over the body. He also had a recurrent growth in the left nostril with anosmia for five years. Before coming to our OPD, the nasal growth was excised twice at a local clinic. He gave history of bathing in local pond. Over the last one year he gradually developed multiple lesions in oral cavity, trunk, limbs, head, genitals and anal verge (Fig. 1).



Fig. 1: a) Tongue growth; b) Mass in nose and shoulders; c) Growth over penis and left wrist; d) Growth at anal verge and right foot

On examination, the lesions were non-tender, firm with lobulated surfaces and some of them showed ulceration. There was no regional lymphadenopathy. The distal pulses were normal and there was no evidence of neurological deficit.

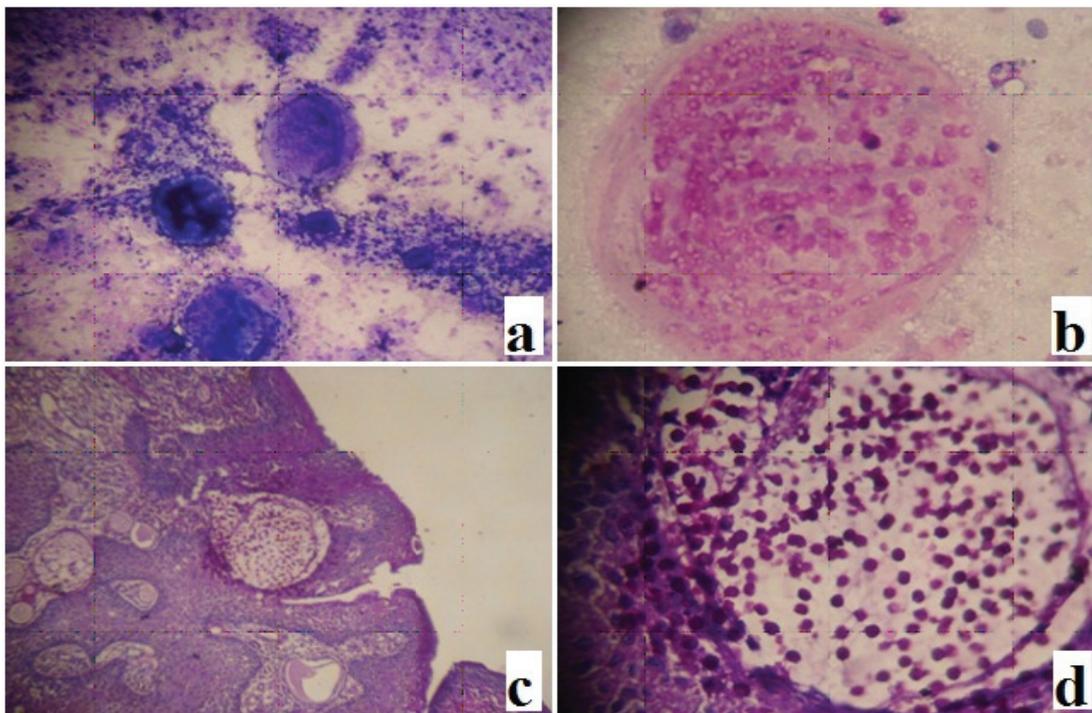


Fig. 2: a) FNAC-Sporangia with spores, (Lieshman Giemsa, $\times 100$).; b) FNAC-Sporangia with spores, (Per Iodic Acid Schiff (PAS), $\times 400$).; c) Biopsy-Sporangia with spores, (Hematoxylin and Eosin stain, $\times 100$).; d) Biopsy-Sporangia with spores, (PAS stain, $\times 400$).

Radiological investigations such as X-ray chest and ultrasonography of abdomen, complete haemogram and blood sugar and lower gastrointestinal endoscopy were within normal limits. Serology for Human Immunodeficiency Virus, Hepatitis B surface Antigen and Hepatitis C antibody were negative.

After obtaining the patient's consent, fine needle aspiration cytology (FNAC) was done from multiple swellings with 23 G needle and stained by Papanicolaou, Lishman-Giemsa and Periodic Acid Schiff stains (PAS) (Fig. 2a, b).

The smears showed many sporangia with endospores along with aggregates of neutrophils and few eosinophils in a necrotic background. Trucut biopsy from left forearm mass and punch biopsy from nasal growth were done. The histopathological examination showed many globular cysts of varying sizes representing immature and mature sporangia in various stages of maturation, beneath the epithelium. The stroma showed mild inflammatory reaction (Fig. 2 c, d). No visceral organ involvement was detected in our patient. Final diagnosis of disseminated cutaneous rhinosporidiosis was made. All his accessible lesions were excised and he was put on oral dapsone (100 mg/day) therapy. He did not show recurrence till six months of therapy, but was lost on follow up after that.

Discussion

Rhinosporidiosis has been known for over a hundred years. It was first reported by Malbran (1892), described as a protozoan by Guellermo Seeber in Argentina (1900) and as a phycomyces by Ashworth (1923) (4). The taxonomy remained debated for long time. It was finally placed in its present position of mesomycetozoa (group related to fish pathogen) by Herr *et al.* in 1999 (5) reconfirmed by Fredericks *et al.* (2000) (6). The incidence is greater in males aged between 20 and 40 years (1,2). In India the highest incidence is seen in coastal states especially Tamil Nadu

and also West Bengal (7, 8).

Rhinosporidiosis is usually limited to the surface epithelium of nasal mucosa, but rarely wide dissemination with cutaneous and/ or visceral involvement can occur (1, 3). Less than 20 cases of disseminated cutaneous rhinosporidiosis have been reported till date. The nasal lesions usually start as a small papule that grows into a polypoidal mass causing obstruction of the nose. Cutaneous lesions in rhinosporidiosis usually start as friable papillomas that become pedunculated (2).

Rhinosporidium seeberi cannot be isolated in synthetic media though it grows in cell culture. It can be diagnosed by its distinctive morphology on histopathology- many round thick walled cysts (sporangia), up to 0.5mm in diameter with endospores (6-7 μ in diameter) in different stages of maturation. These spores are positive for PAS stain. The diagnosis can also be made on cytology (FNAC) by 10% KOH and Papanicolaou stain (9). The disease must be differentiated from *Coccidioides immitis* whose different clinical presentation, smaller sporangia size (<60 μ in diameter) and negative stain by mucicarmine may be clues for identification (9). The disease may also be confused with a neoplastic lesion, as in this case.

Several modes of spread have been postulated for cutaneous rhinosporidiosis: like direct inoculation or autoinoculation through traumatized epithelium and subsequent hemato-lymphoid spread (10). Follow-up is mandatory because of high recurrences, which may be due to inadequate excision or re-infection. There is a possibility that the disease may be disseminated through FNAC. FNAC was done in this case because it was mistaken as multiple tumors and referred to our institute.

Disseminated cutaneous rhinosporidiosis with nasopharyngeal involvement has been reported by some authors (1, 3, 10, 11). Whether lowered immunity leads to widespread dissemination needs to be assessed by larger trials. However case reports as by Tolat *et al.* have not proven such (11). Our case was also immunocompetent

like Tolat *et al.* (11). Kumari R *et al.* reported disseminated cutaneous rhinosporidiosis with pulmonary involvement (12). Our patient had nasal and anal involvement but no lesions in the visceral organs like lungs, liver, kidneys or retroperitoneum were detected. As our patient did not come for further follow up, it cannot be said with certainty if he is free from disease till date.

In spite of its recognition, rhinosporidiosis remains a therapeutic challenge with a high risk of recurrence and occasional widespread and fatal complications (13). Surgical removal and electrodesiccation remain the cornerstones of therapy (10, 12). Several drugs such as dapsone (4, 4-diaminodiphenyl sulphone), ketoconazole, ciprofloxacin and amphotericin B have been tried. Dapsone has been found to have some anti-rhinosporidial effect by arresting the maturation of the sporangia and promoting fibrosis in the stroma, when used as an adjunct to surgery (14).

Conclusion

The case is presented to highlight the importance of early recognition of a common lesion with an uncommon presentation and a proper plan to manage such lesions.

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The authors declare that there is no conflict of interests.

References

1. Anoop TM, Rajany A, Deepa PS, Sangamithra P, Jayaprakash R. Disseminated cutaneous rhinosporidiosis. *J R Coll Physicians Edinb* 2008;38:123-5.
2. Hay RJ, Ajello L. Topley and Wilson's Microbiology and Microbial infection. Medical Mycology. 9th ed. London: Hodder Arnold; 1998.
3. Thappa DM, Venkatesan S, Sirka CS, Jaishankar TJ, Gopalkrishnan, Ratnakar C. Disseminated cutaneous rhinosporidiosis. *J Dermatol* 1998;25 (8):527-32.
4. Ashworth JH. On *Rhinosporidium seeberi* with special reference to its sporulation and affinities. *Trans Royal Soc Edinb* 1923;53: 301-42.
5. Herr RA, Anjello L, Taylor, Arsecularatne SN, Mendoza L. Phylogenetic analysis of *Rhinosporidium Seeberi*'s 18S small-subunit ribosomal DNA groups this pathogen among members of this protistan Mesomycetozoa clade. *J Clin Microbiol* 1999; 37(9):2750-4.
6. Fredericks DN, Jolley JA, Lepp PW, Kosek JC, Relman DA. *Rhinosporidium seeberi*: A human pathogen from a novel group of aquatic protistan parasites. *Emerg Infect Dis* 2000; 6 (3):273-82.
7. Moses JS, Shanmugham A, Kingsly N, Vijayan JC, Balachandran C, Venkateswaren, *et al.* Epidemiological survey of rhinosporidiosis in Kanyakumari district of Tamil Nadu. *Mycopathologia* 1988;101(3):177-9.
8. Saha SN, Mondal AR, Bera SP, Das S, Banerjee AR. Rhinosporidial infection in West Bengal: a Calcutta based hospital study. *Indian J Otolaryngol Head Neck Surg* 2001; 53 (2)100-4.
9. Elder D, Elenitsas R, Jaworsky C, Johnson B. *Lever's Histopathology of the skin*. 8th ed. Philadelphia: Lippincott Williams and Wilkins; 1997.
10. Nayak S, Acharjya B, Devi B, Sahoo A, Singh N. Disseminated cutaneous rhinosporidiosis. *Indian J Dermatol Venereol Leprol* 2007; 73 (3):185-7.
11. Tolat SN, Gokhale NR, Belgaumkar VA, Pradhan SN, Birud NR. Disseminated cutaneous rhinosporidiosis in an immunocompetent male. *Indian J Dermatol Venereol Leprol* 2007;73 (5):343-5.
12. Kumari R, Nath AK, Rajalaxmi R, Adityan B, Thappa DM. Disseminated cutaneous rhinosporidiosis: varied morphological appearances on the skin. *Indian J Dermatol Venereol Leprol* 2009; 75 (1):68-71.
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. Job A, Venkateswaran S, Mathan M, Krisnaswami H, Raman R. Medical therapy of rhinosporidiosis with dapsone. *J Laryngol Otol* 1993;107 (9):809-12.