

Rhinosporidiosis of the upper airways and trachea

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Abstract

Objective: We report an extremely rare case of rhinosporidiosis with involvement of both larynx and trachea, together with coexisting nasal, nasopharyngeal and oropharyngeal lesions, in a 32-year-old man.

Method: Case report and review of the world literature concerning laryngotracheal and disseminated rhinosporidiosis.

Results: A 32-year-old, South Indian man presented with a nasal mass of four years' duration, with progressive hoarseness for one year. Strawberry-like rhinosporidial masses were seen in both nasal cavities. Fibre-optic laryngoscopic examination revealed reddish masses with whitish surface specks, involving the glottis, subglottis and trachea. Computed tomography revealed soft tissue opacities involving both nasal cavities and the nasopharynx and extending to the oropharynx, with involvement of the larynx and trachea. Direct laryngoscopy and rigid bronchoscopy guided excision of the laryngeal and tracheal lesions was performed.

Conclusion: Rhinosporidiosis is a chronic, granulomatous disease which usually affects the mucous membranes of the nose and nasopharynx. Lower dissemination into the trachea is extremely rare. Laryngotracheal involvement poses many diagnostic and therapeutic challenges, due to the potential risk of bleeding and aspiration. In the presented case, local spread of rhinosporidiosis, due to direct spillage of spores from the nasopharynx into the larynx during episodes of bleeding or previous surgery, may have resulted in laryngotracheal involvement, although systemic spread cannot be excluded.

Key words: Rhinosporidiosis; Trachea; Larynx

Introduction

Rhinosporidiosis is a chronic, granulomatous infection caused by *Rhinosporidium seeberi*. It is characterised by the development of granular, polypoidal masses primarily affecting the mucous membranes of the nasal cavity and nasopharynx. The disease is more common in South India and Sri Lanka.¹ Cutaneous, laryngotracheal, genital and bony dissemination are rare, and can mimic soft tissue sarcoma.^{1–3} Following the onset of nasal lesions, it usually takes many years for systemic dissemination to occur.¹

We report an extremely rare case of rhinosporidiosis with involvement of the larynx and trachea, together with coexisting nasal, nasopharyngeal and oropharyngeal lesions.

Case report

A 32-year-old man from an urban region of South India presented complaining of masses in both nostrils for four years, associated with progressive hoarseness of voice for one year. There had been many episodes of bleeding from the nasal masses. One year previously, the patient had noticed the gradual onset of progressive voice change, with blood-stained sputum on coughing. The patient had undergone three surgical procedures for the nasal and nasopharyngeal lesions. The most recent procedure had been one and a half years ago; six months later, the patient had again begun to develop throat symptoms. The patient had never bathed in ponds, and nobody from his neighbourhood had suffered similar illness.

Nasal cavity examination revealed reddish, strawberry-like, nodular, irregularly surfaced lesions studded with

whitish specs on the mucosa of both nasal cavities, including both vestibules (Figure 1), and extending to the nasopharynx and oropharynx (Figure 2).

Tele-laryngoscopy showed multiple, sessile, reddish, nodular, strawberry-like masses with whitish specs on their surface, involving the laryngeal inlet and extending to the subglottis (Figure 3). Fibre-optic laryngoscopic examination revealed multiple sessile lesions occluding the tracheal lumen, more on the right lateral wall than the left.

Computed tomography (CT) revealed soft tissue opacities involving both nasal cavities and the nasopharynx (Figure 4) and extending to the oropharynx, with involvement of the larynx and trachea (Figure 5).

Routine haematological investigations, chest radiography and funduscopy were all normal. The patient was seronegative for human immunodeficiency virus.

A clinical diagnosis of laryngotracheal rhinosporidiosis was made. Diode laser excision of lesions was performed under direct laryngoscopy and rigid bronchoscopy guidance.

Histopathological examination of the excised specimens revealed hyperplastic epithelium with globular cysts of various shapes, representing sporangia at various stages of development (Figure 6).

Post-operatively, the patient was commenced on dapsone 100 mg/day, which he was advised to continue for two years.

Discussion

Rhinosporidiosis was first reported in Eastern India in 1894, by O'Kinealy.⁴ The disease is caused by *Rhinosporidium*

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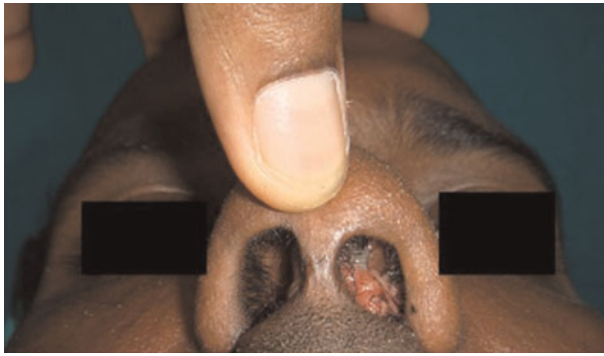


FIG. 1

Clinical photograph showing reddish rhinosporidial mass involving the left nasal cavity up to the vestibule.

seebri, a protist belonging to the mesomycetozoa family of aquatic parasites.^{1,5} In 1923, Ashworth named the organism *Rhinosporidium seeberi*, after Guillermo Seeber, who submitted a doctoral thesis on the topic in 1900.⁴

Although rhinosporidiosis has been reported in the Americas, Europe, Africa and Asia, it is far more common in the tropics, with greatest prevalence seen in South India and Sri Lanka.^{1,6,7} It may be transmitted by direct contact with the spores of *R seeberi* via dust or infected clothing, or through swimming in stagnant water contaminated with the spores. The organism has developed various mechanisms of immune evasion (i.e. immune deviation, suppression of local immune responses, immunoglobulin binding and antigenic variation), making both humoral and cell-mediated immunity less effective against the infection.⁵

Nasopharyngeal lesions are seen predominantly in males, while ocular infection is more prevalent among females.¹ Infection produces granulomatous inflammation of the affected tissues. In the nose, infection produces painless mucosal papules that evolve into large and hyperplastic polypoidal masses which characteristically bleed on touch and have a raspberry-coloured background with white specs (representing the chitinous walls of rhinosporidial sporangia).^{1,6} Masses are usually unilateral and pedunculated, and rarely exteriorise. Other mucosal sites are involved infrequently, including the palpebral conjunctiva, oropharynx,



FIG. 2

Clinical photograph showing reddish, strawberry-like rhinosporidial masses in the oropharynx, hanging down from the nasopharynx behind the uvula.

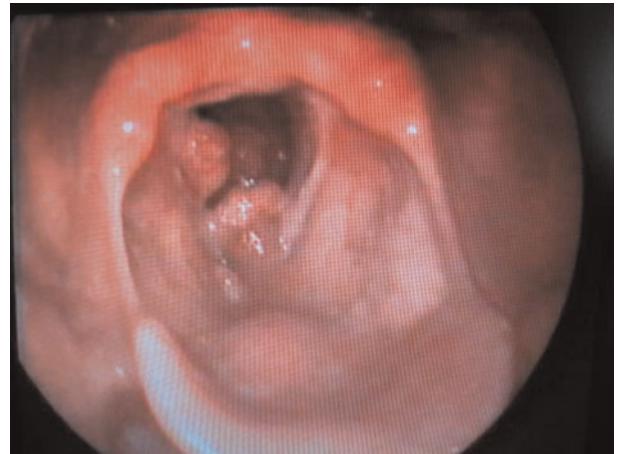


FIG. 3

Tele-laryngoscopic view showing reddish, strawberry-like rhinosporidial mass with whitish spots on its surface, in the laryngeal inlet and involving the glottis and subglottis.

nasopharynx, maxillary antrum, larynx, external ear canal, parotid duct and genitalia.^{3,6,7} Oral and oropharyngeal lesions may interfere with breathing and/or eating.⁶

Systemic dissemination to the trachea, lung, urethra, liver, genitalia, lacrimal sac and bone is very rare.^{1,3,6} Dissemination can be fatal, and occurs more commonly in association with immunodeficiency disorders (such as acquired immunodeficiency syndrome). Infection can spread via body fluids such as ascites and blood. Airway obstruction, profuse bleeding and multiorgan failure often precipitate death.⁴ In our patient, laryngotracheal dissemination could have been due to implantation of spores into the laryngeal and tracheal mucosa during episodes of recurrent bleeding from nasopharyngeal lesions and during previous surgery. Biopsy of such lesions must be avoided due to the risk of bleeding and aspiration, which may prove fatal. Computed tomography of the neck and thorax is preferred in these situations to evaluate the extent of laryngotracheal lesions.

Diagnosis is confirmed on the basis of histopathological demonstration of the characteristic thick-walled giant sporangia (measuring 60–450 μm or more in diameter) in various stages of development and containing

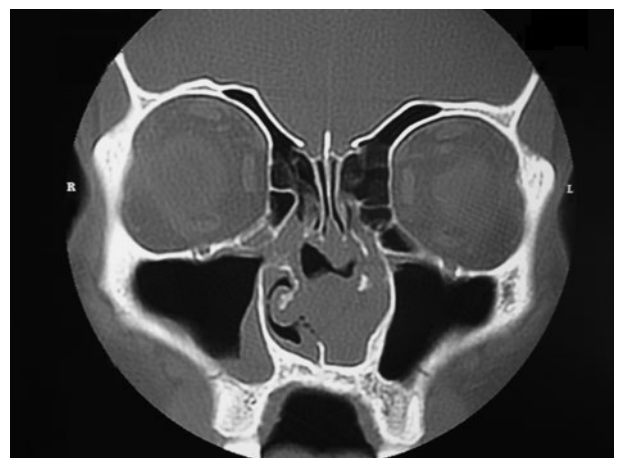


FIG. 4

Coronal computed tomography scan showing soft tissue opacities involving both nasal cavities and the nasopharynx. R = right; L = left

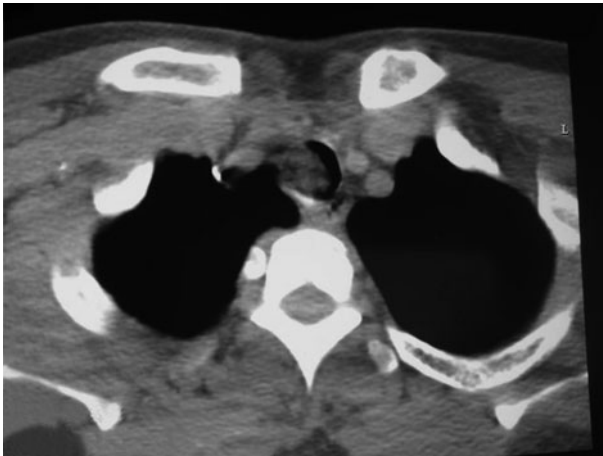


FIG. 5

Axial computed tomography scan showing a soft tissue mass lesion involving the right lateral tracheal wall and partially occluding the tracheal lumen. L = left

sporangiospores (7–15 µm in diameter, and up to 12 000 in number).^{1,6,7} *Rhinosporidium seeberi* is easily identified in haematoxylin and eosin stained smears. Spores and sporangia are better demonstrated by periodic acid Schiff, Mayer's mucicarmine, Verhoff's vonGieson and Grocott Gomori methamine silver stains.^{8,9}

Barring one report by Levy *et al.*, cultivation has not been successful.

Recently, tests for assessing the viability of the organism have been developed. Spherical bodies, provisionally regarded as the electron-dense bodies of endospores, are able to reduce the salt 3-[4,5-dimethyl-2-thiazolyl]-2,5-diphenyl-2H-tetrazolium bromide to a formazan that can be visualised microscopically.^{5,9}

Surgical extirpation is the treatment of choice. Following surgical excision, cauterisation or ablation of the lesion base is strongly recommended, as local recurrences are common due spillage of sporangia.¹ Rigid bronchoscopy combined with tracheotomy is useful in the removal of laryngotracheal lesions. The likelihood of bleeding from these lesions makes surgical intervention difficult.³ In our patient, direct laryngoscopy and rigid bronchoscopy guided

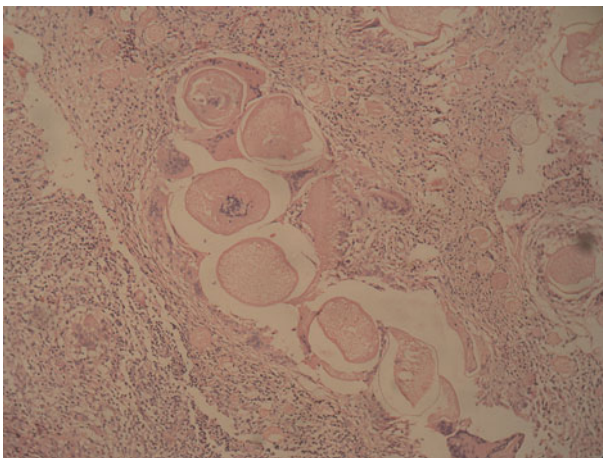


FIG. 6

Photomicrograph of the excised specimen, showing hyperplastic epithelium with globular cysts of various shapes, representing sporangia at various stages of development. (Eosin & Hematoxylin stain, 40 xMagnification)

excision of the laryngeal and tracheal lesions was performed without tracheostomy. No rhinosporidial lesions were seen in either main bronchus.

- **Rhinosporidiosis is a chronic, granulomatous disease that usually affects the mucous membranes of the nose and nasopharynx**
- **Lower dissemination into the trachea is extremely rare**
- **Laryngotracheal involvement poses many diagnostic and therapeutic challenges, due to the potential risk of bleeding and aspiration**
- **In the presented case, laryngotracheal involvement may have been due to local spread with direct spillage of spores from the nasopharynx into the larynx during episodes of bleeding or previous surgery, although systemic spread cannot be excluded**

Nasal lesions recur following surgery in approximately 10 per cent of cases; the incidence of recurrence of laryngotracheal lesions is not known due to their extreme rarity.^{3,10}

Medical treatment of rhinosporidiosis with dapsone may be effective, as it arrests the maturation of sporangia, accelerates their degeneration, increases the granulomatous response with fibrosis, and prevents recurrence by interfering with the folic acid metabolism of the organism.^{1,4,7,10}

References

- 1 Kumari R, Nath AK, Rajalakshmi R, Adityan B, Thappa DM. Disseminated cutaneous rhinosporidiosis: varied morphological appearances on the skin. *Indian J Dermatol Venereol Leprol* 2009;**75**:68–71
- 2 Adiga BK, Singh N, Arora VK, Bhatia A, Jain AK. Rhinosporidiosis. Report of a case with an unusual presentation with bony involvement. *Acta Cytol* 1997;**41**:889–91
- 3 Rekha P, Thomas B, Pappachan JM, Venugopal KP, Jayakumar TK, Sukumaran P. Tracheal rhinosporidiosis. *J Thorac Cardiovasc Surg* 2006;**132**:718–19
- 4 Raveenthiran V. Metastatic rhinosporidiosis in a child. *J Pediatr Surg* 2006;**41**:853–5
- 5 Arseculeratne SN. Rhinosporidiosis: what is the cause? *Curr Opin Infect Dis* 2005;**18**:113–18
- 6 Pang KR, Wu JJ, Lupi O, Tying SK. Tropical dermatology: fungal tropical diseases. *J Am Acad Dermatol* 2004;**50**(suppl 1):109
- 7 Kumar S, Mathew J, Cherian V, Rozario R, Kurien M. Laryngeal rhinosporidiosis: report of a rare case. *Ear Nose Throat J* 2004;**83**:568–70
- 8 Loh KS, Chong SM, Pang YT, Soh K. Rhinosporidiosis: differential diagnosis of a large nasal mass. *Otolaryngol Head Neck Surg* 2001;**124**:121–2
- 9 Nath Ak, Madana J, Yolmo D, D'Souza M. Disseminated rhinosporidiosis with unusual involvement of the nail apparatus. *Clin Exp Dermatol* 2009;**34**:e886–8 10.1111/j.1365-2230.2009.03655.x.
- 10 Pillai OS. Rhinosporidiosis of the larynx. *J Laryngol Otol* 1974;**88**:277–80

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Dr J Madana takes responsibility for the integrity of the content of the paper.

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