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CLINICAL RECORD

Disseminated rhinosporidiosis

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Abstract

Introduction: Rhinosporidiosis primarily affects the mucous membranes of the nose and nasopharynx. The disseminated form of this chronic fungal disease is extremely rare.

Case report: The authors present a case of disseminated rhinosporidiosis in an immunocompetent patient with involvement of the skin, subcutaneous tissue, muscle, bone, penis and urethra, and with a long-standing primary lesion in the nose.

Discussion: A late or atypical presentation of rhinosporidiosis may cause diagnostic dilemma. Fine needle aspiration cytology of the tumoural lesions may establish the diagnosis. Histopathology is confirmatory. The subcutaneous masses may be solid or cystic. Ulceroproliferative lesions need to be differentiated from malignancies.

Conclusion: This is the first reported case of truly disseminated rhinosporidiosis with simultaneous involvement of multiple anatomically unrelated sites in a single patient. This is also the first reported case of cystic rhinosporidiosis. The possibility and sequelae of spontaneous regression of rhinosporidiosis are also discussed.

Key words: Rhinosporidiosis; Rhinosporidium seeberi; Sporangia; Cysts; Skin; Urethra; Bone; Pathology; Otolaryngology

Introduction

Rhinosporidiosis is a chronic granulomatous disease caused by a fungus-like organism, *Rhinosporidium seeberi*, which affects primarily the mucous membranes of the nose and nasopharynx. The disease sometimes produces mucocutaneous lesions in other parts of the body, mainly due to autoinoculation by infected fingernails. Dissemination of rhinosporidiosis is extremely rare. Delayed or atypical presentation and involvement of extranasal sites make the diagnosis difficult.

A case of disseminated rhinosporidiosis with simultaneous involvement of multiple, anatomically unrelated structures is reported for the first time in the literature. This case also represents the first report of a cystic lesion in rhinosporidiosis.

Case report

A 50-year-old man presented with breathing difficulties due to bilateral, pink, polypoid, fleshy, granular masses filling both nasal cavities and hanging down into the oropharynx from the post-nasal space (Figure 1). The patient also had a huge, bilobed, ulceroproliferative growth with everted margins on the left wrist, and a fleshy, bleeding mass on the right buttock. The nasal masses had the typical appearance of rhinosporidiosis, with white dots on the surface, and had been present for the last 20 years. The patient had been operated upon five times in the past for recurrent nasal rhinosporidiosis, the last operation being 10 years previously. The patient had been living with a swollen left wrist for the last three and a half years. There was mild pain but, as movement was unrestricted, he could carry on with his normal activities. He had also noticed swellings on both shoulders at approximately the same time. Three years ago, a fleshy mass had appeared on the glans penis, near the urethral orifice.

The patient had been involved in a road traffic accident two years back, with extensive soft tissue injuries all over the body. Subsequently, abscesses had developed on both shoulders, and the skin over the swollen left wrist had become ulcerated. The shoulder abscesses had ruptured spontaneously and healed with wide, stellate scars (Figure 2). The ulcerated wrist lesion had begun to grow at an increased rate after the accident, and was $10 \times 7.5 \times 5$ cm at the time of presentation (Figure 3). This lesion was fixed to the deeper structures and bled on touch.

A small, polypoid, granular, fleshy mass was seen to arise from the urethral meatus and to involve the glans penis.

Numerous subcutaneous swellings of various sizes were found on the scalp, thigh, chest, forearm and calf. All these lesions were firm, smooth, non-tender and mobile, with more or less well-defined margins.

The cutaneous lesions appeared as small nodules on the cheek or wart-like growths with short pedicles on the back. The patient also had a firm, pink, fleshy, pedunculated, $4.5 \times 3.5 \times 3$ cm growth arising from the perianal skin, which bled on touch.

There was no lymphadenopathy.

X-rays of the chest and shoulders were normal, as was ultrasonography of the abdomen. Screening for human immunodeficiency virus was negative.

A tracheostomy was performed immediately after admission to relieve respiratory obstruction, and the growth arising from the perianal skin was excised to stop the

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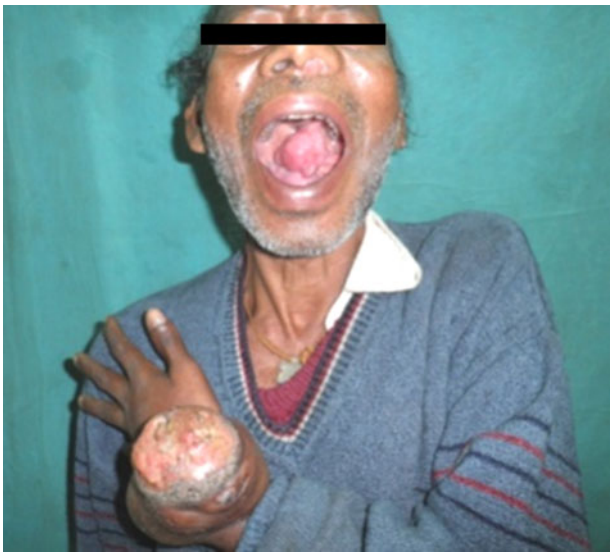

FIG. 1

Clinical photograph showing the rhinosporidiosis lesions in both nasal cavities and the left wrist, and the oropharyngeal mass protruding into the mouth, lying over the tongue.

bleeding. A wedge biopsy was taken from the ulcerated lesion on the left wrist to exclude soft tissue sarcoma. The histopathological diagnosis, for both the lesions, was rhinosporidiosis.

The nasal lesion consisted of multiple growths of varying sizes, with attachments to the vestibule, floor, septum and lateral wall of the nasal cavity as well as to the nasopharyngeal mucosa. The nasal lesions were excised with diathermy coagulation of the base. The oropharyngeal mass, having its attachment on the dorsal surface of the soft palate, was dealt with in a similar fashion.

The penile lesion was a pink, leaf-like mass with finely granular surface attached to the fossa navicularis. Wide excision of the mass with diathermy coagulation was performed, along with amputation of the glans penis.

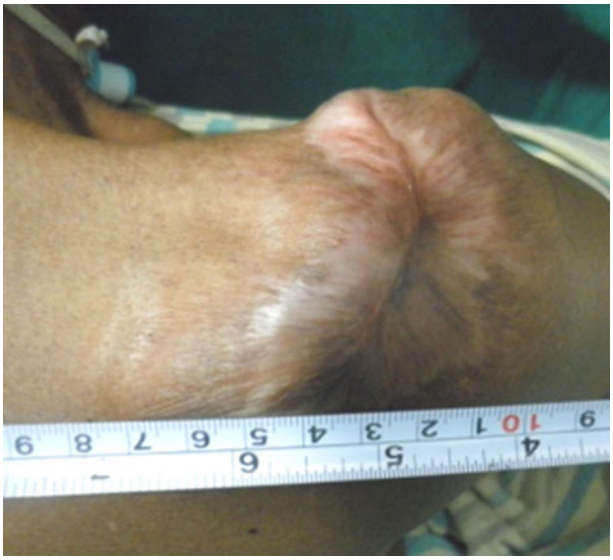

FIG. 2

Clinical photograph showing the scar on the right shoulder.

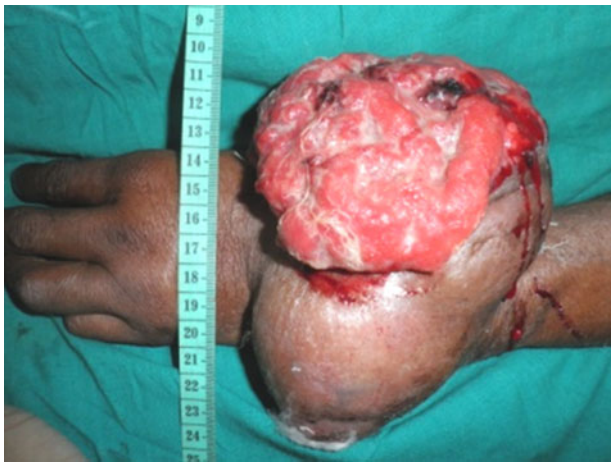

FIG. 3

Clinical photograph showing the bilobed, bleeding, ulceroproliferative rhinosporidiosis lesion on the dorsum of the left wrist.

An X-ray of the left wrist joint showed irregular destruction of the distal part of the radius and ulna. On computed tomography, the lesion was found to infiltrate the bones of the wrist joint, eroding the lower ends of the radius and ulna. A below-elbow amputation of the forearm was performed.

All the subcutaneous lesions were excised, after confirmation of the diagnosis by fine needle aspiration cytology (FNAC). Macroscopically, most of the masses had a smooth, glistening, convoluted, pale yellow, lobulated

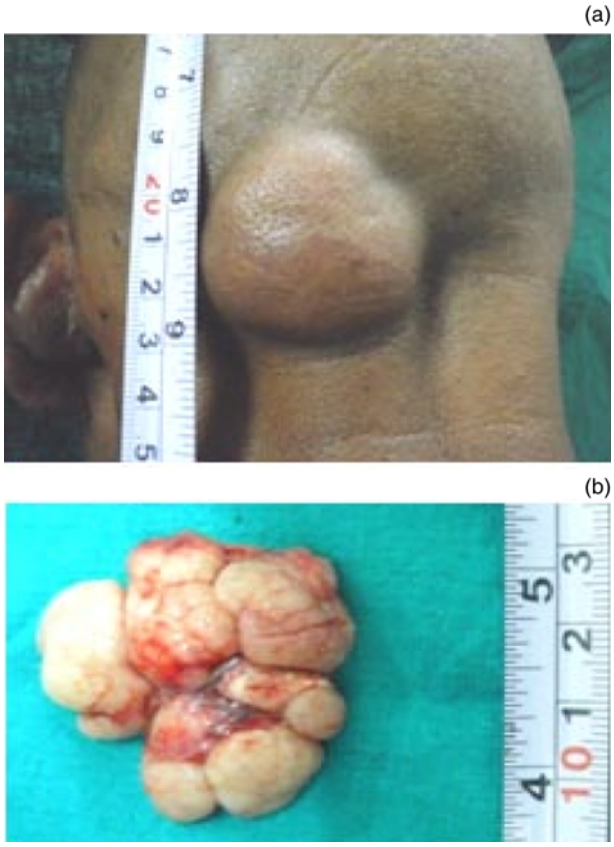

FIG. 4

Clinical photographs showing (a) subcutaneous rhinosporidiosis on the occipital scalp, and (b) the lobulated, cerebriform surface of the excised growth.

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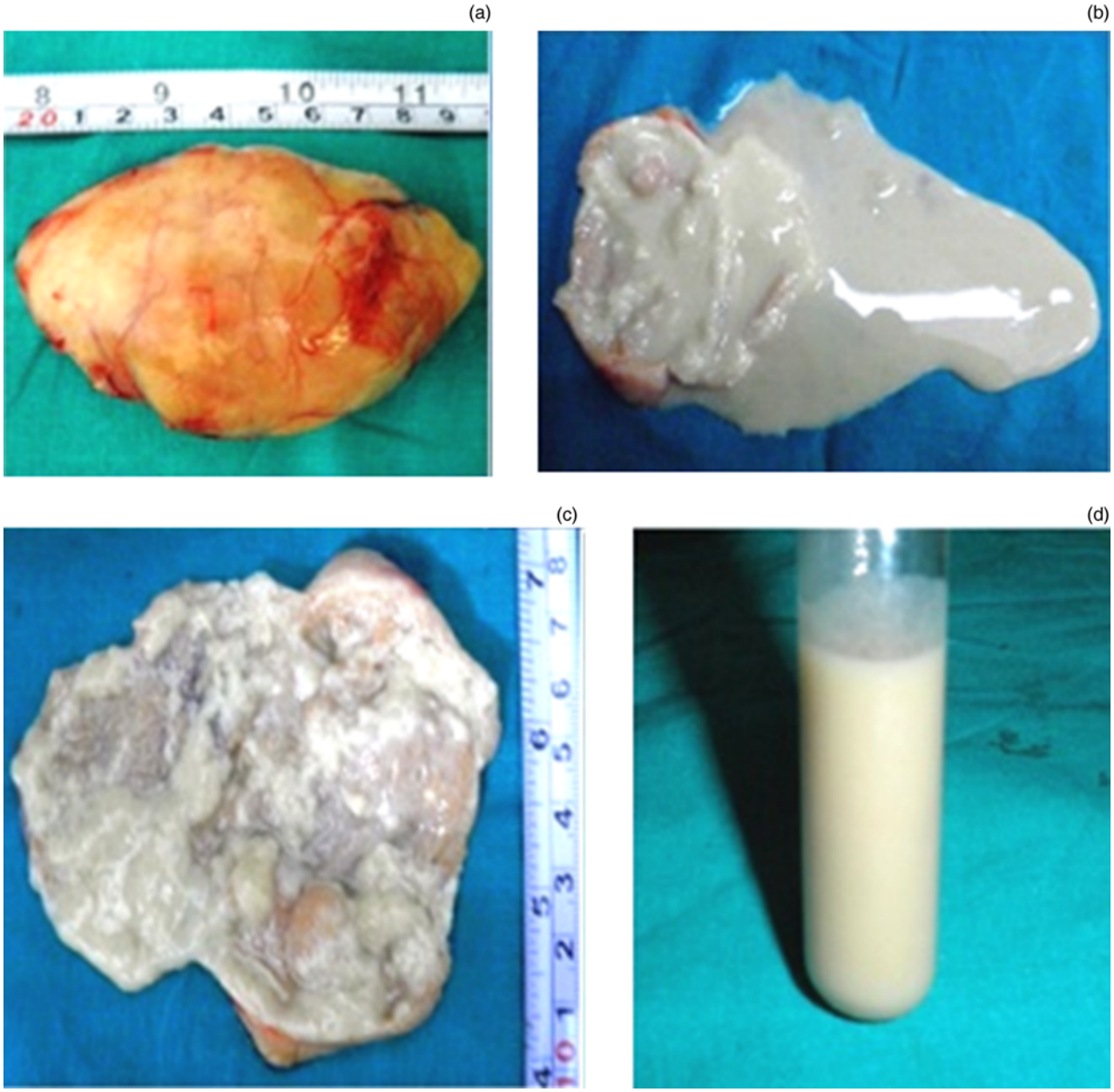

FIG. 5
Clinical photograph showing: (a) the cystic rhinosporidiosis lesion enucleated from the right calf; (b) thick fluid released upon incising the cyst; (c) the interior of the cyst; and (d) the creamy cyst fluid collected in a test tube.

surface, with a cut surface which was homogeneous and pale yellow with the consistency of brain tissue (Figure 4).

The mass removed from the right calf was pale yellow, smooth, ovoid, tense, cystic and $9 \times 6 \times 5$ cm in size, and contained approximately 20 ml of thick, cream-coloured, non-odorous fluid (Figure 5). The cyst wall was composed of thick, fibrous tissue and did not appear to contain any embedded rhinosporidiosis spores. Microscopic examination of the fluid revealed the presence of a large number of sporangia in different stages of maturation. Culture of the fluid failed to grow any organism.

Discussion

The spores of *Rhinosporidium seeberi* are possibly transmitted to humans from its natural aquatic habitat, e.g. stagnant ponds, via traumatised epithelium (i.e. transepithelial infection), most commonly the nasal mucosa.[1] Rhinosporidiosis spreads through the submucosal lymphatics and may involve multiple anatomically related and contiguous sites such as the lips, palate, uvula, conjunctiva, lacrimal sac, epiglottis, larynx or, occasionally, trachea. The infection is usually limited to the surface epithelium. The vulva, vagina and penis have also been reported to be affected infrequently.[2] Involvement of the skin, bone and other viscera is rare.[3]

Rhinosporidiosis is characterised by the formation of vascular, friable lesions which are papillomatous or polypoidal with a granular surface. Other types of presentation make the diagnosis difficult: presentation of rhinosporidiosis as a tumour,[4–6] ulceroproliferative growth[6,7] or arthropathy[3] may cause diagnostic dilemma.

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The patient was a pond-bather. The habit of nose-picking might have traumatised the mucosa, facilitating implantation of *R seeberi* spores. The patient's nasal disease recurred several times after surgical removal. Total excision of the mass with diathermy coagulation of the base is recommended to avoid recurrence. Complete excision of rhinosporidiosis is sometimes difficult, especially if lesions are sessile. In addition, it may be difficult to access the base of even polypoid lesions when they are located in the posterior nasal cavity, the meatuses or the inaccessible areas of the nasopharynx, due to limited manoeuvrability of the surgical instruments. Compromised visibility due to brisk bleeding is one important reason for inadequate removal of multiple lesions. Recurrence of rhinosporidiosis may actually represent regrowth of residual disease.

Our patient's nasal disease remained localised for more than a decade before dissemination. Once the disease began to spread, however, the involvement of different sites was quite rapid and almost simultaneous. The mucocutaneous lesions of the face, perianal region and penis might have been caused by direct contact inoculation of spores via infected fingernails.[8] However, the tumoural lesions of the shoulders, arm, thigh, leg and wrist, affecting the soft tissues and bones and without initial involvement of the overlying skin, suggest possible haematogenous spread.[6]

Trauma, in the form of a road traffic accident, modified the course of our patient's disease in two distinct ways. While the breach in the skin overlying the wrist lesion resulted in rapid growth into a fungating mass, the subcutaneous swellings over both shoulders grew rapidly to rupture and discharge pus-like fluid. The ulcers ultimately healed to leave stellate, depressed scars. The intramuscular, cystic mass enucleated from the right calf contained a large amount of thick, cream-coloured fluid enclosed in a thick, fibrous capsule. The bacteriologically sterile fluid was teeming with the sporangia of *R seeberi*, but the fibrous capsule was devoid of any spore or granuloma. The spores found in the fluid were in various stages of maturation, signifying that the organism was actively multiplying inside the cyst. Any such cystic lesion in the subcutaneous plane, if infected, may rupture and evacuate completely. As the fibrous capsule was not involved, the ulcer may have eventually healed, albeit with some scarring. The swellings on the patient's shoulders may also have been cystic lesions which ruptured and healed spontaneously. A literature search identified no previous reports of cystic rhinosporidiosis lesions.

The diagnosis of rhinosporidiosis can be made by FNAC,[9] but this may also be inconclusive.[10] The 'gold standard' for rhinosporidiosis diagnosis is histopathological identification of the pathogen, showing different stages of maturation, in biopsied or resected tissue.

Rhinosporidiosis rarely involves the bones. A total of eight cases[2,3,5–7,11–13] of osteolytic lesions have been reported to date. Wide excision or partial amputation is the treatment of choice.[2,3,5,11]

- **A case of disseminated rhinosporidiosis is presented**
- **The patient had simultaneous involvement of skin, subcutaneous tissue, genitalia, bone and muscle, with a long-standing primary nasal lesion**
- **The patient also had a cystic lesion, previously unreported in rhinosporidiosis**

Urethral rhinosporidiosis presents as a discrete, friable, painless, slow-growing, polypoidal, and pedunculated or sessile mass, mainly in the navicular fossa, and resembling condylomata acuminata or lata.[14] Surgical excision of the lesion with diathermy coagulation of the base has been advocated as the only treatment for genitourinary rhinosporidiosis.

No systemic or local medical treatment is usually effective. However, antimicrobial therapy is indicated for disseminated disease, and therapy with dapsone (4,4′-diaminodiphenyl sulphone) has sometimes been successful.[4] Dapsone may arrest sporangia maturation and accelerate their degenerative changes. The effete organisms are then removed by an accelerated granulomatous response.[8] Our patient was treated with dapsone (100 mg daily orally) post-operatively, to arrest further dissemination.

Conclusion

Disseminated rhinosporidiosis is extremely rare. The presented patient represents the first reported case of disseminated rhinosporidiosis with simultaneous involvement of the skin, subcutaneous tissue, genitalia, muscles, bone and joints, along with a huge nasal and nasopharyngeal mass. All lesions were dealt with surgically in a staged manner. There have been no previous reports of cystic lesions in rhinosporidiosis, as seen in the presented patient. We assume that cystic lesions which rupture may heal spontaneously.

Although autoinoculation of *R seeberi* spores by infected fingernails may explain the occurrence of rhinosporidiosis in various anatomically unrelated parts of the body, the involvement of bone and the appearance of a soft tissue mass with intact overlying skin suggest the possibility of additional spread of rhinosporidiosis via a haematogenous route.

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