Rhinosporidiosis of the Neck: A Rare Occurrence

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Rhinosporidiosis is a chronic granulomatous disease caused by a fungus Rhinosporidium seeberi. It frequently involves the nasopharynx and presents as a painless, friable, polypoidal growth hanging anterior to the nares or posteriorly into the pharynx. The lesions appear pink with minute white dots, which give it a strawberry-like appearance. Histopathologically, the organisms appear as sporangia containing large numbers of round or ovoid endospores. Several drugs have been tried, but Dapsone has proven to be most effective. In the present case, the lesion was seen on the anterior border of the mandible which is a very rare site for this infection. The growth was pedunculated and proliferative mimicking an aggressive lesion. On histology, also a rare finding of involvement of lymph nodes by the sporangia was noted.

Keywords: Granulomatous, Lesion, Nasopharynx

INTRODUCTION

Rhinosporidiosis has been defined as a chronic granulomatous disease characterised by production of polyps and other manifestations of hyperplasia of the nasal mucosa. The etiologic agent, Rhinosporidium seeberi, is an aquatic protozoan and was previously considered to be a fungus, however its taxonomic position is unclear.¹ It was first described by Guillermo Seeber in 1900 from Buenos Aires.² The first case from India was reported by O’Kineay in 1903.³ The life cycle of this fungus was described by Ashworth in 1923.⁴ The organism has never been isolated in vitro.⁵ The disease is endemic in India and Sri Lanka but has also been reported from the United States, South America, and Iran.⁶ It is more common in adult men and is possibly transmitted to humans by direct contact with spores through dust, through infected clothing or fingers, and through swimming in stagnant waters.⁶ Rhinosporidiosis frequently involves the nasopharynx presenting as a painless, friable, polypoidal growth, which may hang anterior to the nares or posteriorly into the pharynx. The lesions are pink or purple-red and studded with minute white dots, which are sporangia containing the spores.⁷ Infection of the nose and nasopharynx is observed in 70% of patients of rhinosporidiosis; infection of the palpebral conjunctivae or associated structures (including the lacrimal apparatus) is observed in 15%.⁵ Face and neck are rare sites of infection.

CASE REPORT

A 69-year-old male patient visited the Nitte Meenakshi Institute of Craniofacial Surgery, K. S. Hegde Medical Hospital with a chief complaint of a growth on the skin over left mandibular region since 4 years. The lesion showed a gradual increase in size since the last 2 months. It was associated with pain for the last 2 weeks. Patient gave a history of nasal growth that was surgically removed 6 months back. More details were not available. Patient also gave a history of swimming in the community pool. Patient was a known hypertensive since 2 years and was under medication for the same. No relevant family history was reported.

There was a single, well-defined, pedunculated proliferative lesion approximately 6 cm × 4 cm in size seen over the left submandibular area. It had a cobbled surface and the skin overlying the lesion was ulcerated (Figure 1). There was bleeding with pus discharge. No fixity to underlying structures was seen. Lymph nodes were bilateral level IB, palpable and non-tender.

The lesion was excised along with part of salivary glands and lymph nodes and sent to the Department of Oral Pathology, Yenepoya Dental College.

Histological Features

Microscopically the section showed the lesional tissue with the presence of numerous round sporangia with an average size of more than 40 μm, containing the fungal spores (Figures 2 and 3). Dissemination of the endospores was also seen. Surrounding these was dense infiltration of chronic inflammatory cells, chiefly lymphocytes. Pseudocysts arranged in “Swiss cheese” pattern also seen. Epithelium present was stratified squamous hyperplastic with...
obstruction of the nose and nasopharynx. Following this, there may be seen development of small cysts and pseudocysts which may discharge a combination of pus, mucus, and organisms. This discharge creates tiny white dots on the surface of the lesion that are clearly visible with a hand lens. These features give the entire lesion a characteristic strawberry-like appearance. The lesions are highly vascular and bleed easily. Cutaneous lesions in rhinosporidiosis are rare and usually start as friable papillomas that over a period become pedunculated. Cutaneous rhinosporidiosis sometimes may also be seen as warty papules and nodules with whitish spots, crusting, and bleeding on the surface.

The mode of transmission of rhinosporidiosis is still a matter of debate. Contaminated water and soil have long been established as the main source of infection. The microorganisms gain entry into the body via any traumatized epithelium.

**DISCUSSION**

Rhinosporidiosis has been known for over a 100 years and was first described in Argentina. The causative agent for this disease is *R. seeberi*, which was initially believed to be a sporozoan, but it is now considered to be a fungus. Rhinosporidiosis is, usually, limited to the surface epithelium of the nasal mucosa, but, rarely, wide dissemination with visceral involvement can occur. Anterior nares and conjunctiva are the most common site of infection, but other rare sites such as nasopharynx, larynx, maxillary antrum, skin of limbs, lachrymal sac, urethra, vagina, parotid duct, bone, vagina and rectum may also get involved. The most common presentation of the nasal lesions is an often pruritic papule that grows into an erythematous polypoidal mass that may cause

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**Figure 1:** Clinical picture showing the growth

**Figure 2:** Histological picture showing the on the mandible lesional tissue with numerous round sporangia

**Figure 3:** Sporangia containing the fungal

**Figure 4:** Section of lymph node showing spores. The presence of sporangia and spores
Cutaneous rhinosporidiosis may characteristically present as the following three types: 1. Satellite lesions, in which skin adjacent to the nasal rhinosporidiosis is involved secondarily; 2. Generalized cutaneous type with or without nasal involvement, occurring through hematogenous dissemination of the organism; 3. Primary cutaneous type associated with direct inoculation of organisms onto the skin. Several modes of spread have been postulated for cutaneous rhinosporidiosis. Demellow’s theory of direct transmission propounded by Demellow had its acceptance for quite some time. He postulated that the infection always occurred as a result of direct transmission of the organism. When nasal mucosa comes into contact with infected material while bathing in common ponds, infection found its way into the nasal mucosa. Karunaratne accounted for satellite lesions in the skin and conjunctival mucosa as a result of autoinoculation. Rhinosporidiosis affecting distant sites could be accounted for only through hematogenous spread. Apart from these a possibility of lymphatic spread has also been considered but it is extremely rare.

The diagnosis of cutaneous rhinosporidiosis can be done by checking for the presence of typical nasopharyngeal lesions and through careful inspection of the surface of warty lesions. Histopathologically the organisms appear as sporangia containing large numbers of round or ovoid endospores. The epithelium is hyperplastic with papillomatosis and deep invaginations, some of which form pseudocysts. Numerous globular cysts of varying shape, representing sporangia in different stages of development, give the corium a distinctive “Swiss cheese” appearance. There is a surrounding dense, mixed inflammatory infiltrate with lymphocytes and histiocytes, including occasional giant cells, plasma cells, neutrophils, and eosinophils. This disease must be differentiated from coccidioidomycosis whose different clinical presentation and smaller sporangia size (<60 μ in diameter) allow for an easy distinction.

The lesions of cutaneous rhinosporidiosis very rarely, if ever, regress spontaneously. The lesions, however, should be treated early in order to prevent extension or dissemination. Determining the drug sensitivity has been difficult since growing the organisms in vitro or in vivo has been a failure. Many clinicians prefer diathermy excision as the treatment of choice, but incomplete excision may result in recurrences. Drug therapy for rhinosporidiosis has not been very successful. Several drugs have been tried but dapsone alone is proven to be somewhat effective in a dose of 100 mg/day for a period of 6 months. The most probable mode of action of dapsone is to arrest the maturation of the sporangia and induce stromal fibrosis. Other authors suggest that dapsone may arrest the maturation of sporangia and accelerate degenerative changes in them. The organisms are then removed by an accelerated granulomatous response.

In the present case, rhinosporidiosis infection was manifested on the external aspect of the mandible, which is a rare site for this disease. The lesion presented as a proliferative growth which gave the impression of an aggressive lesion and could influence a clinician to make a diagnosis as a malignant neoplasm like verrucous carcinoma or squamous cell carcinoma. Another interesting feature was the presence of fungal spores in the lymph node sections which hinted towards a possible lymphatic mode of spread, which again is a rarity.

CONCLUSION

The present case highlights the rare occurrence of cutaneous rhinosporidiosis. Since the lesion is not frequently seen on the skin, it is very easy to misdiagnose the case. However careful history taking along with detailed histopathologic examination can lead to accurate diagnosis and initiation of prompt treatment.

REFERENCES