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Case Series

Ophthalmology Section

Ocular Rhinosporidiosis and Recurrence Postsurgery: A Case Series

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ABSTRACT

Rhinosporidiosis, a chronic granulomatous disease, is caused by *Rhinosporidium seeberi*. It is an endosporulating microorganism causing polypoidal ocular mass. A clinician needs to have high degree of suspicion for oculosporidiosis, as it is more common in nasal cavity and other respiratory passage as compared to ocular lesion. The authors intend to increase awareness among ophthalmologists through this case series. It was intended to evaluate profile, clinical presentation and recurrence rate of ocular rhinosporidiosis in a tertiary care setting in Eastern India. The present case series is about 15 patients diagnosed with rhinosporidiosis, that was reported within one year. Age range was 6-50 years, with male:female was 9:6. All patients underwent total excision with around 2 mm lesion free margin and electrocautery of base. Postoperatively, antibiotic and lubricant eye drops were given, a follow-up schedule was day 1, 7, 30, six months and one year. Majority of the patients showed involvement of lower tarsal conjunctiva (eight), followed by upper (four), then bulbar conjunctiva (one), lower lid (one), and lacrimal sac (one). All patients recovered completely postoperatively. At one year follow-up no patient had any recurrence. Ocular rhinosporidiosis may have diverse presentation from chalazion, papilloma, keratoconjunctivitis etc. This disease is often missed because of poor pathological back up at many centres. In an endemic area, a high degree of suspicion should be there to detect this relatively rare entity.

Keywords: Cautery, Conjunctival mass, Dapsone, Oculosporidiosis, Sporangia

INTRODUCTION

Rhinosporidiosis is a chronic, granulomatous disease, clinical picture being reddish polypoidal masses that are hyperplastic and friable, can be pedunculated or sessile. The disease is caused by *Rhinosporidium seeberi* [1]. It was previously considered to be a fungus and was classified as a fungal disease under International Classification of Diseases (ICD) 10 [2]. It is now considered as a protist classified under Mesomycetozoa. The mucous membranes of the nasal cavity and nasopharynx and anterior nasal septum are the most common sites affected by this pathogen. Extranasal involvement is rare. Conjunctiva, lips, uvula, palate, trachea, larynx, and bone are some of these rare extranasal sites [3].

The most common presentation is a soft polypoidal pedunculated mass. Polypoidal conjunctival mass is the most common presenting feature of ocular rhinosporidiosis [4], while the other presentations include diverticulum of lacrimal sac, recurrent chalazion, cyst or chronic follicular reaction of conjunctiva especially in contact lens users, keratitis, scleral melting, ciliary staphyloma or eyelid tumours or periorbital skin, leading to mechanical entropion [5]. If the lacrimal sacs are involved, it presents as bloody tear [6]. The typical conjunctival lesion is a red, fleshy, pedunculated with polypoid mass arising from the palpebral conjunctiva. These masses usually have multiple pale-yellow dots denoting mature sporangia on the surface. It has been hypothesised that when the lesion arises and spreads in the bulbar conjunctiva, there is no space to grow as the lids over the conjunctiva exert a flattening force. Therefore, the lesions in the bulbar conjunctiva are usually sessile and flatter in appearance [7]. The presence of yellowish pin head-sized spots on the surface of lesion, is a good indicator of a possible diagnosis of rhinosporidiosis [8].

It is hyperendemic in Sri Lanka and Southern India, with the hot tropical weather favoring the infection [9]. The most probable mode of infection is through transepithelial route via traumatised nasal epithelium [10].

Diagnosis is easier at nasal sites because of typical features, but becomes difficult when lesions involve extranasal sites. In doubtful cases, history of bathing in stagnant water gives an indication and histopathological examination of the excised mass confirms the diagnosis [11]. Contaminated water coming in contact with traumatised epithelium is the commonest mode of spread of infection with highest incidence of cases reported among river-sand workers in India and in Sri Lanka [12]. Epithelial abrasions caused by sand particles facilitate the spread. Inhalation of field dust contaminated by the spore bearing feces of infected animals can also spread the infection [13].

For confirmatory diagnosis, histopathological evaluation of excised tissue is carried out. Rather than the stromal and cellular responses of the host, the identification of pathogen in its different stages in resected tissue is indicative of diagnosis. Most typical finding is of thick-walled sporangia containing numerous endospores within fibrovascular stroma [7,8]. Spontaneous regression is rare. Most effective mode of treatment remains surgical. Rhinosporidial lesions may recur years after primary excision [14]. Total excision of the mass preferably by electrocautery is recommended to reduce the chance of recurrence. Postoperatively, oral Dapsone is recommended by some authors as a measure to prevent recurrence [6,15].

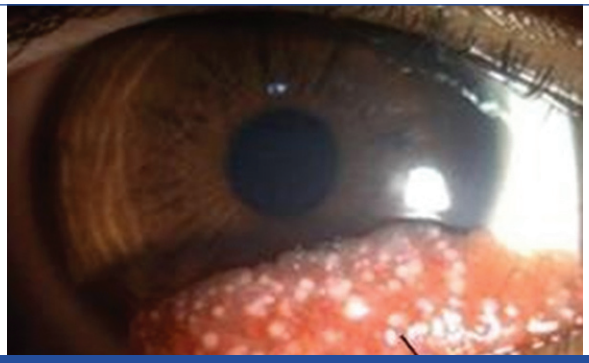
In Southwestern India, largest case series of 462 cases was reported, 81.1% cases were found to involve the nose and nasopharynx while only 14.2% cases affected the eyes [16]. A case involving multiple mucous membranes has also been reported in India [10]. There has been only few reported case series from Northern India though several reported cases have come from Southern India [4-10]. The present series highlights the importance of histopathological evaluation of all conjunctival lesions which helps in the proper diagnosis of the cases and its appropriate management keeping in mind a high suspicion of rhinosporidiosis in all such lesions.

CASE SERIES

This present case series comprises, 15 patients who visited the Department of Ophthalmology of the hospital, over a period of six months. The diagnosis was based on clinical features and results of examination of biopsied tissues samples.

All patients presented with vascular flat or elevated, fleshy, pendunculated or sessile papillomatous lesions with yellowish nodule on surface of conjunctiva and eyelid. They were admitted after routine examination and COVID-19 screening. All conjunctival growths were excised and the base was cauterised under general and local anaesthesia. Patients who were <10 years, uncooperative were subjected to general anaesthesia and others were operated under local anaesthesia.

There were 9 males and 6 females. The youngest patient was a male aged 6 years and the oldest one also was a male aged 50 years. The palpebral conjunctiva was involved in 11 and lacrimal sac in one patient [Table/Fig-1]. The most common site of infection was the lower tarsal conjunctiva (53.3%) followed by upper tarsal conjunctiva (26.6%) [Table/Fig-2,3]. All patients had history of bathing in the pond.



[Table/Fig-1]: Clinical picture showing papillomatous lesion with visible sporangium.

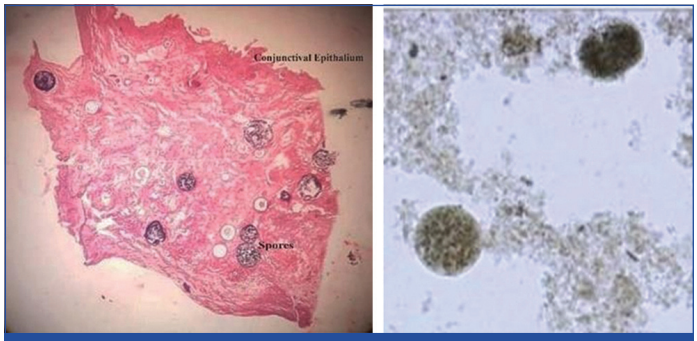
Location	No. of cases
Upper eye lid	0
Lower eye lid	1 (6.7%)
Upper tarsal conjunctiva	4 (26.6%)
Lower tarsal conjunctiva	8 (53.3%)
Bulbar conjunctiva	1 (6.7%)
Lacrimal sac	1 (6.7%)

[Table/Fig-2]: Location of ocular rhinosporidiosis.

[Table/Fig-3]: Various ocular presentations: bulbar, lower tarsal, upper tarsal.

All resected samples were sent for Histopathological Examination (HPE) and for microbiological work-up for supportive evidences. Collected tissues were processed and wet mounts were prepared in 10% KOH (potassium hydroxide) solution. These preparations were studied under (40X) microscope. Diagnosis was done by HPE and microbiological evaluation. HPE with Haematoxylin and Eosin (H&E) stain showed very vascular fibromyxomatous connective tissue covered by stratified squamous epithelium with pathogen in its diverse stages [Table/Fig-4].

Examination of Potassium Hydroxide (KOH) mount of resected tissue revealed the presence of multiple sporangia filled with endospores in various sizes and stages of development of the pathogen [Table/Fig-5].



[Table/Fig-4]: Showing conjunctival cellular infiltrate with spores (H&E,40X).
[Table/Fig-5]: Showing numerous sporangia containing endospores (KOH,100X).
(Images from left to right)

Postoperatively, all patients were advised tab. amoxycillin clavulanic acid (AmoxiclavR), paracetamol in calculated dose with topical antibiotic and lubricant drops for one week. Follow-up schedule was day 1, 7 then three monthly for one year. No recurrence was seen till 12 months follow-up. Visual acuity was not affected, and there was no visual axis involvement in any patient.

DISCUSSION

Rhinosporidiosis is four times more common in males. Usual age of presentation is between 10-40 years of life. Infection of nose and nasopharynx is most common site of presentation (70-85% cases) followed by eye (9-15%), penile urethra, external ear and bones [17,18]. There are various theories on mode of spread of infection, exact nature is unknown. One possible mode can be via migrants from endemic areas [11-13]. Lesions are always associated with the presence of the pathogen. It is believed that transmission is through discontinuous nasal mucosa. The mode of spread to other sites is via autoinoculation, another mode of ocular spread to is via lacrimal sac to plica of conjunctiva [11].

In the eye, conjunctiva (50-77.6%) and lacrimal sac (24-33%) are the most commonly affected areas. In conjunctiva, both tarsal and bulbar sides may be affected, with predominance in the tarsal region [19]. Apart from this, sclera and eyelid are also affected. In index series study, all patients presented with either conjunctival mass (93.33%) with one unusual presentation of lacrimal diverticulum (6.7%). Most common site of presentation was lower tarsal conjunctiva (53.3%) followed by upper tarsal conjunctiva (26.6%), lower eyelid, bulbar conjunctiva and sac (6.7%). These presentations were in accordance with literature presentation [4-6].

All patients in the present series had history of bathing in pond. The stagnant water, the chemical constituents and synergistic relation with other microbes may be the reason for persistence of this parasite [13]. The children were predominantly affected in the study group. Only one adult case was detected, rest all cases were below 16 years. Male predominance was which matched with previous studies [1,12-19].

The definitive diagnosis was made by HPE on biopsied or resected tissues, with the identification of the pathogen in its diverse stages, with the stromal and cellular responses of the host. The growth consists of very vascular fibromyxomatous connective tissue. It is covered by stratified squamous epithelium [12]. In all the cases, the diagnosis was confirmed using HPE [Table/Fig-4,5] and KOH mount.

Medical therapy against rhinosporidiosis is not effective. Surgical therapy is the mainstay of treatment in this disease. The surgical outcome in case of lacrimal sac involvement ranged from 28.5-92.3% [13,14]. In addition to surgical therapy, medical therapy with dapsone can be started. It inhibits the spores, arrests maturation

of sporangia and accelerates their degenerative changes. It also inhibits the uptake of para-aminobenzoic acid by the microorganism by competitively inhibiting the enzyme, folate synthetase impairing Deoxyribonucleic Acid (DNA) synthesis. It causes shrinkage and fibrosis of lesion [14,15]. Antiseptics like cetrimide-chlorhexidine, povidone-iodine, and silver nitrate solutions show some anti-rhinosporidiosis activity [16,17]. In the present series, no medical therapy was used.

Recurrence of the growth after surgical removal is a common feature, as it is not possible to eradicate entirely the subepithelial extensions [12]. In this series, none of the patients have recurrence after one year of follow-up. Histopathologically, edges were free of the disease in all cases. Complete excision of the mass along with electrocautery of base is recommended. Cases of spontaneous regression have been reported in literature but they are rare. Postoperatively, oral dapsone is recommended by studies [20].

The cases must be differentiated from papilloma, haemangioma, pyogenic granuloma, various arteriovenous malformation etc., [10]. The distinguishing feature of rhinosporidiosis is pale yellow spherules [11]. This relatively rare disease requires a high degree of suspicion, especially in endemic regions. Indian subcontinent is reported to have highest number of cases Europe and North America also have few foci [11].

CONCLUSION(S)

Ocular rhinosporidiosis is relatively rarely diagnosed case, often goes undiagnosed or under reported also. This disease is endemic in few countries, although sporadic cases are found across the world. In case of nasal rhinosporidiosis, which is more common, ocular involvement should always be looked for. While dealing with any polypoidal mass of conjunctiva, oculosporidiosis should be considered as differential diagnosis, irrespective of topo-geographical presentation. In most of the cases, clinical features suggest diagnosis, though histopathology and microbiological evidence is required to exclude mimickers. Surgery is the main treatment modality. This is a curable disease with minimum recurrence postsurgery, assuming complete excision with wide base cauterisation has been done. Regular follow-up is required to detect recurrence in case of microscopic residual disease.

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