

## Structural and Functional Outcome of Scleral Patch Graft for Ciliary Staphyloma in Rhinosporidiosis

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### Abstract

A twenty year old female presented with mass in left eye, gradually increasing in size with diminution of vision since last 2 years. There was history of conjunctival mass excision from the same eye 16 years back. The visual acuity was 20/2000 with a large ciliary staphyloma of 11 X 9 X 4 mm superiorly with adjacent flat, fleshy conjunctival mass. The mass biopsy was indicative of rhinosporidiosis. A tectonic scleral grafting with complete removal of the conjunctival mass was done. Over 10 months of follow up, the visual function improved with partial regression of the staphyloma. Staphyloma formation is rare manifestation of rhinosporidiosis. Scleral grafts are readily available with us and hold good strength. The covering of thinned area contributes to scleral re-epithelization and thus helps in partial regression of staphyloma as well as infection control. Thus, scleral grafting is an effective method for management of large anterior staphylomas for preserving ocular integrity and giving reasonable cosmeses.

**Keywords:** *Staphyloma; Anterior Staphyloma; Rhinosporidiosis; Scleral Patch Graft*

### Introduction

Rhinosporidiosis is a chronic granulomatous infection affecting the mucosal tissue layer and commonly manifests as vascular soft polypoidal pedunculated or sessile mass. The most common sites of infliction are the Nose and nasopharynx in almost 70% cases. In the eye, conjunctiva and lacrimal sac are most commonly involved. Ocular involvement is seen in 15% of the total cases. If involvement of the eye and adnexa occurs it is called oculosporidiosis [1].

The causative organism is *Rhinosporidium seeberi* which is a unique unicellular pathogen belonging to a class of organism known as micrometazoa, which are neither fungi nor protozoa. They live in ponds and lakes and infect the nasal and oral cavities of human and other animals which come in contact with contaminated water. Developing countries, with poor hygiene and use of same water source for animals and humans, like India, Sri Lanka and Pakistan account for more than 90% of the cases [2].

The lesion presents as fleshy, vascular, pedunculated or polypoidal mass, which often bleed on touch. The pathognomonic feature is presence of small grey white spores within the mass. The incidence of scleral involvement in rhinosporidiosis is very rare and presents as ectasia and staphyloma formation.

This article describes the use of donor scleral patch graft as method to preserve the structural and functional integrity of eyes with large anterior staphyloma secondary to rhinosporidiosis.

## Case Presentation

A 20 years age old girl with no contributory medical illness presented with mass in left eye, gradually increasing in size with diminution of vision since last 2 years. There was history of excision of a mass from conjunctiva of the same eye 16 years back but no details were available. She gave a history of frequently bathing in public ponds which were frequented by cattle. There was negative history for recurrent attacks of pain and redness, watering, diplopia, trauma or any intraocular surgical procedure.

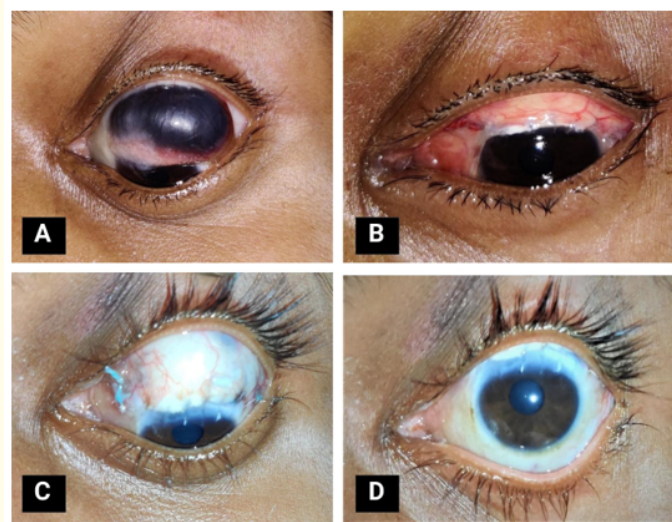
The best corrected visual acuity (BCVA) for left eye was 20/2000. There was mild restriction of the movement of eyeball superiorly with hypotropia because of the staphyloma. Rest of the ocular movements were full, free and painless. Superiorly there was a ciliary staphyloma measuring 11 X 9 X 4 mm. Superior limbus was also involved. Rest of the cornea was clear. The overlying conjunctiva showed a sessile mass with multiple pale, yellowish white dots over it. Rest of the anterior segment was within normal limits. Choroidal folds could be appreciated in the fundus. The eye was slightly hypotonous on digital tonometry with an intraocular pressure of 8.5mm Hg by non-contact tonometry. Lacrimal involvement was not noted and nasolacrimal duct was patent on syringing. Right eye examination was normal.

Otorhinolaryngology consultation was done to rule out the involvement of nasal and nasopharyngeal complex. No lesions were seen. Autoimmunity was ruled out by lack of any significant history or related ocular or general examination finding.

Since no other cause for staphyloma was found and there was presence of typical conjunctival mass with pinhead spots, a diagnosis of ocular rhinosporidiosis was made. Conjunctival biopsy was taken. Histopathological examination of the excised conjunctiva showed thick walled sporangia of large size (100 - 250 $\mu$ ) with mixed inflammatory cells which confirmed the diagnosis.

Complete excision of conjunctival mass with tectonic scleral patch graft was planned.

Intraoperatively, conjunctiva excision was done with a 1.5 mm margin all around except at the limbus where it was cut. A fornix based conjunctival flap was raised to be used later. A 13 x 11 mm size of scleral graft was used and tightly sutured onto healthy sclera with 5-0 nylon. The corneal epithelium was scraped superiorly and the scleral graft slightly flanged over the superior cornea. The loosened conjunctiva was then hooded onto the scleral graft and sutured with 8-0 vicryl. Postoperatively, patient was put on topical prednisolone acetate 1%, antibiotics and cycloplegics. As it has been proven effective earlier, oral dapsone 100 mg for 6 days in a week for 6 months was given. She was reviewed at the end of the week 1 and monthly thereafter till 3 months. The last follow up was at 10 months postoperative period. At the third month review, there was satisfactory cosmeses with BCVA improved to 20/125. There was decreased palpebral fissure of left eye. Resolution of the choroidal folds was noted. The staphyloma showed reduction in height. There was no recurrence of rhinosporidiosis (Figure 1).



**Figure 1:** (A) Superior perilimbal area showing ectatic sclera measuring 11 X 9 X 4 mm with incarceration of uveal tissue with multiple small, discrete, yellowish white mucoid spots on the surface of overlying conjunctival mass. (B) Post-operative day 1 showing the conjunctival hooding over the graft with vicryl 8-0 used for suturing. (C and D) Post-operative 3 months and 10 months showing good graft uptake and vascularization. There is reduction in height of staphyloma with dissolution of vicryl sutures.

### Discussion

As previously documented in multiple studies, *Rhinosporidium seeberi* is generally acquired by bathing in ponds contaminated by animal faeces in warmer climates, but still there is no proven theory about the complete life cycle of the organism [3-5]. The direct contact of conjunctiva to water during bathing lead to its infection. Age range of 3 and 39 years has shown higher prevalence of oculorhinosporidiosis but most cases are sporadic [4]. Our patient gave a typical history of stay in endemic area, young age and history of bathing in ponds.

Little is known about the pathogenesis of the disease but the organisms are seen to elicit specific antibodies in humans at 37 degree Celsius in *in-vitro* studies. The *in vitro* production of extracellular proteases and lipases has been postulated to play a role in the limited virulence of these organisms [6]. The proteases and protease-inhibitors system has to stay balanced to maintain homeostasis at tissue and cellular level. But in higher concentrations these enzymes have the potential to cause tissue injury as they catalyze hydrolysis of certain peptides in target proteins [7]. Histopathological picture of oculorhinosporidiosis shows neutrophils, lymphocytes, plasma cells, and multinucleated giant cells present in the submucosal layer underlying normal columnar or squamous epithelium. Often there is hyperplasia of Papillomatous nature and increase in vascularity in these lesions [3]. Hence, we postulate that the release of proteases from the overlying conjunctival rhinosporidiosis might be the cause of scleral ectasia in the patient.

Over the years, fascia lata, cartilage, synthetic Gore-Tex, skin, amniotic membrane, cadaveric aortic tissue, tibial periosteum, autologous sclera, and homologous sclera have all been tried as scleral patch grafts [8-12]. In a case similar to ours, Jacob., *et al.* performed a tectonic corneal graft over the ectasia. They concluded that cornea does not need to be covered, even with active inflammation and this was a great advantage [13].

We decided to use donor sclera for its strength, easy handling and ready uptake by the host with little inflammatory reaction and rare rejections. Many studies have proven its benefit in scleral ectasias [14,15]. Avascularity of the graft may lead to its necrosis or melting. If it is not firmly secured, dehiscence and postoperative endophthalmitis may occur. A conjunctival flap or amniotic membrane is hence recommended by many surgeons to help epithelization and vascularization of the graft [9,16,17]. Thus, we took the decision for hooding with conjunctiva.

In the postoperative period, the eye recovered with good cosmeses and no signs of recurrence of rhinosporidiosis or graft failure and decreased height of the staphyloma. This reduction was attributed to scleral fibre rearrangement and re-epithelization. Hence we propose scleral patch graft with conjunctival hooding as a good method for large staphylomas secondary to rhinosporidiosis.

### Conclusion

We recommend the use of donor scleral patch graft as method to preserve the structural and functional integrity of eyes with large anterior staphyloma secondary to rhinosporidiosis.

### Disclosure

No financial interest or any conflict of interest exists.

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