CASE REPORT

A RARE CASE OF CONJUNCTIVAL RHINOSPORIDIOSIS MIMICKING A NEOPLASM IN GHANA.

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Abstract

Introduction: Rhinosporidiosis has not been reported in the literature, in Ghana. It is however endemic in India, mostly presenting as tumorous growths within the nasal cavity and conjunctiva. Rhinosporidiosis is a chronic granulomatous infection caused by *Rhinosporidium seeberi* an organism initially thought to be a fungus, but now newly classified as belonging to a group of fish parasite DRIP clade (Mesomycetozoa) located in the middle (-meso) of the animal(-zoea) and fungal(-myceto) divergence. It has been proposed to be acquired by direct inoculation of traumatized mucosal membranes through contact with contaminated water bodies with aquatic animals as natural hosts. It usually presents as a red granular polypoid mass which may mimick a neoplasm.

Case Presentation: An 8-year-old boy was seen at the Eye clinic with a four-week history of a painless right palpebral conjunctival growth which progressively increased in size over the period. It was excised because it was suspicious for a neoplasm (Papilloma). Histopathological examination confirmed conjunctival Rhinosporidiosis. We report the clinical and histopathological findings of this entity and review the existing literature.

Conclusion: Conjunctival Rhinosporidiosis although rare occurs in our setting. *Rhinosporidium seeberi* is found in river bodies within our communities and Health care givers should be aware of this infection, which may mimick a neoplastic lesion. It should be included in their differential diagnoses of individuals who present with polypoid conjunctival and nasal masses, who hail from areas with stagnant freshwater bodies in which they routinely bathe or swim.

Key words: *Conjunctival Rhinosporidiosis, Papilloma, Rhinosporidium seeberi, Mesomycetozoa, Ghana*

Introduction

Rhinosporidiosis is a chronic granulomatous disease affecting the mucous membranes with common sites being the nose and nasopharynx (81.1%), and the eye (14.2%). Rare sites of infection include the penis, lips, skin, and uvula[1-6]. Rhinosporidiosis involving the eye is known as Oculosporidiosis[3] and the commonest sites reported in the ocular and adnexal region are the conjunctival surface (90%)[7,8] followed by the lacrimal sac (5-24%)[7-10]. The condition mostly affects males between the second and fourth decades of life[7].

The disease occurs in the Americas, Europe, Africa, and Asia but is most common in the tropics with the highest prevalence in Sri Lanka[9]. A survey of school children from Pallam, India found 11 cases in 781 children (prevalence of 1.4%)[11]. In Africa, cases of oculosporidiosis have been reported in South Africa, Malawi, Zambia, Kenya, Tanzania, Congo, Ivory Coast, and Cameroon with no report in the literature from Ghana[1]. Studies have linked infections to swimming or bathing in freshwater ponds, lakes, or rivers[11,12].

The causative agent of the disease, *Rhinosporidium Seeberi*, is believed to be an eukaryotic parasite that continues to elude growth in culture[13,14,15]. *R. Seeberi* was considered a fungus although it was originally thought to be a protozoan parasite[12] but is now classified under the class Mesomycetozoa, using ribosomal DNA analysis[7]. This class is in the middle (meso-) of the fungal (-myceto) and animal (-zoa) divergence with its morphological characteristics resembling those of *Coccidioides immitis*[1,9]. Both organisms have mature stages that consist of large, thick-walled spherical structures containing smaller daughter cells (endospores)[9]. *R. seeberi* is visualized

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with fungal stains such as methanamine silver and periodic acid-Schiff as well as mucicarmine. [9].

R. seeberi begins its life cycle as a parasite measuring 8µ and grows by nuclear division until it reaches a size of about 200 to 300µ containing at this stage, over 4,000 nuclei, which form up to 16,000 spores[16]. The mature parasite, now called Sporangium, has a double-contoured chitinous envelope with a germinal spore through which the spores are discharged[16]. Each spore subsequently develops into a separate Sporangium[16]. This organism occurring in the eye may give rise to oculosporidiosis, as its manifestations in the eye and its adnexa are more profound than those in the nose (rhinosporidiosis)[16].

Transmission of the disease occurs through traumatized epithelium (transepithelial infection), coming into contact with contaminated water and/or soil[17,18]. Patients subsequently develop a mass with polypoid morphology, associated with bleeding, pruritus, and sneezing for nasal involvement[19,20]. Clinically, Rhinosporidiosis is a slow-growing, tumor-like mass (polyp), usually of the nasal mucosa or ocular conjunctivae. It is friable and causes a foreign body sensation, with irritation and watering from the eye with no effect on visual acuity according to some isolated case reports[7]. Diagnosis is established through histopathology with the demonstration of granulomatous chronic inflammation with stromal fibrosis around several thick-walled sporangia at different stages of maturation and containing numerous endospores[1,9]. The differential diagnoses include papilloma and pyogenic granuloma.

Case Report

An 8-year-old boy, who hails from a coastal town and is a usual swimmer in the community stagnant freshwater pond presented to the Eye clinic with a four-week history of a painless right palpebral (upper eyelid) mass which progressively increased in size over the period with an associated sensation of a foreign body in the eye resulting in tearing and mild discharge. It started as an itch with no associated loss of vision. Routine laboratory investigations revealed only microcytic anaemia and his medical history was otherwise unremarkable. A diagnosis of conjunctival papilloma was made, with surgical excision of the mass for histopathological analysis. We received a tan polypoid mass in formalin measuring 13x10mm at the histopathology lab. All the specimen was processed for examination. Haematoxylin and eosin-stained sections showed a partly ulcerated lesion covered by residual stratified squamous epithelium in areas. The underlying stroma was laden by heavy mixed inflammatory cells predominantly lymphoplasmacytic in nature and surrounding numerous thick-walled sporangia containing nucleated basophilic endospores. In a focus, few of the sporangia had ruptured with spilling of endospores into stroma inciting a neutrophilic abscess reaction. These are captured in ***figure 1a*** to ***figure 1h*** below. A Periodic Acid Schiff stain (PAS) was done to outline the cuticle layer of the sporangia and confirm the histologic diagnosis of Rhinosporidiosis as shown in ***figure 2.***

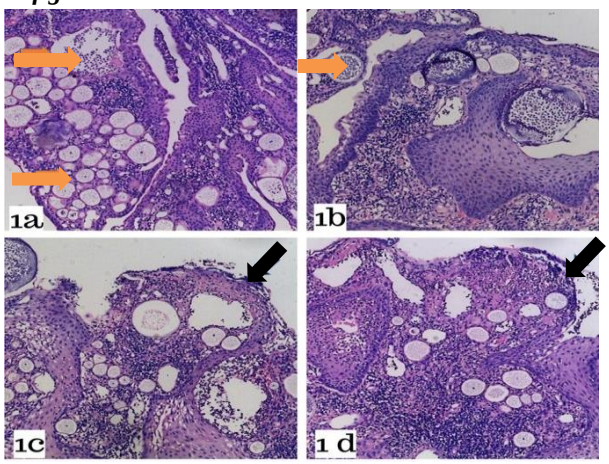

Figure 1a and Fig 1b *- (H&E x 100) - Numerous thick-walled spherical sporangia (orange arrows) containing endospores with surrounding dense lymphoplasmacytic infiltrate.*

Figure 1c *and* ***Figure 1d*** *– (H&E x 100) –Partly ulcerated surface epithelium with focal granulation tissue (black arrows).*

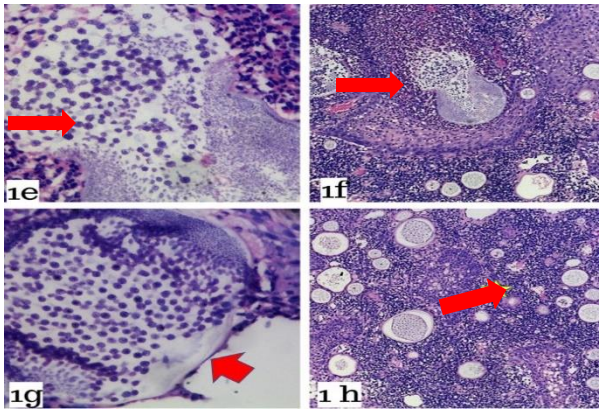

Figure 1e *and* ***Figure 1f****- (H&E x 100&400) - Ruptured sporangia (long arrow) with spilling of endospores into stroma inciting neutrophilic abscess (red arrow).*

Figure 1g *– ((H&E x400) – Sporangium containing numerous endospores located within the surface epithelium.*

Figure 1h *- (H&E x 100) - section shows lymphoplasmacytic infiltrate around sporangia. (Red arrow)*

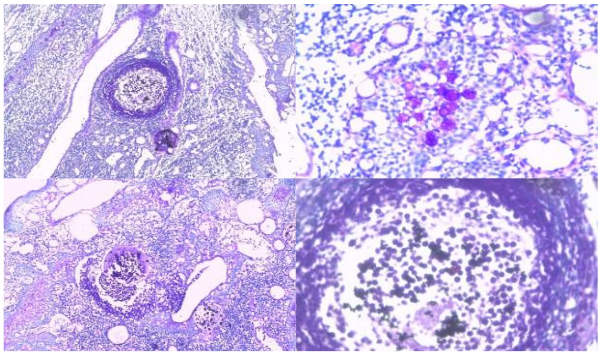

Figure 2: *PAS-stained slides showing various stages of Rhinosporidiosis*

Discussion

Rhinosporidium Seeberi is the identified etiologic agent of Rhinosporidiosis, a mucosal chronic granulomatous inflammatory disease transmitted by bathing in stagnant ponds in which infected animals also bathe. Our patient frequently swims in a stagnant water body in the community. There was however no prior significant history of trauma to the right eye.

The life cycle of *R. Seeberi* has 2 phases, invitro and in human host. It develops spherical cysts with endospores (stage 1). In vitro the released endospores (stage 2) give rise to uniflagellated infectious zoospores (stage 3). When the zoospores (infecting units) infect the host, they encyst (stage 4) and increase in size (stage 4, 5) and undergo cleavage into endospores (stage 1). The endospores can also be directly released within the host's tissues when the sporangia rupture and the cycle is repeated inside the host (stages 1, 4, 5, 1). A pictorial representation of the life cycle of *Rhinosporidium Seeberi* is illustrated in (***figure 3)***. This results in the expanding tumorous lesion with the associated chronic inflammatory changes. This was initially diagnosed in our patient as a conjunctival papilloma.

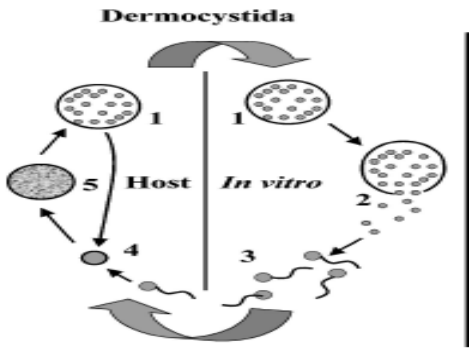

Figure 3*- Depiction of the putative life cycle of members of the orders Dermocystida, from***: *https://doi.org/10.1146/annurev.micro.56.012302.160950*

Clinical features depend on the site of involvement. The lesions are usually seen in the nasal cavity and nasopharynx in majority of patients. These are reddish polypoidal, bulky, friable mucosal masses. Conjunctiva, mouth, larynx, genitalia and skin are the other rare sites of involvement.[1-8,24] For our case we were unable to get a gross photo of the eye lesion before excision. The lesion was initially diagnosed as a papilloma a rather common benign tumor seen in the eye clinic. Our patient presented with a four-week history of foreign body sensation, a swelling of the eye with associated tearing and mild discharge. Clinical presentation however depends on the presence of a tumorous mass at the locations stated early on.

Microscopy showed characteristic morphology, spherical sporangia of various sizes with heavy lymphoplasmacytic infiltrate located in the stroma of the polyp. The overlying mucosa may show focal ulceration or metaplastic changes depending on the site. The largest of the sporangia are usually immediate subepithelial in location which may or may not show evidence of rupture. For our case, there was focal epithelial ulceration as well as several subepithelial and intraepithelial sporangia containing numerous endospores within a stroma laden with heavy lymphoplasmacytic infiltrate. In areas, the sporangia were ruptured releasing endospores into adjacent stroma with surrounding abscess formation **(*figure 1e)***. Periodic Acid Schiff special stain highlights the thick cuticle cyst wall of sporangia and the round nucleated basophilic endospores (***Figure 2***).

The treatment of choice is usually excision of the lesion. However, excision with local cautery is considered the most effective treatment[22,24]. Our patient had excision of the lesion and was given DEXATROL (dexamethasone neomycin polymyxin b) eye drops to be reviewed in a month. Cautery is used to reduce the risk of recurrence which is caused by endospores being released into nearby mucosa. Medical treatment with dapsone and amphotericin B remains controversial, as determining drug sensitivity has been impossible without the ability to grow R. seeberi in vitro. However, dapsone has had some success in treating Rhinosporidioisis; it is believed to act by arresting sporangia maturation and promote fibrosis[7]. Recurrence rates have been noted to vary by infection site with conjunctival and lacrimal sac recurrences being relatively low compared to nasopharyngeal recurrence rates which have been reported in up to 10% of patients[9,21]. An important ocular complication is the formation of staphyloma, which occurs due to scleral

thinning and herniation of the intraocular content[7]. This can lead to rupture and loss of the intraocular content.

Conclusion

Conjunctival Rhinosporidiosis although rare occurs in our setting. It is found in river bodies within our communities and Health care givers should be aware of this infection, which may mimick a neoplastic lesion. There should be a high index of suspicion in individuals who present with polypoid conjunctival and nasal masses, who hail from areas with stagnant freshwater bodies in which they routinely bathe or swim. Routine histopathological examination of all conjunctival lesions in these areas could confirm the endemicity of this condition.

Ethical Considerations

Ethical review is not required for reporting cases in our institution. Informed consent was however sought from the patient's guardians and institutional assent was obtained from the Cape Coast Teaching Hospital.

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