Rhinosporidiosis – unusual presentations

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Abstract Rhinosporidiosis is an infection caused by *Rhinosporidium seeberi* that frequently presents as polypoidal nasal lesions. Here, we report two cases of rhinosporidiosis with unusual presentations. The first case presented in our department with chronic dacryocystitis of left side for endoscopic dacryocystorhinostomy (endoscopic DCR) operation. The second case presented as a long hanging mass arising from the right side of nasal septum. The diagnosis was established on the morphological basis by the identification of endospores and sporangia. The clinicopathological and immunologic features were discussed and the literature was reviewed.

Keywords *Rhinosporidium seeberi* · Rhinosporidiosis · Lacrimal sac · Endoscopic DCR

Introduction

Rhinosporidiosis was previously thought to be a fungal infection caused by *Rhinosporidium seeberi*. However recent evidence suggests that *R. seeberi* phylogenetically appears to belong to a number of so called DRIPs, clade of fish parasite [1], which affects not only the human but also animals. This disease is endemic in India and Sri Lanka. It is a chronic disease striking selectively two different target zones: nasal and conjunctival mucosa. In tissues, the life cycle of this organism starts with a 6–10 µm round endospore. The endospore grows to become a thick-walled sporangium of 150–350 µm size containing thousands of endospores. Clinically, these lesions usually present as polyps, tumours, papillomas or warty lesions in the nose, conjunctiva or the urethra. They are soft, hyperplastic and highly vascular and they can be either sessile or pedunculated. Other clinical presentations include osteolytic bony lesions in the disseminated form of disease [2].

The disease is treated by surgical excision combined with electro-coagulation of the base of the lesion. Dapsone therapy was considered in recurrent cases. Complications of the disease include extremely rare life-threatening dissemination, local secondary bacterial infection, and recurrence [3].

Case report

A 45-year-old male patient from rural background presented to the outpatient clinic at Medical College and Hospital, Kolkata with history of epiphora and purulent discharge from the left eye of three months duration. Clinical examination revealed presence of a fluctuant swelling at medial canthus of the left eye (Fig. 1). On pressing the swelling, mucoid secretion came out of the medial canthus. Clinically, rhinosporidiosis was not suspected. Endoscopy did not reveal any abnormality of the nasal cavity. Chronic dacryocystitis with mucocoele was considered as the clini-

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Fig. 1 Clinical photograph of lacrimal sac rhinosporidiosis of left side

cal diagnosis and endoscopic dacryocystorhinostomy was performed. On removing the ascending process of the maxilla and the adjacent bones, the lacrimal sac herniated into the nasal cavity. On excising the medial aspect the sac wall, mucosa appeared to be polypoidal, pale and reddish in colour. The sac wall was sent for histopathological examination. Histological examination (PAS stain) revealed the presence of typical features of rhinosporidiosis (Fig. 2). After receiving the histological diagnosis, patient was subjected for dacryocystectomy operation for total clearance of disease process.

A 18-year-old young man from rural Bengal presented to our OPD with atypical long mass coming out of right nasal cavity, which he could blow out of the nose and again take back inside the nasal cavity on his own wish. The mass was long, pedunculated and reddish studded with white dots (Fig. 3, Fig. 4). It was arising from the right side of nasal septum. It was endoscopically removed and cauterization of the base was done. Histopathologically the mass was diagnosed as rhinosporidiosis and 3-year follow up of the patient did not show any recurrence.

Discussion

Rhinosporidiosis in humans presents several enigmas that have yet to be explained: (a) the chronicity of the disease that could last for several years, (b) recurrence after surgical

Fig. 2 Histopathological microphotograph of rhinosporidiosis (100X, PAS stain)

Fig. 3 Clinical photograph of Patient No. 2

excision of the initial polyps that most commonly occur in the anterior respiratory passages, and (c) occasional dissemination from these sites to the limbs and viscera. (Malignant rhinosporidiosis) [4].

Fig. 4 Clinical photograph of Patient No. 2

According to the site of lesion, rhinosporidiosis are divided into following categories:

1. Nasal, 2. Nasopharyngeal, 3. Mixed and 4. Bizarre which includes conjunctival, tarsal and cutaneous rhinosporidiosis [4]. The nasal lesion appears as reddish, papillomatous mass resembling ripe strawberry, usually restricted to nasal cavity and extending towards nasopharynx. On MEDLINE search through PUBMED, many unusual presentations of rhinosporidiosis have been found in recent literature like- cutaneous [5], pulmonary [5], bone [6], and disseminated [5] involvement. A hanging rhinosporidial mass which patient could take back inside the nose on his own wish is an uncommon presentation, which was found in our second case. Lacrimal sac rhinosporidiosis is not an uncommon lesion and Kameswaran postulated that nearly in all cases infection ascends from the nose [4]. Once it has reached lacrimal sac the infection has a tendency to spread diffusely underneath the skin [4]. However analyzing a series of 48 cases of lacrimal sac rhinosporidiosis, Mukherjee et al observed nasal involvement only in 6 cases [7].

Characteristics of rhinosporidiosis of lacrimal sac are- diffuse, soft and non-tender swelling; limited superiorly by the roof of the orbit, medially by nasal cavity; extending diffusely under the lower lid; the skin over the swelling showing small pits, giving an appearance of orange peel; tendency to erode surrounding bone; absence of regurgitation and epiphora [7]. However, these typical features are not seen in all the cases and an otolaryngologist is prompted to perform an endoscopic DCR operation. In our first case typical features of rhinosporidiosis was absent and nasal endoscopy also did not reveal any rhinosporidial mass.

The histological features of rhinosporidiosis are typical. First, hyperplasia of the ciliated columnar epithelium covering mucosa. The hyperplastic areas have vascular fibro-connective tissue cores formed of fibroblasts, myofibroblasts and occasional inflammatory cells. Second, sporangia full of spores in different stages of development. The giant spo-

rangia characteristically have thick-walls. Third, hypervascularity in the submucosa with multiple thin walled blood vessels. Fourth, increased submucosal inflammatory cell infiltrates including neutrophils, lymphocytes, plasma cells, histiocytes and multinucleated giant cells.

Finally, foci of necrosis were seen both in the epidermis and the submucosa. Histochemical stains such as Periodic Acid Schiff and Methenamine silver were used to

establish the diagnosis. Cytological criteria for diagnosis included the presence of 5–10 μm endospores and 50–1000 μm sporangia.

Cell-mediated immune (CMI) responses were recently demonstrated by the assay of lymphoproliferative responses to rhinosporidial antigens and the T-cell mitogen. The occurrence of CMI responses was also suggested by the marked presence in rhinosporidial polyps of many plasma cells, fewer CD68+ macrophages, a population of CD3+ T lymphocytes, and CD56/57+ NK lymphocytes, which were positive for CD3 as well. CD4+ T helper cells were scarce. CD8+ suppressor/cytotoxic-cytolytic cells were numerous. Most of the CD8+ cells were TIA1+ and therefore of the cytotoxic subtype. CD8+ T cells were not sub-typed according to their cytokine profile; IL2, IFN-gamma (Tc1); IL4, IL5 (Tc2) [8]. The role of antibody mediated anti-rhinosporidial immunity has however not been investigated although high titers of anti-rhinosporidial antibody have been detected in patients with localized or disseminated disease.

The treatment modality of this chronic recurrent disease is surgery with excision of the polypoid mass with wide margin with diathermy cauterization of the base, but recurrence is very common. Endoscopic surgery has simplified the removal of rhinosporidial polyp and vastly improved the visibility inside the nasal cavity [4]. In a recent study KTP laser has been found to be a useful adjunct to surgery in comparison to conventional diathermy excision [9]. Treatment of lacrimal sac rhinosporidiosis is controversial, Kameswaran et al described surgical intervention in the lacrimal sac is uncalled for subcutaneous spread [4]. But in another study, Mukherjee et al described dacryocystectomy as the surgical management of the lacrimal sac rhinosporidiosis [7]. In a recent study, Jayant et al described endoscopic DCR with postoperative Dapsone therapy as an alternative treatment option to dacryocystectomy for lacrimal sac rhinosporidiosis [10]. No medical treatment has been found to be effective in controlling rhinosporidiosis. Drugs such as griseofulvin and amphotericin have been tried but without much benefit. Dapsone (diaminodiphenylsulfone) is a relatively safe drug to use, and no major side effects were noticed [11].

Conclusion

Rhinosporidiosis, a fungal infection due to *Rhinosporidium seeberi*, frequently produces polypoid lesions in the nose.

Sites like the conjunctiva, larynx, trachea, nasopharynx, skin and genitourinary tract are usually involved; lung and bone are involved less frequently. Rhinosporidiosis presenting as a long hanging mass and lacrimal rhinosporidiosis mimicking chronic dacryocystitis are rare presentations. Caution should be taken while dealing with this type of lesion and every bit of suspected tissue should be sent for histopathological examination.

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