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Case report

Rhinosporidiosis: Report of an extra-ductal facial lesion

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ABSTRACT

Rhinosporidiosis is a chronic granulomatous mucocutaneous disease that is endemic in South Asia. It commonly affects men in the second to fourth decade of life. The most common site of infection is the nose or nasopharynx with primary involvement of the parotid duct noted very rarely; only four cases reported in literature. We report a case of a 77 year old male patient who presented with a subcutaneous cheek swelling with no other clinical features to suggest the diagnosis of rhinosporidiosis. An unusual affliction in a patient in the seventh decade of life, at a site not reported previously with no past history of the same makes it imperative to ascertain the known facts about both the organism and the disease. However the histopathological report confirmed the diagnosis which reiterates the need to further study the pathogenesis of this condition.

Introduction

Rhinosporidiosis is a chronic granulomatous mucocutaneous disease, endemic in Southern India, that commonly affects the mucous membranes of the nose and nasopharynx with reported involvement of extranasal sites such as oropharynx, larynx, trachea, bronchi, eye, ear, skin and genitourinary tract. There have been reports of rhinosporidiosis affecting the parotid duct resulting in cystic swellings of the face. We report a case of a 77 year old male patient from Malappuram in Kerala who presented with a swelling over the left cheek. The clinical differential diagnosis did not favour the possibility of rhinosporidiosis that was confirmed postoperatively by histopathological examination of the surgical specimen. This unusual case of rhinosporidiosis presenting as a subcutaneous cystic swelling left cheek adherant to the parotid duct not actually arising from it, is first of its kind and makes it imperative to think whether there is much more to explore about this disease and organism. Furthermore our case emphasises the need for clinicians to be aware of such a possibility in endemic areas and to ascertain further facts regarding the pathogenesis of this disease.

Case report

A 77 year old male from Malappuram district of Kerala (Southern India) presented to our ENT outpatient department with a swelling over the left cheek for one month. The swelling was small to begin with, approximately the size of a marble when he first noticed it. It then gradually increased in size with no alteration in size with meals and no history of pain or difficulty in mouth opening. He did not give any history of trauma, fever or dip baths i.e bathing in nearby ponds or any waterbody in the locality. On local examination a single, soft, discrete, mobile swelling approximately 4.5×3 cm on the left side of his face was noted. It was non tender with a smooth surface and the overlying skin appeared glossy without local warmth. The intraoral examination was normal with no cervical lymphadenopathy. The examination of the ear, nose, nasopharynx and systemic examination did not reveal any other abnormalities. The routine laboratory investigations were within normal limits except for a raised ESR of 90 mm/h. Screening tests including HIV testing done were negative. Aspiration done in our procedure room revealed serous fluid which was sent for cytology and culture. Cytology showed yeast forms/spores of fungi with acute inflammatory cells in background, suggestive of fungal infection and fungal culture was advised. Fungal culture was negative.

The swelling reduced in size post procedure but within 2 weeks the patient came to the outpatient department with further increase in size of the swelling associated with pain Fig. 1. An ultrasonography done showed a hypoechoeic solid cystic lesion in the left cheek in the subcutaneous plane anterior to the masseter muscle with increased vascularity. The adjacent parotid gland

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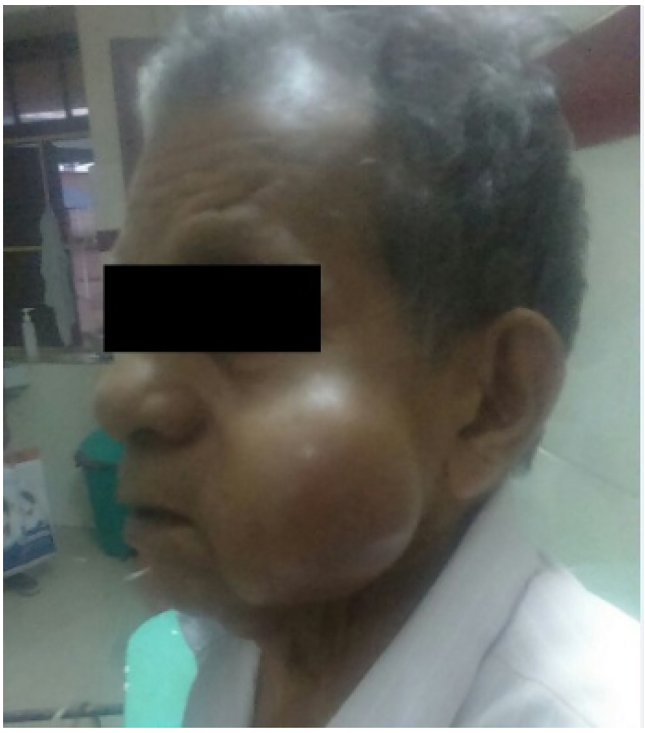

Fig. 1. Clinical presentation of our patient with a cystic swelling over the left cheek which increased in size post aspiration.

showed normal echoes. The possibility of a minor salivary gland neoplasm was suggested. Contrast Enhanced CT (CECT) scan of the face was done which showed a well defined peripherally enhancing collection in the subcutaneous plane in the left cheek with no evidence of deep extension Fig. 2. A fine needle aspiration cytology(FNAC) was carried out which reported a possibility of rhinosporidiosis. In view of this a diagnostic nasal endoscopy was done to examine the entire nasal cavity and nasopharynx which were normal with no evidence of any lesion in these areas Fig. 3.

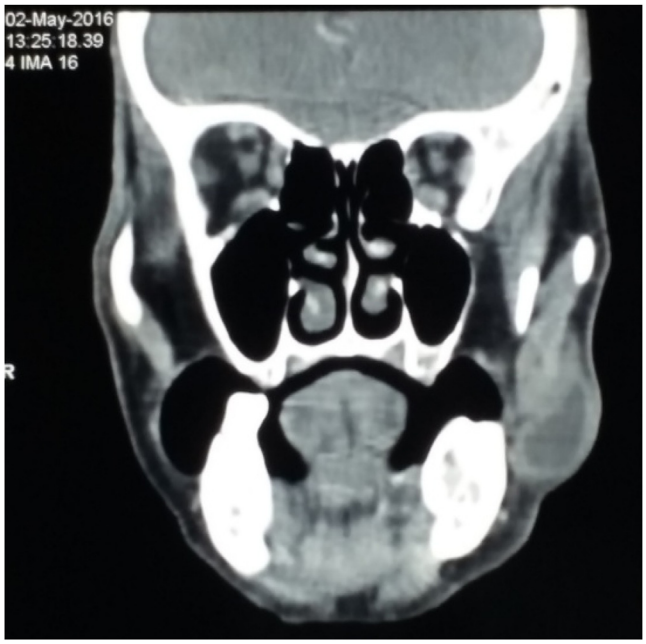

Fig. 2. Contrst Enhanced CT Scan showing the lesion in subcutaneous plane over masseter muscle(Lt).

The plan thereafter was a total excision of the lesion under general anaesthesia. In view of acquired valvular heart disease and high risk to undergo surgery under general anaesthesia the procedure was done under local anaesthesia. A wide excision was done with careful clearance of the lesion from the peripheral branches of the facial nerve. A portion of the sac adherant to the parotid duct was separated meticulously and removed in toto.

The post operative period was uneventful and patient discharged. The patient has been on follow up with no recurrence noted Fig. 4. The histopathological report confirmed the diagnosis of rhinosporidiosis Fig. 5. There have been reports of rhinosporidiosis of the parotid duct presenting as swelling of cheek but this case of a subcutaneous swelling of the cheek outside the parotid duct proper is a rare form of presentation of rhinosporidiosis; a first of its kind to be reported.

Discussion

Rhinosporidiosis caused by *Rhinosporidium seeberi* is a chronic mucocutaneous infection endemic in South Asia with largest number of cases reported in India and Sri Lanka [1]. The life cycle and taxonomy of the causative agent, *Rhinosporidium seeberi*, are incompletely understood and controversial [2]. Therefore this disease till date continues to be associated with diagnostic and treatment related challenges.

The organism *Rhinosporidium seeberi* was first described by Seeber in 1900 in the nasal region in his doctoral thesis in medicine. Initially thought to be a fungus, on further research it was considered to be a protozoan by Seeber and a phycomycete by Ashworth. An understanding of its life cycle and phylogeny has mostly been defined by observations in tissue culture, scanning electron microscopy and molecular studies. Through phylogenetic analysis of *Rhinosporidium seeberi* 18S rRNA gene, this group of pathogens was originally identified by Ragan et al. as part of the DRIP clade (acronym derived from *Dermocystidum*, *rosette agent*, *Ichthyophonus* and *Psorospermium*) [3] Herr et al replaced Rhinosporideacae with the term Mesomycetozoa (between fungi and animals) [4]. The class Mesomycetozoa has two orders, that is, the Dermocystida and the Ichthyophonida. In the order Dermocystida is the family Rhinosporideaceae which includes *Rhinosporidium seeberi*, *Dermocystidium* spp. and the rosette agent. In the order Ichthyophonida, the class Ichthyophonae has members with phylogenetic features in common with the genus *Ichthyophonus* and *Psorospermium*. The most recent classification of *R. seeberi* is a eukaryotic pathogen in the Mesomycetozoa class [10]. The mature phenotype of the organism includes a thick walled spherical structure termed sporangium containing smaller spherical structures termed sporangiospores. Microscopically, the involved tissue contains numerous sporangia ranging from 10 to 300 μm in diameter in various stages of development with the larger lesions closer to the mucosal surface. The sporangiospores are 2–5 μm in diameter. The limitation of growing the organism in the culture media is responsible for poor understanding of the pathogenicity of the organism [2,5].

The common mode of spread of infection is by transepithelial transmission or autoinoculation and is thought to be mediated by exposure to spores present in contaminated dust, soil or stagnant water. Owing to the natural aquatic habitat of the organism, a high incidence is reported in individuals who bathe in water contaminated by diseased cattle. But our patient had no history of bathing in ponds or stagnant waters and hence alternative mode of spread needed to be thought of. The pre-existing damage to the epithelium and poorly understood host factors have been implicated [5].

In patients with rhinosporidiosis of the parotid duct, passage of the spores into the parotid duct through the natural patulous

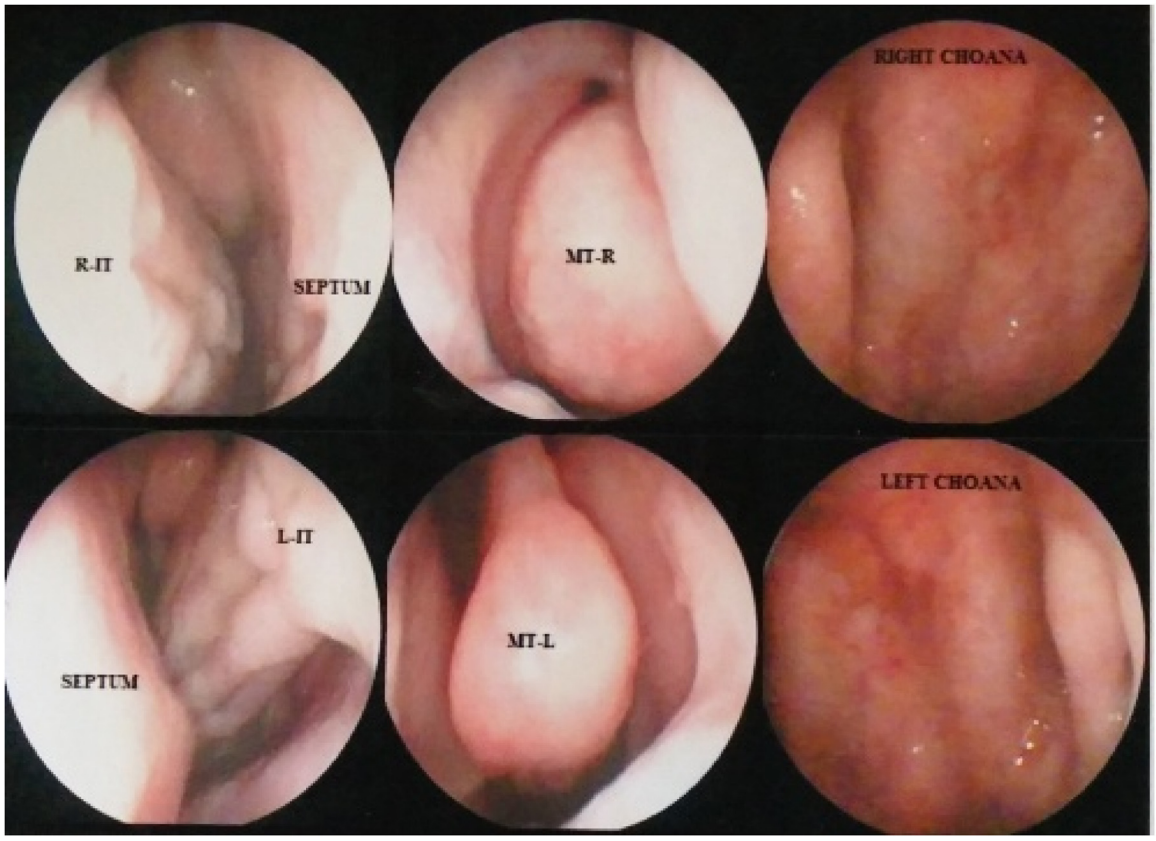

Fig. 3. Diagnostic Nasal Endoscopy of the patient that was normal with no evidence of the lesion in the nasal cavity or nasopharynx. (IT: Inferior turbinate; MT: Middle turbinate; R: Right side; L:Left side).

Fig. 4. Post operative picture when the patient came for follow up.

opening of the parotid duct from the oral cavity with subsequent replication has been thought to cause ductal stenosis and subsequently development of the cyst [6]. In our case the spore had migrated from the parotid duct to the surrounding tissues resulting in subcutaneous cyst formation. This compels us to think on the possibility of the organism being motile or possessing the potential to migrate to adjacent tissues. Hematogenous and lymphatic spread have been reported and thought to be responsible for the dissemination of the disease to anatomically unrelated sites [7]. The lesion usually presents as a friable, polypoid, exophytic mass with submucosal studding of the erythematous surface with white dots giving it a “strawberry” like appearance. It is a highly vascular lesion that bleeds easily on touch. Lesions may be associated with slow clinical progression and a prolonged incubation can be seen before initial exposure [2]. A high degree of clinical suspicion is necessary especially in patients who live in endemic areas and in patients with delayed presentation. The need for long term follow-up is also critical in this disorder given the potential for slow progression of a persistent disease [8].

The primary treatment for rhinosporidiosis is wide surgical excision of lesion with cauterization of the base to promote the destruction of all spores. Anatomic considerations of the surgical resection are based on the site of involvement. In rhinosporidiosis of parotid duct surgical excision along with ligation of the Stensen duct in cases involving the parotid duct forms the mainstay but minimally invasive surgery together with dapsone may be tried in selected cases [9]. In our patient a large cystic lesion was identified overlying the masseter muscle on the left which was dissected free from the branches of the facial nerve and a part of it adherant to the parotid duct was carefully separated ensuring total excision of the lesion. The complications of surgical therapy include recurrence at the primary or adjacent sites, bacterial superinfection and treatment related morbidity like injury to the surrounding critical neurovascular structures like the facial nerve in our case. The need for long term follow-up in this disease becomes important due to recurrences noted.

In patients with high risk of recurrence the use of dapsone following surgery is suggested as it may stop the growth cycle of

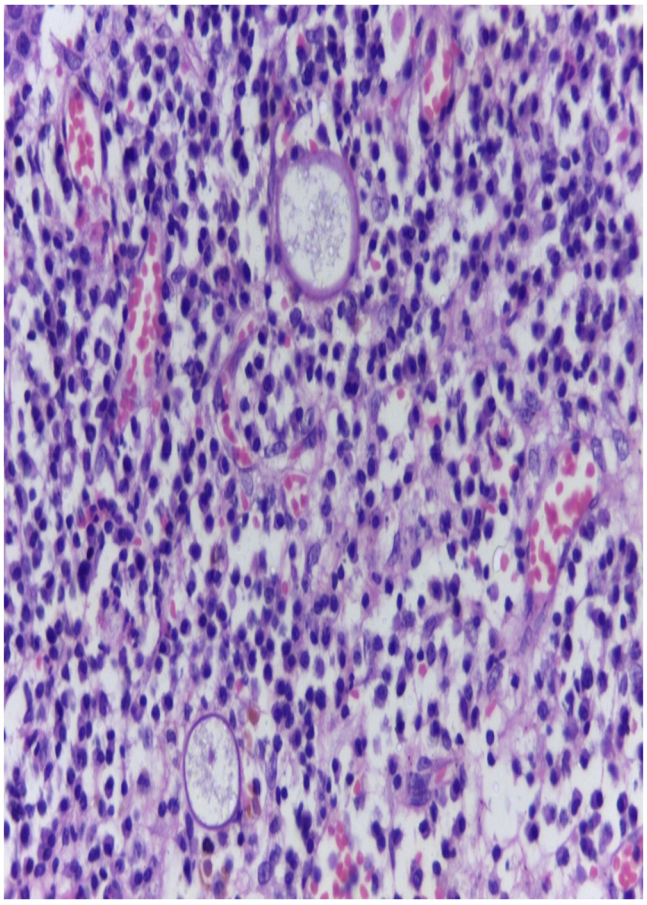

Fig. 5. Histopathological report confirmatory of Rhinosporidiosis.

the sporangia and induce fibrosis of the surrounding stroma [5]. The inability to establish experimental rhinosporidiosis and failure to propogate the organism *in vitro*, have prevented the determination of the sensitivity of *R.seeberi* to drugs that might have clinical application. While several anti-bacterial and anti-fungal drugs have been tested clinically, but unsuccessfully, the only drug which was found to have some anti-rhinosporidial effect is dapsone (4,4-diaminodiphenyl sulphone) The role of dapsone is considered to be adjunctive in cases of incomplete surgical resection and in multi recurrent or disseminated disease or in involvement of critical structures [11].

Conclusion

Rhinosporidiosis is an unique granulomatous mucocutaneous infection caused by *Rhinosporidium seeberi*. The difficulty in culturing the organism and its atypical cellular features have been responsible for its taxonomic enigma. The endemicity and rarity of this disease has further complicated matters with many questions related to it remaining unanswered to date. The organism is endemic to Southeast Asia and transmission are most commonly mediated by exposure to spores from contaminated dust, soil and water. The rare presentations of this disease as in our case reiterates the need to explore more on the pathogenesis. Endoscopic examination, imaging and histopathology are needed for the diagnosis and treatment planning. The primary treatment modality is surgical excision with cauterization of the base of the lesion. This could be challenging especially when the lesion abuts critical neurovascular structures like the facial nerve in our case. Dapsone may be used as an adjunct to surgery, in recurrent and disseminated cases; although its role is yet to be proven.

Conflicts of interest

None.

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