RHINOSPORIDIOSIS CHEEK: A VERY RARE PRESENTATION

SHAH R[1], FAKIR MAY[2], TALUKDER DC[2], KHAN AFM[3]

Abstract:

Rhinosporidiosis is a chronic granulomatous infection of the mucous membranes that usually manifests as vascular friable polyps in the nasal cavity. It frequently involves the nose and nasopharynx. But occasionally the lesions may be found in anatomically distant sites like conjunctiva, rectum and external genitalia. Traumatic inoculation from one site to others is common as in Laryngeal rhinosporidiosis. The buccal cavity (cheek) is only rarely affected. We report here a unique case of rhinosporidiosis left cheek in a 32 year old man presented as an ulcerated mass on left side of cheek.

Key words: *Rhinosporidiosis, cheek.*

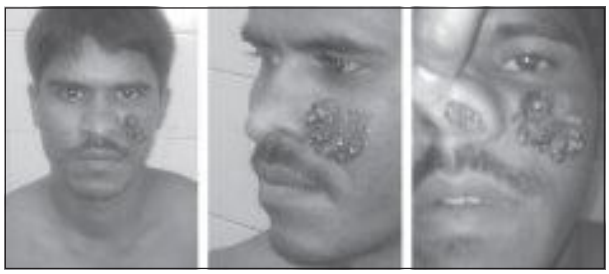
J Dhaka Med Coll. 2012; 21(2) : 242-244.

Introduction:

Rhinosporidiosis is a granulomatous disorder caused by Rhinosporidium seeberi. It is a chronic disease affecting primarily the mucosa of the nose, conjunctiva and urethra. Approximately 90% of all known cases occur in India, where the prevalence has been estimated at 1.4%[1], and in Sri Lanka. The disease has also been reported in America, Europe and Africa[2]. Except in ocular rhinosporidiosis, this has a female preponderance, rhinosporidiosis in respiratory sites shows a higher incidence in males. Infection is usually caused by contact with stagnant ground water such as that in lakes and ponds. Clinically, the disease presents as polypoid, tumour-like, papillomatous or warty lesions which are hyperplastic, highly vascularised and friable. They may be sessile or pedunculated. The common sites are the nose (the primary site of infection), the eye and its appendages, conjunctiva and the urethra. The larynx, trachea, skin and lungs are less frequently involved. The most common sites in the upper respiratory tract are the anterior nares and the inferior turbinates, the septum and the floor of the nasal cavity. Posteriorly, rhinosporidial polyps occur in the nasopharynx, larynx and soft palate. The buccal cavity (cheek) is only rarely affected. About 15% of cases of rhinosporidiosis are ocular in location, in the bulbar and palpebral conjunctiva. Osteolytic bone infiltration is another clinical presentation. Generalised rhinosporidiosis with skin and visceral involvement is extremely rare[3]. It is an infective but not an infectious, contagious or toxic disease. History and histopathology of specimen with high index of suspicion are helpful for proper diagnosis. At present, surgical excision with cauterization of base followed by a course of oral dapsone seems to have convincing result[4].

Case Report:

A 32years old manual laborer male was admitted to the dept of ENT & Head Neck Surgery, Dhaka Medical College Hospital with a painless ulcerative growth on left cheek for about 2 years & left sided nasal mass for about 9 years.

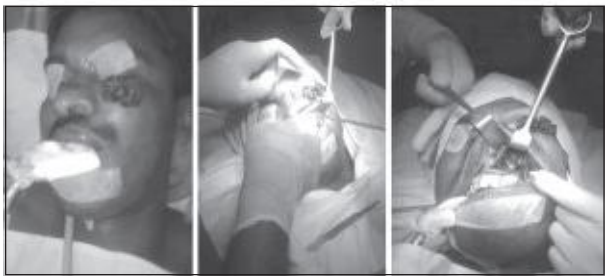


1. Dr. Rameez Shah, M.S. (Thesis Part), Dept. of ENT & Head-Neck Surgery, Dhaka Medical College Hospital, Dhaka.
2. Dr. Mohammad Abu Yousuf Fakir, Dr. Debesh Chandra Talukder, Assistant Professor, Dept. of ENT & Head-Neck Surgery, Dhaka Medical College Hospital, Dhaka.
3. Dr. A.F.Mohiuddin Khan, Professor & Head, Dept. of ENT & Head-Neck Surgery, Dhaka Medical College Hospital, Dhaka.

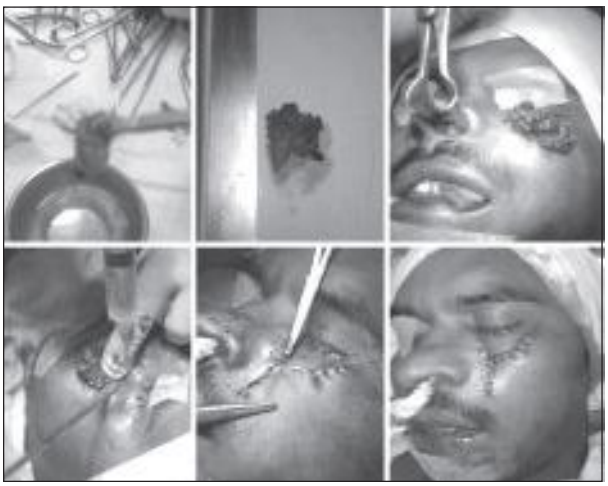
For correspondence: Dr. Rameez Shah, M.S. (Thesis Part) student, Dept. of ENT & Head-Neck Surgery, Dhaka Medical College Hospital, Dhaka. Email: rameezshah527@yahoo.com

On examination a non-tender, cauliflower-like, ulcerative growth about 3cm × 3cm was present on left cheek. Oral cavity, parotid gland, was not involved & there was no lymphadenopathy. On examination of nose, a pink colored mass was seen in left nares anteriorly, occupying the whole space, but posterior nares was not involved. Examination of ear revealed no abnormality. Overall systemic examination was normal. His blood counts, ESR, blood sugar, liver and renal function tests, chest x-ray and abdominal ultrasonography were within normal limits. CT-scan of PNS with contrast was done and was suggestive of invasive fungal infection involving the left nasal cavity extending to adjacent left cheek. Serology for HIV was negative. Patient was operated under general anesthesia.

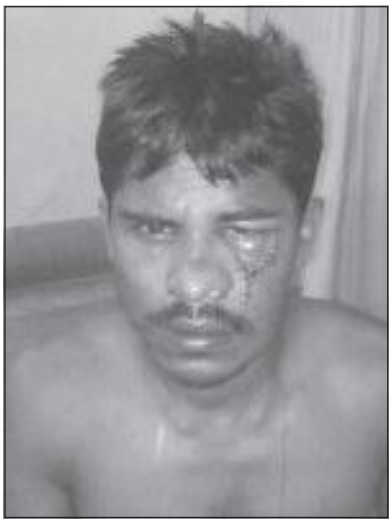
Per operative findings revealed that both the masses were separate.



The nasal mass was removed, turbinectomy was done and the growth on cheek was removed by broad base excision & wide base cauterization. Skin was apposed by 3-0 proline.



Both the tissues were sent for histopathological examination, which revealed rhinosporidiosis. Patient was given dapsone therapy (100 mg/ day), and was advised regular follow up to assess response to dapsone. There was no recurrence in six month period.



Discussion:

Rhinosporidiosis is known for over a hundred years and was first described in Argentina.[5] *Rhinosporidium seeberi* is the etiological agent. Morphologically, it resembles both fungi and protozoa, and its taxonomy has been debated for decades. The micro-organism has not been isolated or grown in microbiological culture,[6] and its taxonomic position was only resolved in 1999 when Herr et al,[7] using molecular biological techniques, definitively placed it in a new class, the Mesomycetozoea. This classification was independently confirmed in 2000.[8] Rhinosporidiosis is endemic in south Asia, notably in southern India, Bangladesh, Nepal and Sri Lanka[9]. It is commonly seen in young males. It is thought to occur through contact with stagnant water[10]. Bathing in infected ponds with local trauma resulting in abrasion to mucous membrane and skin may precipitate the initiation of the disease[11]. Transepithelial infection through traumatized epithelium is the presumed mode of infection from the natural aquatic habitat of the organism[5].Cutaneous lesions,

although rare, can occur due to autoinoculation[18] or haematogenous spread[19] and are classified as:

1. Satellite lesions around the nasal polypoidal lesions
2. Disseminated lesions with visceral involvement
3. Primary cutaneous lesions without internal organ involvement.[20]

Usually it presents with epistaxis, nasal obstruction and a mass inside in nasal cavity. The mass is leaf like, pinkish, granular with strawberry like appearance and the surface studded with whitish spots. The mass is pedunculated, friable, and bleeds on touch. Histopathology is diagnostic showing multiple budding sporangia embedded in a fibrovascular stroma infiltrated by chronic inflammatory cells.[4,13] Spontaneous regression of lesions can rarely occur, however it should be treated early in order to prevent extension of lesions or dissemination. Mode of treatment is mainly surgical. Diathermy excision is the treatment of choice, but in spite of this recurrences have occurred.[14] Total excision of the polyp, preferably by electrocautery, is recommended. The only drug that has been found to have some anti-rhinosporidial effect is dapsone (4,4 diaminodiphenyl sulphone), which appears to arrest the maturation of the sporangia and to promote fibrosis in the stroma when used as an adjunct to surgery.[15] Dapsone is usually given in an attempt to cure or prevent the recurrence of rhinosporidiosis.[16]

Conclusion:

Rhinosporidiosis is a chronic granulomatous infection caused by *Rhinosporidium seeberi*. Mucosal involvement of the nose, nasopharynx and soft palate in the form of sessile or pedunculated vascular polyps is the most common presentation. Disseminated cutaneous lesions without mucosal involvement and/ separate involvement of nose and cheek are extremely rare.[17] The mode of spread in our patient could have been due to autoinoculation but is more likely to have been haematogenous spread.

References:

1. Moses JD, Shanmugham A. Epidemiological survey of rhinosporidiosis in man – a sample survey in a high school located in a hyperendemic area. Indian Vet J 1987; 64: 34-8.
2. Loh KS, Chong SM, Pang YT et al. Rhinosporidiosis: differential diagnosis of a large nasal mass. Otolaryngol Head Neck Surg 2001; 124: 121-2.
3. Kumari R, Laxmisha C, Thappa DM. Disseminated cutaneous rhinosporidiosis. Dermatol Online J 2005; 11(1): 19.
4. Morelli L, Polce M, Piscioli F, Del Nonno F, Covello R, Brenna A, et al. Human nasal rhinosporidiosis: an Italian case report. Diagn Pathol 2006; 1: 25.
5. Arseculeratne SN. Recent advances in rhinosporidiosis and Rhinosporidium seeberi. Ind J Med Microbiol 2002; 20: 119-31
6. Ahluwalia KB. New interpretations in rhinosporidiosis, enigmatic disease of the last nine decades. J Submicrosc Cytol Pathol 1992; 24: 109-14.
7. Fredricks DN, Jolley JA, Lepp PW et al. Rhinosporidium seeberi: A human pathogen from a novel group of aquatic protistan parasites. Emerg Infect Dis 2000; 6(3): 273-82.
8. Herr RA, Ajello L, Taylor JW et al. Phylogenetic analysis of Rhinosporidium seeberi's 18S small-subunit ribosomal DNA groups this pathogen among members of the protoctistan Mesomycetozoa clade. J Clin Microbiol 1999; 37(9): 2750-4.
9. Arseculeratne SN. Recent advances in rhinosporidiosis and Rhinosporidium seeberi. Indian J Med Microbiol 2002; 20: 119-31.
10. Mandalik GS. A record of Rhinosporidial polyp: with some observation on mode of infection. Indian Med Gaz 1937; 72: 143-7.
11. Satyanarayana C. Rhinosporidiosis with a record of 255 cases. Acta Otolaryngol 1960; 51: 348-66.
12. Lupi O, Tyring SK, McGinnis MR. Tropical dermatology: Fungal tropical diseases. J Am Acad Dermatol 2005; 53: 931-51.
13. Arun BN, Manjula BV, Balasubramanyam AM, Ravi CN. Endoscopic removal of naso-oropharyngeal rhinosporidiosis: a report. Internet J Otorhinolaryngol 2009; 9.
14. Shah AK, Ingle MV. Tracheal rhinosporidiosis: A case report. J Postgrad Med 1985; 31: 210-1
15. Job A, Venkateswaran S, Mathan M et al. Medical therapy of rhinosporidiosis with dapsone. J Laryngol Otol 1993; 107: 809-12.
16. Krishnan KN. Clinical trial of diamino-diphenyl sulphone in nasal and nasopharyngeal rhinosporidiosis. Laryngoscope 1979; 89: 291-5.
17. Madhavan M, Ratnakar C, Mehdiratta KS. Rhinosporidial infection of the forehead. J Postgrad Med 1978; 24(4): 235-6.
18. Karunaratne WAE. Rhinosporidiosis in man. London: The Athlone Press; 1964.
19. Rajam RV, Viswanathan GC. Rhinosporidiosis: a study with a report of a fatal case with systemic dissemination. Ind J Surg 1955; 17: 269-98.
20. Kumari R, Laxmisha C, Thappa DM. Disseminated cutaneous rhinosporidiosis. Dermatol Online J 2005; 11(1): 19.