

## Case Report

# Rhinosporidiosis-an atypical presentation as a facial swelling in left cheek

E. Siva Kumar<sup>1</sup>, C. A. Swapna<sup>1</sup>, Lavanya Karanam<sup>1\*</sup>, Ram Kumar<sup>2</sup>

<sup>1</sup>Department of ENT, Sri Narayani Hospital and Research Centre, Vellore, Tamil Nadu, India

<sup>2</sup>Department of Plastic Surgery, Sri Narayani Hospital and Research Centre, Vellore, Tamil Nadu, India

**Received:** 17 September 2020

**Revised:** 09 October 2020

**Accepted:** 13 October 2020

**\*Correspondence:**

Dr. Lavanya Karanam,

E-mail: lavvi87@yahoo.com

**Copyright:** © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

### ABSTRACT

Rhinosporidiosis is a chronic granulomatous disease of mucous membranes caused by *Rhinosporidium seeberi*. Most frequently affected sites are nose and nasopharynx followed by eye. Report a rare case of 13-year-old female with rhinosporidiosis in a bizarre way of presentation as a cyst in left cheek. A 13-year-old female child hailing from West Bengal presented to ENT OPD with complaints of swelling in the left cheek for 6 months gradually increasing in size. On examination left side of cheek a cystic swelling extending from angle of mouth to lateral side of face was seen. Magnetic resonance imaging (MRI) neck plain and contrast showed large well defined T2 hyperintense cystic lesion in subcutaneous plane measuring about 5.7 cm (transverse)×6.0 cm (cephalocaudal)×5.5 cm (anteroposterior) with post contrast wall enhancement. Patient planned for excision and histopathological examination was consistent with rhinosporidiosis. Rhinosporidiosis continues to be a conundrum necessitating further literature and research on this. The difficulty in culturing the organism and its preternatural presentations add up to difficulties in diagnosing. Emphasise the quaint feature of its presentation as a cyst in cheek as primary site without any focus of infection in the respiratory tract and with negative history of traumatic inoculation.

**Keywords:** Rhinosporidiosis, Facial swelling, Histopathological examination

### INTRODUCTION

Rhinosporidiosis is a chronic granulomatous disease of mucous membranes caused by *Rhinosporidium seeberi*. Most frequently affected sites are nose and nasopharynx followed by eye. It is an endemic disease with more cases being reported from India and Sri Lanka.<sup>1</sup> Even though nose and nasopharynx are common sites there have been cases reported involving larynx, oropharynx, conjunctiva, lacrimal sac, trachea, bronchus, bone, vulva, vagina, penis, urethra, skin and subcutaneous tissue.<sup>2</sup> Rarely dissemination can occur with involvement of limbs, trunk, viscera and brain causing fatal outcome.<sup>1</sup>

Report a rare case of 13-year-old female with rhinosporidiosis in a bizarre way of presentation as a cyst in left cheek. It's one of the very few cases in such a presentation. Highlight the need for cautious clinical evaluation and discuss differential diagnosis and treatment.

### CASE REPORT

A 13-year-old female child hailing from West Bengal presented to ENT OPD with complaints of swelling in the left cheek for 6 months gradually increasing in size. She had no complaints of pain. There was no change in size of swelling while chewing or eating food. No history of halitosis or any discharge draining into mouth. No

complaints of nasal obstruction, nasal discharge and bleeding from nasal cavity.

On examination left side of cheek was very tense. A cystic swelling approximately 10×8 cm size extending from angle of mouth to lateral side of face was seen. Lower border of mandible was well defined. Swelling was soft, fluctuant, nontender and not attached to underlying structures. Skin over the swelling was normal with no local rise of temperature and no evidence of sinus/fistula. Intra oral examination was normal. There was no lymphadenopathy. Diagnostic nasal endoscopy was found to be normal. No evidence of any lesion in the nasal cavity and nasopharynx. Rest of the otorhinolaryngological examination was normal. Routine laboratory investigations were within normal limits.

CECT of maxillofacial region shows a well-defined peripherally enhancing hypodense measuring 4.3 cm (transverse)×3.7 cm (cephalocaudal)×3.5 cm (anteroposterior) features suggestive of abscess. Previous history of aspiration at an unknown centre was inconclusive with recurrence of swelling within a week.

MRI neck plain and contrast showed large well defined T2 hyperintense cystic lesion in subcutaneous plane measuring about 5.7 cm (transverse)×6.0 cm (cephalocaudal)×5.5 cm (anteroposterior). It closely abuts masseter muscle with internal T2/T1 fluid debris. Post contrast wall enhancement present extending from above infratemporal fossa up to angle of mandible closely abuts superficial lobe of parotid.



**Figure 1: MRI with contrast large well defined T2 hyperintense cystic lesion.**

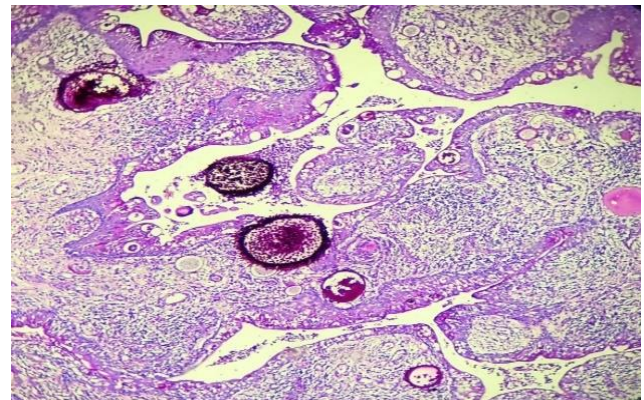
Provisional diagnosis of infected 2<sup>nd</sup> brachial cleft cyst, unilateral cystic hygroma, lipoma, parotid gland cyst and tumours of muscular origin were considered. Patient was planned for excision under GA. A modified parotid incision was given in front of left external auditory canal and lateral part of cyst was excised. Cyst was abutting the superficial lobe of parotid but gland was free. Medial part of cyst was accessed using a sublabial incision along left gingivobuccal sulcus. Cyst over the anterior wall of maxilla was excised in toto and sent for histopathological

examination. Patient had no complications in the postop period.

Histopathological examination showed chronic inflammatory granulation tissue with varying sized sporangia containing endospores and trophocytes ranging in size from 30-300 microns. Fat necrosis was present and showed no evidence of malignancy. The sporangia and endospores were positive for periodic acid-Schiff (PAS) and Gomori methenamine silver (GMS). It was suggestive of rhinosporidiosis with granulomatous inflammation. In view of this patient was started on tab Dapsone 100 mg once a day for 6 months. There is no recurrence till date.



**Figure 2: Histopathological examination for rhinosporidiosis positive for GMS.**



**Figure 3: Histopathological examination of rhinosporidiosis positive for PAS.**

## DISCUSSION

Rhinosporidiosis is chronic granulomatous disease affecting mucous membranes in both humans and animals. It was first discovered by Malbran in 1892 and later cases in cattle were reported in India in 1894.<sup>3</sup> First was reported by Guillermo Seeber in 1900 in a 19-year-old agricultural labourer suffering from nasal polyp, hailing from Buenos Aires. Wernicke named it as *Coccidium Seeberia*.<sup>4</sup> Herr et al 1999 classified the

organism *Rhinosporidium seeberi* as Mesomycetozoa based on biological analysis of 18S rRNA.<sup>5</sup>

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 rosette agent. In the order Ichthyophonida, class *Ichthyophonae* has members with phylogenetic features in common with genus *Ichthyophonus* and *Psorospermium*.<sup>6</sup>

In India, rhinosporidiosis is mostly reported from Chhattisgarh, Kerala, Tamil Nadu, Orissa and West Bengal. The patient was hailing from West Bengal. Most commonly affects age group between 20-35 years and males are more frequently affected compared to females with M:F ratio of 4:1. Due to natural aquatic habitat of the organism the usual mode of infection is transepithelial infection through traumatised epithelium.<sup>7</sup>

Various modes of spread have been identified as: autoinoculation through spillage of spores from polyps after trauma or surgery, haematogenous dissemination to distant sites, lymphatic, sexual.<sup>8</sup> Most commonly affects the nasal cavity presenting as a fleshy polypoidal growth. In this case due to swelling in the parotid region, MRI with contrast and CECT helped in identifying the exact site and extent of lesion. But the diagnosis was made only on histopathological examination.

On histopathological examination varying sized sporangia containing endospores and trophocytes ranging from 30-300 microns were seen. They were positive for PAS and GMS. This is the typical picture for diagnosis of rhinosporidiosis. Histopathological differential diagnoses include myospherulosis and coccidioides immitis.<sup>9</sup> Myospherulosis is an iatrogenically induced reactive process caused by the interaction of red blood cells with petrolatum, lanolin or traumatized human adipose tissue in the nose, paranasal sinuses and sub-cutaneous tissue. Coccidioides immitis has similar mature stages represented by large, thick-walled, spherical structures containing endospores, but the spherules are smaller, contain small endospores and do not stain with mucicarmine.

Primary treatment of choice is wide surgical excision with cauterisation of base in case of nasal polyps. In this case the cyst was removed using both a modified parotid incision and a sublabial approach with careful dissection due to proximity to parotid gland and duct. There were no complications like injury to parotid duct and facial nerve. There was a similar case reported previously.<sup>10</sup>

Postoperatively patient was put on tab Dapsone as it stops the growth cycle and induces fibrosis of surrounding stroma.<sup>11</sup> As rhinosporidiosis has a very slow progress a long term follow up is needed.<sup>12</sup> Recurrences can occur due to spillage of spores due to trauma or surgery. In this case there was no recurrence till date.

## CONCLUSION

Rhinosporidiosis is a very rare and endemic disease. It continues to be a conundrum necessitating further literature and research on this. The difficulty in culturing the organism and its preternatural presentations add up to difficulties in diagnosing. Emphasise the quaint feature of its presentation as a cyst in cheek as primary site without any focus of infection in the respiratory tract and with negative history of traumatic inoculation.

In this case the diagnosis was made only on histopathological examination, which is the gold standard investigation. It should be kept in mind in all cases hailing from endemic areas that rhinosporidiosis can be a possibility. Surgery along with dapsone forms crucial part of treatment. A long term follow up is needed as recurrence can occur after a protracted clinical course.

## ACKNOWLEDGEMENTS

Authors would like to thank Prof. Dr. Balaji Nandagopal, director and trustee of Sri Narayani hospital and research centre for giving us access to use the necessary clinical data for manuscript preparation.

*Funding: No funding sources*

*Conflict of interest: None declared*

*Ethical approval: Not required*

## REFERENCES

1. Arsecularatne SN. Recent advances in rhinosporidiosis and rhinosporidium seeberi. Indian J Med Microbiol. 2002;20(3):119-31.
2. Sudarshan V, Gahine R, Daharwal A, Kujur P, Hussain N, Krishnani C et al. Rhinosporidiosis of the parotid duct presenting as a parotid duct cyst-A report of three cases. Indian J Med Microbiol. 2012;30:108-11.
3. Ayyar VK. Rhinosporidiosis in cattle, a case recorded in a bullock. Transact Far-Eastern Asso Trop Med. 1927;3:658.
4. Karunaratne WA. The pathology of rhinosporidiosis. J Path Bact. 1936;42:193-202.
5. Herr RA, Ajello L, Taylor JW, Arsecularatne SN, Mendoza L. Phylogenetic analysis of *Rhinosporidium seeberi*'s 18S small subunit ribosomal DNA groups this pathogen among members of the ProtoctistanMesomycetozoa Clade. J Clin Microbiol. 1999;37(9):2750-54.
6. Ajello L, Mendoza L. The phylogeny of *Rhinosporidium seeberi*. 14<sup>th</sup> Meeting of the International Society for Human and Animal Mycology. Buenos Aires, Argentina. 2000:78.
7. Lupi O, Tyring SK, McGinnis MR. Tropical dermatology: Fungal tropical diseases. J Am Academy Dermatol. 2005;53:931-51.
8. Capoor MR, Khanna G, Batra RK, Nair D, Venkatchalam VP, Aggarwal P. Rhinosporidiosis in

- Delhi, North India: case series from a non-endemic area and mini-review. *Mycopathologia*. 2009;168(2):89-94.
9. Mounabati M, Banushree CS. Two rare cases of rhinosporidiosis of parotid duct: Case reports and review of literature. *Ann Maxillofacial Surg*. 2014;4(2):234-6.
  10. Nambiar SS, Radhakrishnan S, Vijayan A. Rhinosporidiosis: report of an extra-ductal facial lesion. *ID Cases*. 2017;7:40-3.
  11. Das S, Kashyap B, Barua M, Gupta N, Saha R, Vaid L et al. Nasal rhinosporidiosis in humans: new interpretations and a review of the literature of this enigmatic disease. *Med Mycol*. 2011;49:311-5.
  12. Chen L, Buonocore D, Wang B, Tabaee A. Delayed recurrence of sinonasal rhinosporidiosis. *Am J Otolaryngol-Head Neck Med Surg*. 2015;36:778-80.

**Cite this article as:** Kumar ES, Swapna CA, Karanam L, Kumar R. Rhinosporidiosis-an atypical presentation as a facial swelling in left cheek. *Int J Otorhinolaryngol Head Neck Surg* 2020;6:2142-5.