

Case Report

Lacrimal sac rhinosporidiosis: case report and review of literature with a new grading system to optimize treatment



Nishi Gupta^{a,*}; Poonam Singla^a; Bibhu Pradhan^b; Urmila Gurung^b

Abstract

Rhinosporidiosis is a chronic granulomatous disease affecting the mucous membrane primarily and is caused by *Rhinosporidium seeberi*, an aquatic protistan parasite. The nose is the most common site of involvement and is seen in 83.3% cases, followed by ocular involvement in 11.2% cases and other sites like larynx, trachea and bronchus in 5.5% cases. In various oculosporidiosis case series, lacrimal drainage system involvement was seen to vary from 14.3% to 59.6% cases. Isolated lacrimal sac involvement in rhinosporidiosis was found in 45.8% (72 out of 157) cases of the lacrimal drainage system in a review of 31 studies.

A variety of surgical procedures have been used to treat rhinosporidiosis of lacrimal sac like dacryocystorhinostomy, Dacryocystectomy, lateral rhinotomy and local lesion excision with a success rate varying from 28.5% to 92.3%. This wide variation in the success rate was due to the fact that a uniform surgical procedure was performed in all the cases of a particular series irrespective of the extent of disease.

Grading the lacrimal sac rhinosporidiosis to decide the extent of surgical excision may help achieve better results. We present a grading system based on our own experience in a case of extensive rhinosporidiosis of lacrimal sac and review of 31 studies published in the literature.

A 24-year-old male from Nepal presented with the complaints of watering from his right eye of 13 years duration, swelling in the right medial canthal area with an extension to the inferior part of the orbit for 12 years and nasal blockage for 1.5 years. The patient had a history of previous intervention in which biopsy was taken from the nose and sent for histopathology that confirmed rhinosporidiosis. An extended intranasal endoscopic dacryocystectomy was done along with debridement and coblation of the lesion over the septum and nasopharynx. Intraoperatively a large rhinosporidiosis mass was seen filling the sac and was removed in toto along with the sac and nasolacrimal duct. Recurrence of a tiny lesion after 6 months in our case despite wide excision with the drilling of bony nasolacrimal duct and coblation, made us review the literature.

Keywords: Rhinosporidiosis, Lacrimal sac (LS), Grading, Dacryocystorhinostomy (DCR), Dacryocystectomy (DCT), Nasolacrimal duct (NLD)

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Introduction

Rhinosporidiosis is a chronic granulomatous disease caused by *Rhinosporidium seeberi*. It is a rare aquatic protistan parasite.¹ It commonly affects the mucous membrane but can also affect other structures including larynx, trachea, skin,

genitalia, lungs and rectum. Isolated lacrimal sac rhinosporidiosis is very rare.¹

Kuriakose in 1963 coined the term oculosporidiosis for rhinosporidiosis of the eye.² A PubMed search of all the articles published in English on rhinosporidiosis of lacrimal sac and oculosporidiosis was performed. The relevant cross-referral

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Table 1. Review details of 31 studies on Lacrimal sac rhinosporidiosis published in literature.

| SN | Author | Year | n | Presentation | ROPLAS | NLD/syringing | Epiphora | Site/spread | CT scan findings | Treatment | Recurrence | Follow up | Medical therapy | Pond bath | Location |
|----|---------------------------------------|------|----|---|--|--|--|--|--|--|----------------|---------------|--|--------------|-----------------------------|
| 1 | Rajesh Raju and Sandeep ¹⁹ | 2018 | 13 | Swelling over LS area, Blood stained nasal discharge | Doughy swelling medial to medial canthus | Partial or complete block (Numbers NM) | Present in 2/13 cases | LS, NLD& Nose | Not done | Endoscopic DCR with NLD excision | 1/13 | 16 months | Not given | Present | Kerela(South India) |
| 2 | Prabhu et al. ³⁶ | 2018 | 4 | No details available (Radiological study) | NM | NM | NM | LS, NLD & nose | LS pyocele & NLD mucocele seen as a result of NLD block. Enhancing soft tissue mass in NLD and nose with same attenuation showing an upward extension. | Sac was excised, it was full of pink vascularised polypoidal growth | NM | NM | NM | NM | Tamilnadu (India) |
| 3 | Suneer and Sivasnkari ³³ | 2018 | 2 | NM | NM | NM | NM | LS & NLD | Not done | Excision of lesion no details | NM | NM | NM | Present | Kanyakumari (South India) |
| 4 | Chakraborti et al. ²⁴ | 2017 | 1 | Gradually increasing, soft, painless swelling of left lower eyelid | Serosanguinous | Patent | Nil | LS | Soft tissue enhancing mass near medial canthus of Left orbit. DCG showing diverticula | Via Ant orbitotomy through sub ciliary approach diverticula was removed leaving sac behind. It recurred with fistula with discharging spores | 1st postop day | 1 year | Iodine + Amoxyclav | Present | West Bengal (India) |
| 5 | Girish and Prathima ²⁷ | 2017 | 1 | Diffuse nontender infra orbital swelling of the left eye | Mucopurulent discharge | NM | Intermittent | LS + NLD | Hyperdense lesion in subcutaneous plane in left infraorbital region | DCT with enbloc resection of NLD. | NM | NM | 100 mg x3 months | Present | North Eastern part of India |
| 6 | Jamison et al. ³⁴ | 2016 | 1 | Swelling at nasal aspect of left lower lid. | NM | Patent | Absent | LS, NLD & nose | CT-DCG- donut distribution i.e. contrast passed through NLD in circumferential manner till it drained in IM, with asymmetry in lateral wall of nose | Gelatinous lesion attached to superior wall of lacrimal sac extending into NLD. Details of excision not mentioned. | NM | 5 months | NM | NM | Pakistan |
| 7 | Basu et al. ²⁰ | 2016 | 1 | Pinkish swelling over left lower orbital area for 3 years with a deep T scar from the previous intervention | NM | Patent | Nil | LS & NLD | Enhancing soft tissue mass in lacrimal sac region extending to superior aspect of right maxillary antrum, disruption of medial wall of the left orbit and blurred adjacent fat planes. | DCT, excision of mass with sac | NIL | 6 months | 100 mg OD X6 days for 6 months | NM | Kolkata (India) |
| 8 | Mishra et al. ¹ | 2015 | 1 | Soft, non-tender swelling at the medial canthus of left eye, 4cmx2cm in size | NM | Blocked | Present | LS + NLD | Hyperdense lesion with mean CT attenuation of 48 hounsfield unit lesion is extending to upper part of NLD | An elliptical incision was made over the medial canthus of left eye. Mass with sac removed. Silastic tube placed from punctum to nose | NIL | 1 month | 100 MG OD X6 months | NM | Bhuvneshwar (India) |
| 9 | Sah et al. ¹⁴ | 2014 | 1 | Left medial infraorbital diffuse nontender swelling | Reddish mucopurulent discharge | Partial patent | Intermittent | LS + NLD | Isodense lesion with mild enhancement in preseptal compartment | Multiple tiny vascularized growth seen. Sac was sutured and removed enbloc with NLD | Nil | 2 Yrs | 100 mg x3 months | NM | Terai (Nepal) |
| 10 | Nuruddin ¹⁰ | 2014 | 18 | Soft doughy swelling in LS area with epistaxis and blood-stained discharge from puncta. | NM | Blocked only in 4 cases, patent in rest 14 | Blood stained discharge only in 4 patients | LS n = 16, lacrimo-cutaneous fistula n = 2 | Not done | Modified DCR done. All sac content along with medial and lateral wall was excised, a small portion of the sac around common canaliculi was left, DCR tube placed | 2 out of 18 | 1 year | Dapsone not used Povidone iodine for 2 minutes | 12 out of 18 | Bangladesh |
| 11 | Guru and Pradhan ²³ | 2014 | 10 | Blood-tinged discharge from eye | Blood tinged discharge | Blood tinged discharge on irrigation | Blood tinged discharge | LS and nose n = 7, NLD and nose n = 3 | Not done | DCT, cauterisation of base. Debridement of mucous membrane of NLD | NM | NM | Dapsone | NM | Burla Sambalpur (India) |
| 12 | Mukherjee et al. ²² | 2013 | 1 | Recurrent painful swelling below right lower lid | Negative | NM | Patent | LS | small oval swelling in the medial canthal area, deviated nasal septum maxillary sinus polyp with concha bullosa | DCT with wide excision with cauterization | NM | Not mentioned | Povidone iodine + 100 mg Dapsone OD × 6mnths | NM | Chennai (India) |

| | | | | | | | | | | | | | | | | |
|----|----------------------------------|------|----|--|--------------------------------------|-----------------------------------|--|---------------------------------------|---|---|--|---|----------|--|---------|--|
| 13 | Belliveau et al. ²¹ | 2012 | 1 | Bloody tear with mild tender swelling in the medial canthal area of the left eye. | Mucopurulent discharge | Patent | | Bloody tear Present | LS and NLD | Left Lacrimal sac mass, no bony destruction. | Open excision biopsy done, frozen section followed by external DCR | NIL | 5 years | No Dapsone | NM | Canada (Migrated from Bangladesh) |
| 14 | Mithal et al. ²⁵ | 2012 | 13 | Soft fluctuating swelling in medial canthus area | NM | | | Blood tinged discharge | LS + NLD | Not done | DCT with en bloc resection of growth in NLD was done, pink vascularised growth with finger like extension was seen. | 1 | 6 months | not given | NM | South India |
| 15 | Pushkar et al. ⁴ | 2012 | 1 | Large painless ill defined, boggy swelling 5x5cm in size below medial canthus, | Mucopurulent discharge | Blocked with mucopurulent disease | | Present | LS with extension into NLD | DCG showed a dilated LS with intraluminal filling defect. Soft tissue mass lesion within LS and NLD extending up to IM. Expanded NLD had displaced the IT medially. | Sac wall was incised, pink vascularised, polypoidal growth was seen, sac was sutured, DCT done. Extension of growth in nasopharynx removed en bloc with the sac. | NIL | 6 months | dapsone given | NM | Delhi (patient's native place not mentioned (India)) |
| 16 | Rogers et al. ²⁶ | 2012 | 1 | Swelling left inner canthus. | Watery discharge from nose | NM | | Present | LS | Dacryocystogram showed passage of dye but significant constriction of the nasolacrimal duct. | External DCR done | Recurrence noted | 10yrs | Not given | NM | Sydney (Patient origin (Bangladesh)) |
| 17 | Satya narayana ³ | 2009 | 3 | Non-compressible boggy swelling of the lacrimal sac | NM | NM | | NM | LS with skin infiltration as was evident from skin ulceration on pressure with discharge of thick mucous secretion containing spores. | Not done | Excision cautery, details not known | not mentioned | NM | NM | NM | Madras (India) |
| 18 | Ghosh et al. ²⁸ | 2008 | 1 | Fluctuant swelling in medial canthus area with epiphora and purulent discharge from eye. | Mucoid secretion from medial canthus | NM | | Purulent discharge from the left eye | LS | Not done | DCT | NM | NM | NM | NM | Kolkata (India) |
| 19 | Varshney et al. ³⁵ | 2008 | 1 | Right facial swelling, nasal obstruction and intermittent epiphora. | NM | Blocked | | watering | LS, Nose& oropharynx | Cystic fluid air filled lacrimal sac with irregular filling defect, no bony erosion. LS diverticulum | Lateral rhinotomy with sac excision | Nil | 3 months | NM | NM | Dehradun (India) |
| 20 | Ghorpade et al. ²⁹ | 2007 | 1 | Gradually progressive, painful swelling under right eye for 8 months with scanty bloody nasal discharge. | NM | NM | | NM | Small nasal mass with extension in LS & NLD | Not done | The mass was reached through an incision in the right naso-optic sulcus. A pinkish mass with whitish small dots on its surface was found to occupy the lacrimal sac and projecting into the lacrimal duct. It was extracted from the upper opening of the duct and the part in the inferior meatus was excised separately from the right nasal chamber followed by electrocautery. | Nil | 8 months | Dapsone 100 mg daily orally, and has not had a recurrence since last 8 months. | Present | Bhilai (India) |
| 21 | Chowdhury RK et al. ¹ | 2007 | 3 | Epistaxis | NM | NM | | NM | LS n = 1, LS with subcutaneous spread n = 2 | Not done | DCT done, pink vascularized finger like extension was seen in all the cases | Two out of three that had subcutaneous spread | NM | NM | Present | Western Orrisa (India) |
| 22 | Watve et al. ⁹ | 2006 | 1 | Nontender swelling Rt medial canthal area | Fluid | NM | | Continuous epiphora purulent at times | LS | Not done | Endoscopic DCR done, a mass popped out of sac during surgery | NIL | NM | 100 mg alternate day × 1 yr | NM | Western Maharashtra (India) |

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Table 1 (continued)

| SN | Author | Year | n | Presentation | ROPLAS | NLD/syringing | Epiphora | Site/spread | CT scan findings | Treatment | Recurrence | Follow up | Medical therapy | Pond bath | Location |
|----|---|------|----|--|---|--|--------------------|--|--|---|---------------|-----------|------------------------|--|-----------------------------|
| 23 | Nerurkar et al. ¹⁸ | 2004 | 1 | Diffuse, soft, non-tender Rt infraorbital swelling. diffuse | NM | Blocked | Intermittent | LS | Isodense lesion with mild enhancement in preseptal compartment. There was enhancement within extraconal compartment of right orbit medially suggestive of a lesion in NLD & inferior aspect of right orbit | Endoscopic DCR | Present | 1 Week | 50 mg daily | NM | Migrant from Orrisa (India) |
| 24 | Thakur et al. ¹⁵ | 2002 | 3 | Watering eye, no swelling, fistula in one case | Regurgitation produced a reddish discharge from the upper punctum | NM | watering present | LS n = 2, LS with fistula/previous history of DCR with silastic tube n = 1 | Not done | DCT | Not mentioned | NM | NM | Present | Nepal |
| 25 | Shreshtha et al. ¹¹ | 1998 | 6 | Soft, fluctuating swelling around LS with bleeding from eye & nose | Regurgitation was soft slimy and sprinkled with fine white granular particles | Patent | Purulent discharge | LS in 3, LS with spread in surrounding area n = 3 | Not done | DCT done sac was opened, vascularised growth with finger like extension seen. Typical white spots on the surface of growth gave an appearance resembling that of bunches of fish eggs. Sac was sutured and removed. | 1 | 2.5 years | Topical betadine | Present | Nepal (Lahan & Dharan) |
| 26 | Krishnan et al. ³⁰ | 1986 | 1 | Swelling at the inner canthus of the right eye with epiphora and occasional blood-stained discharge | Serosanguinous discharge | Blocked | NM | LS | Dacrycystography showed diverticulum of LS | DCT (sac + diverticula) | NM | NM | NM | NM | South India (Pondicherry) |
| 27 | Mukherjee et al. ³¹ | 1982 | 48 | Diffuse swelling over the sac n = 45 Widening of the nose bridge in n = 42, swelling extending under the lower lid n = 30, localised swelling over the sac n = 3 | Nil | Blocked in 75% (36) cases, partial block in rest(12 cases) | Nil | LS n = 42, LS & nose n = 6 | Not done | DCT | NIL | NM | NM | NM | Raipur (MP India) |
| 28 | Suseela and Subramaniam ¹³ | 1975 | 7 | Epistaxis main symptom as the lesions involved nose, | Negative | Patent | NM | LS & nose | Dacrycystography showed dilated sac and duct | Excision biopsy | 5 out of 7 | NM | NM | Present | Kerala (India) |
| 29 | Jain and Sahai ³² | 1974 | 1 | Gradually increasing painless swelling underneath, the skin on rt lateral side of the bridge of nose | NM | patent | Absent | LS | Not done | DCT (sac with growth excised) | NIL | NM | NM | Agriculturist | Udaipur (India) |
| 30 | David and Sivaramasubrahmanyam ⁷ | 1973 | 4 | Swelling lower lid | Regurgitation of blood in nose | NM | NM | LS + Nose & Limbus | Not done | DCT | 1 | NM | NM | 1 case had injury by fall + diving in tank | Tamilnadu (India) |
| 31 | Kuriakose et al. ² | 1963 | 6 | Soft fluctuant, non-tender swelling of sac extending to lower lid with epistaxis. | Present | Patent | NM | NM | Not done | Details not mentioned, complete excision was difficult because of severe bleeding. Silver nitrate was applied after excision | Present | NM | Silver nitrate cautery | NM | Kerala (India) |

NM: Not mentioned, LS: Lacrimal sac, NLD: Nasolacrimal duct, DCR: Dacryocystorhinostomy, DCT: Dacrycystectomy, ROPLAS: Regurgitation on Pressure Over Lacrimal Sac Area, n: Number of cases, IT: Inferior turbinate, Rt: Right, Lt: Left, IM: Inferior meatus, DCG- Dacrycystography.

of all these articles was also reviewed. The first description of this parasite *Rhinosporidium seeberi* was given by Malbran from Buenos Aires, in 1892, who described it as a sporozoan parasite in nasal polypus followed by Guillermo seeber in 1986, who referred it as sporozoan belonging to subdivision coccidia.³ It is now considered an aquatic protistan parasite belonging to the class mesomycetozoa.^{1,4,5,6}

Kirkpatrick published the first case of lacrimal sac (LS) rhinosporidiosis in 1916.^{1,7,8}

Though oculosporidiosis is worldwide in distribution, it is relatively more prevalent in Southern India, Srilanka and Southeast Asia⁹ and accounts for 15% of the cases of rhinosporidiosis.^{4,6,10} The most common age group affected is 15–40 years with a predominance in males.^{1,2,3,7,10–12}

In various oculosporidiosis case series, lacrimal drainage system involvement was seen to vary from 14.3% to 59.6 %

cases.^{2,4,11–13,14–16} Frequency of isolated lacrimal sac involvement was seen in 45.8% (72/157) cases of the lacrimal drainage system in total of 31 studies reviewed.

History of treatment of lacrimal sac rhinosporidiosis dates back to 1949 when Rambo did incision and curetting of the LS region and packed it with sulphonamide without much relief.^{2,17} Kuriakose quoted that recurrence is always the rule and often a LS fistula results which are very resistant to further treatment.² Excision of LS polyp has been reported to be unsatisfactory as complete removal is difficult due to excessive bleeding¹¹ and the recurrence is inevitable.¹⁰ Advancement in technology with the introduction of high definition camera and endoscopes along with coblator have made it possible to perform meticulous excision of the lesion but the subepithelial extension cannot be eradicated completely.^{7,11,12}

Table 2. Grading of Lacrimal sac rhinosporidiosis based on the extent of the disease.

| S. N. | Author | Year | n | Site of involvement | Treatment | Recurrence |
|----------------|--|------|----|---|---|---------------------------------------|
| Grade 1 | | | | | | |
| 1 | Suneer and Sivasnkari ³³ | 2018 | 1 | LS + NLD | DCT | NM |
| 2 | Chakraborti et al. ²⁴ | 2017 | 1 | LS diverticula | Via Ant orbitotomy through sub ciliary approach diverticula was removed leaving sac behind. | 1/1 with fistula discharging spores |
| 3 | Girish and Prathima ²⁷ | 2017 | 1 | LS + NLD | DCT with en bloc resection of NLD | NM |
| 4 | Basu et al. ²⁰ | 2016 | 1 | LS + NLD | DCT | Nil |
| 5 | Mishra ¹ | 25 | 1 | LS + NLD | DCT with intubation from punctum to nose | Nil |
| 6 | Sah ¹⁴ | 2014 | 1 | LS + NLD | DCT with en bloc resection of growth in NLD | Nil |
| 7 | Nuruddin et al. ¹⁰ | 2014 | 16 | LS | Modified DCR | Nil |
| 8 | Mithal et al. ²⁵ | 2012 | 13 | LS + NLD | DCT with en bloc resection of NLD | 1/13 |
| 9 | Rogers et al. ²⁶ | 2012 | 1 | LS | External DCR | 1/1 |
| 10 | Ghosh et al. ²⁸ | 2008 | 1 | LS | DCT | Nil |
| 11 | Chowdhury et al. ¹² | 2007 | 1 | LS | DCT | Nil |
| 12 | Watve et al. ⁹ | 2006 | 1 | LS | Endoscopic DCR | Nil |
| 13 | Nerurkar et al. ¹⁸ | 2004 | 1 | LS | Endoscopic DCR | 1/1 |
| 14 | Thakur et al. ¹⁵ | 2002 | 1 | LS | DCT | NM |
| 15 | Shrestha et al. ¹¹ | 1998 | 3 | LS | DCT | Nil |
| 17 | Krishnan et al. ³⁰ | 1986 | 1 | LS diverticulum | DCT | NM |
| 18 | Mukherjee et al. ³¹ | 1982 | 42 | LS | DCT | Nil |
| 16 | Jain and Sahai ³² | 1974 | 1 | LS | DCT | Nil |
| Grade 2 | | | | | | |
| 1 | Rajesh Raju and Sandeep ¹⁹ | 2018 | 13 | LS + NLD + Nose | Endoscopic DCR with NLD excision | 1/13 |
| 2 | Prabhu et al. ³⁶ | 2018 | 1 | LS + NLD + Nose | DCT | NM |
| 3 | Suneer and Sivasnkari ³³ | 2018 | 1 | LS + NLD + Nose | DCT + en bloc resection of NLD | NM (states high chance of recurrence) |
| 4 | Jamison et al. ³⁴ | 2016 | 1 | LS + NLD + Lateral wall of inferior meatus | Lesion excision en mass | NM |
| 5 | Guru and Pradhan ²³ | 2014 | 10 | LS & nose n = 7, NLD & nose n = 3 | DCT | NM |
| 6 | Pushkar et al. ⁴ | 2012 | 1 | LS + NLD + Nose | DCT + en bloc resection of sac | Nil |
| 7 | Varshney et al. ³⁵ | 2007 | 1 | LS + Nose + Oropharynx | Lateral rhinotomy + DCT | Nil |
| 8 | Shrestha et al. ¹¹ | 1998 | 3 | LS + surrounding areas | DCT | 1/3 |
| 9 | Mukherjee et al. ³¹ | 1982 | 6 | LS + Nose | Nil | Nil |
| 10 | Suseela ¹³ | 1975 | 7 | LS + NLD + Nose | Excision biopsy | 5/7 |
| 11 | David and Sivaramasubrahmanyam et al. ⁷ | 1973 | 4 | LS + nose + limbus | DCT | 1/4 |
| Grade 3 | | | | | | |
| 1 | Nuruddin et al. ¹⁰ | 2014 | 2 | LS + Nose + canaliculi + Lacrimo-cutaneous fistula | Modified DCR | 2/2(100%) |
| 2 | Satyanarayana ³ | 2009 | 3 | LS with skin infiltration as was evident from skin ulceration that occurred on pressure over the swelling | NM | NM |
| 3 | Chowdhury et al. ¹² | 2007 | 2 | LS + subcutaneous spread | DCT | 2/2(100%) |
| 4 | Thakur et al. ¹⁵ | 2002 | 1 | LS with skin fistula | Excision of growth | NM |

LS: Lacrimal sac, NLD: Nasolacrimal duct, DCT: Dacryocystectomy, DCR: Dacryocystorhinostomy, NM: Not mentioned, n: Number of patients.

Various authors have published their results using surgical procedures like dacryocystorhinostomy (DCR) involving endoscopic DCR^{18,19} external DCR,⁹ modified DCR¹⁰ and Dacryocystectomy (DCT)^{3,11,20,21,22,23} with a success rate varying from 28.5% to 92.3%.

A review of published literature on LS rhinosporidiosis was conducted (Table 1) and it was observed that there were no set criteria used for choosing a particular procedure based on the extent of disease.

As a result, when a uniform procedure was applied to all the cases irrespective of the spread, those in grade 1 as per our grading, showed good results while those falling in grade 2&3 recurred.¹⁰⁻¹² A grading system has been devised to understand the disease extension, surgical plan and recurrence pattern (Table 2).

Case report

A 24-year-old boy from Nepal presented with the complaints of watering from his right eye for 13 years, right medial canthal swelling extending to the inferior part of the orbit for 12 years and nasal blockage for 1.5 years. External local examination revealed a large diffuse boggy swelling in right infraorbital area extending superiorly along the nasal bridge reaching up-to-the eyebrow and inferiorly along the infraorbital margin. The overlying skin was inflamed, tender with no regurgitation through the punctum on gentle pressure over the swelling though, the nasolacrimal duct was blocked on syringing. There was a history of biopsy from the nose that showed rhinosporidiosis. On endoscopic examination, there was a granular, lobulated, reddish pink growth over the posterior part of the septum at the bony cartilage junction with oedema and inflammation in the middle meatus area and another 2 × 2 mm lesion in the nasopharynx.

CT scan showed a hyperdense mass filling the right lacrimal sac area with subcutaneous extension and bony destruction. Right inferior turbinate was absent with just a stump seen with the air-filled nasal cavity on the right side. There was a bony erosion of the lacrimal fossa with soft tissue extension into the nose (Fig. 1A and B).

Written informed consent was obtained from the patient. Institutional review board approval was obtained. Intraoperatively a large rhinosporidiosis mass was seen in the LS lumen (Fig. 1C and D) that was removed along with the walls of the sac and NLD followed by coblation of the base and surrounding area to avoid seeding of the spores. This was achieved by an extended endonasal endoscopic DCT involving complete extirpation of the sac with nasolacrimal duct (NLD) up to its distal end with uncinectomy and clearance of ethmoids. Bone overlying the lacrimal fossa and the frontal process of the maxilla was drilled. The lateral wall of the nose in and around the bony NLD was removed, septal and nasopharyngeal lesions were debrided and coblated followed by irrigation with 5% povidone-iodine for 2 minutes. Histopathology revealed the sporangia of rhinosporidiosis at various stages of the life cycle. There was dense chronic inflammatory cell infiltrate mainly plasma cells and lymphocytes (Fig. 1E and F).

Regular check endoscopies were done at 1 week, 2 week, 4 weeks. Though the patient remained asymptomatic for 6 months. At 6 months follow up, a check endoscopy showed a small granular lesion on the lateral nasal wall. The patient was happy and satisfied as he was asymptomatic. His swelling

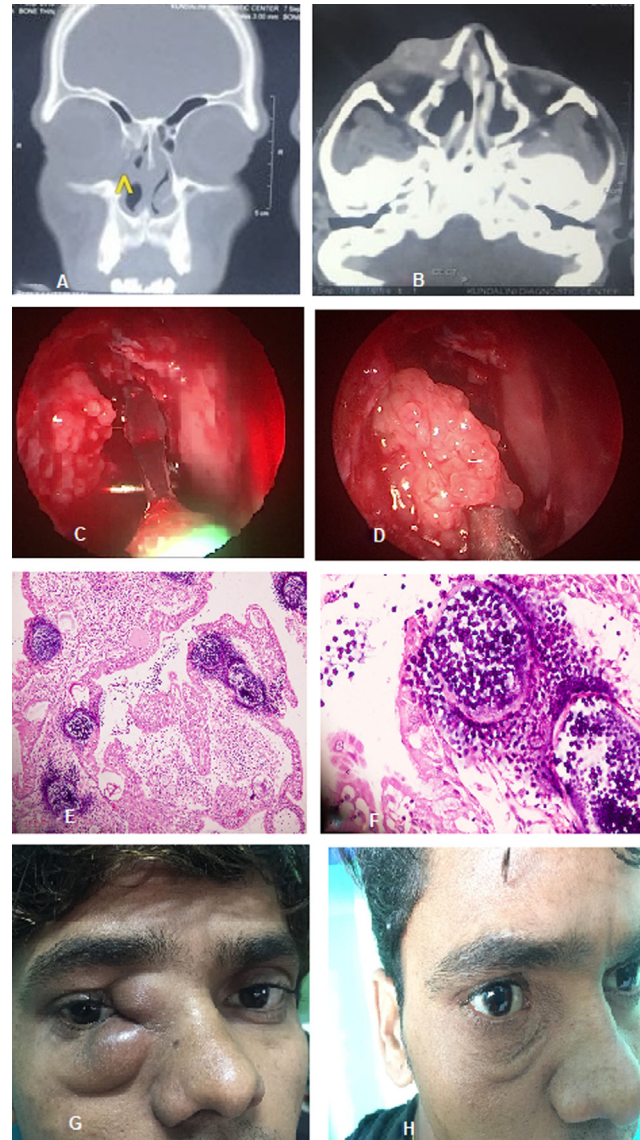


Fig. 1. A and B: Computed tomography scan of orbit & paranasal sinuses, axial & coronal view revealed soft tissue mass in the lacrimal sac fossa with a bony breach (arrow head) and extension into the nose. C and D: Endoscopic view of the right nasal cavity showing rhinosporidiosis mass in the lacrimal sac with probe insitu. E and F: Microphotograph showing epithelium of lacrimal sac with multiple sporangia and sporocysts of rhinosporidiosis with chronic inflammation. G and H: Preoperative and postoperative clinical photograph of the patient.

in the medial canthal area had completely disappeared with well-healed skin and no epiphora (Fig G and H). However, in view of the tiny recurrent lesion, he was advised endoscopic excision and electrocautery but he refused any further intervention.

Discussion

Rhinosporidiosis is presumably a waterborne disease and generally, occurs after swimming in stagnant freshwater ponds, lakes, or rivers, but is also suspected to occur from dust or air.² The natural hosts of the aquatic parasite are fish and amphibians.²²

Patient presents with swelling in the medial canthal area with or without epiphora. Blood stained discharge from the

eye or nose may be present. The nose is the most common site of involvement in rhinosporidiosis and is seen in 83.3% cases, followed by ocular adnexa in 11.2% cases, while other sites are involved in 5.4% cases.¹⁵ The other sites involved are the mucous membrane of larynx, trachea, bronchus and genitalia.¹⁵ In various oculosporidiosis case series, lacrimal drainage system involvement was seen in 14.3% to 59.6% cases.^{2,4,11-13}

The current study presents a review of 31 studies published in the literature (Table 1). Most of the cases presented with swelling in the medial canthal area extending along the infraorbital margin.^{1,2,3,7,9-11,17,14,21,24-33} The swelling was fluctuant^{1,16} and boggy^{7,9,33} and felt like a bag of worms.²⁴ It was painless in most of the studies,^{1,2,4-7,9,10,15,16,18,26,3} while some of them reported tenderness over the swelling.^{21,22,31}

An associated symptom of epistaxis or blood-stained discharge from nose or eye^{2,10,13,22,33} with bloody tears^{21,30} should raise a high index of suspicion of rhinosporidiosis, especially in endemic areas. The nature of discharge on regurgitation test varied from serosanguinous^{17,24,30} to purulent^{4,22,27} and slimy sprinkled with white granular particles¹¹ to reddish discharge with granular material.¹⁵

Epiphora may be absent in some cases of nasolacrimal rhinosporidiosis^{32,34} because the spread of infection is pericanalicular and perisaccal.²⁷ However, epiphora when present, may be continuous^{3,9,11} intermittent^{9,14,18} or blood stained.^{10,21,23} NLD was fully patent in some studies,^{11,20,24,32,34} partially patent in others¹⁴ and completely blocked in the rest.^{1,3,27,30,35}

Our patient presented with the characteristic features of large boggy swelling in the medial canthal area with continuous epiphora. The skin over the swelling was inflamed with puckering and impending fistula thus, firm pressure was not applied as ulceration of skin over the swelling on pressure has been reported.¹ This indicates subcutaneous spread with skin infiltration classifying it into grade 3.

There are various schools of thoughts on the possible route of spread in lacrimal sac involvement in rhinosporidiosis. While some felt the sac is involved by upward extension of the mass from the nose^{13,23,35} the others commented that the LS cannot be involved through NLD because the lacrimal folds act as a valve to prevent the secretion of the nose from being driven up into the duct.⁷

LS could get involved by the spread of infection from lacrimal canaliculi to the sac^{3,27} through permeation along the subepithelial connective tissue¹ or via the subepithelial lymphatic channels.⁷ In our case, the LS was the primary site of involvement with spread to nose through NLD as epiphora and swelling remained the only symptoms for 12 years and nasal symptoms appeared after more than a decade.

Isolated rhinosporidiosis of the LS is very rare^{11,20} and it becomes difficult to suspect a LS swelling to be due to infestation by rhinosporidium⁵ and thus imaging modalities are helpful. On computed tomography, rhinosporidiosis is commonly seen as a homogeneously enhancing lobulated lesion in the inferior nasal cavity with adjacent bone erosion.³⁶ Bone involvement is seen as irregularity, rarefaction, partial or complete erosion of inferior turbinate, thinning of the medial maxillary wall and septal erosion.³⁶

Contrast enhanced CT has an important role in delineating the site and extent of disease as well as bone and NLD involvement. NLD extension was defined as the extension

of soft tissue mass within the NLD, having similar attenuation as the mass in nasal cavity.³⁶

Characteristic findings of "donut distribution" i.e. contrast passes through NLD in a circumferential manner till it drained in the inferior meatus on computed tomographic dacryocystography (CT DCG) with normal sac wash out.³⁴

The swelling in the medial canthal area with a patent NLD on CT DCG in an endemic area should raise the suspicion of LS rhinosporidiosis. Dilated sac and dilated NLD were also seen in other studies.^{3,12}

Computed tomography helped us to judge the extension of the lesion in our case to the nose through a bony breach with the subcutaneous spread.

The aim of grading is to facilitate the planning of a particular surgical procedure e.g. DCR, modified DCR, DCT, Extended DCT and the need for multi-disciplinary approach wherever needed.

- **Grade 1:** Lesion limited to LS lumen as a pedunculated or sessile mass or polyp ± NLD but no lesion in nose or eye.
- **Grade 2:** Lesion involving LS, NLD and nose or eye.
- **Grade 3:** Lesion involving LS, NLD and nose or eye and spread to skin ± Lacrimo-cutaneous fistula.

Grading of the cases in various studies has been listed (Table 1). In the same series, cases were in different grades but a uniform surgical procedure was applied to all leading to recurrence in the advanced cases.

The proposed strategy for surgical management of LS rhinosporidiosis is that the cases in Grade 1 with just a pedunculated or sessile mass limited to LS alone can be treated with DCR or modified DCR may be preferred modality.¹⁰ In cases of grade 1 with mass completely filling the sac & reaching NLD, it is preferred to plan DCT with complete NLD resection and cauterization of the base. In grade 2, DCT with en bloc resection of NLD with thorough excision and cauterization of nasal or eye lesion is recommended. It might require a multidisciplinary approach based on the extent of the lesion. In grade 3 extended DCT involving uncinectomy, ethmoidectomy, complete extirpation of the sac with en bloc removal of NLD along with fistulectomy in cases with presence of fistula should be done. Extensive drilling of the bone of lacrimal fossa and nasolacrimal duct i.e. frontal process of maxilla, lacrimal bone and lateral wall of the inferior meatus is done additionally. Bipolar cauterization of the periorbita if infiltrated and removal of orbital fat if the lesion involves the orbit should be done. The patient needs to be explained the need for rigorous follow-up and regular endoscopic checks.

Due to the availability of the high definition camera and best endoscopes, it has become possible to address the extensive lacrimal sac, Naso-orbital and even skull base lesions endoscopically through the nose. Some studies advocated, an endonasal endoscopic approach citing the risk of seeding of the spore at the incision site in external DCR.¹⁸ We, however, feel that the route of surgical intervention does not make much difference provided the removal is complete. This understanding is based on the assumption that a multidisciplinary approach will always be used in handling lesions extending extension beyond the sac into the nose or eye.

Grading the disease as a part of preoperative assessment not only helps in deciding the extent of excision but also

helps in predicting the outcome and explaining the prognosis to the patient.

These observations suggest that limited disease has a better prognosis while the involvement of more than one site, subcutaneous spread and lacrimo-cutaneous fistula have guarded prognosis. This could also explain recurrence in our case as it presented in grade three with associated nasal lesion with the subcutaneous spread.

Role of Dapsone (Diamino-diphenyl-sulfone) in reducing recurrence rate is attributed to an arrest of maturation of spores and an accentuated granulomatous response.^{9,15} Topical application of 5% Povidone-iodine for 2 minutes was performed as it causes metabolic inactivation of endospores.¹⁰

However, in view of the molecular characterization of the aetiological agent, further research to discover effective treatment options is urgently needed.³⁷

Declaration of Competing Interest

The authors declare no conflicts of interest.

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