

Ocular adnexal sporotrichosis: A case series



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INTRODUCTION

Sporotrichosis, which is caused by fungi of the genus *Sporothrix*, is the most common subcutaneous mycosis.¹ This infection has a worldwide distribution, with areas of high endemicity in Latin America, including the metropolitan region of Rio de Janeiro.² The clinical presentations are highly variable and poorly understood. The most common presentations are the lymphocutaneous and the fixed cutaneous forms. The disseminated cutaneous and the extracutaneous/disseminated forms are rarer and are usually associated with immunosuppression or other routes of transmission.³ The distinction between the affected structures in the eye and its appendages can be difficult, as can be the therapeutic approach, because of the low prevalence of these rare forms. Three cases of sporotrichosis compromising the ocular adnexa are illustrated and described here, with a focus on differentiating them from intraocular sporotrichosis. Classifying these forms according to the 4 clinical presentations (lymphocutaneous, fixed, disseminated cutaneous, and extracutaneous)¹ is important for early diagnosis and correct management. Informed consent was obtained for the 3 patients reported here.

CASE SERIES

Case 1

A 65-year-old woman presented with a 1-month evolution of multiple erythematous papules on the

left upper eyelid, with progressive growth into infiltrated nodules, extending to the medial canthus, which evolved with superficial crusts (Fig 1). Initially, direct mycologic examination with potassium hydroxide and culture for fungi were negative. A biopsy of the skin lesion was inconclusive. A second biopsy was performed, and culture finally revealed growth of *Sporothrix* species. Histopathologic examination revealed acanthosis, hyperkeratosis, and dense granulomatous lymphomononuclear infiltrates (suppurative granuloma).

Case 2

A 7-year-old girl presented with a 3-week history of a painful erythematous papule on the inferior right eyelid. The family had a cat that had recently died from sporotrichosis, and the girl's mother was being treated for the mycosis. On examination, there was dacryocystitis with a fistula and drainage of purulent exudate (Fig 2). A swab of the fistula was positive for *Sporothrix* species.

Case 3

A 47-year-old woman had a 4-week history of an evolving membranous conjunctival lesion involving the plica semilunaris and caruncle of the left eye, with purulent exudate. There was no history of previous trauma. Physical examination showed a membranous yellowish lesion, with a smooth, shiny surface associated with conjunctival hyperemia and

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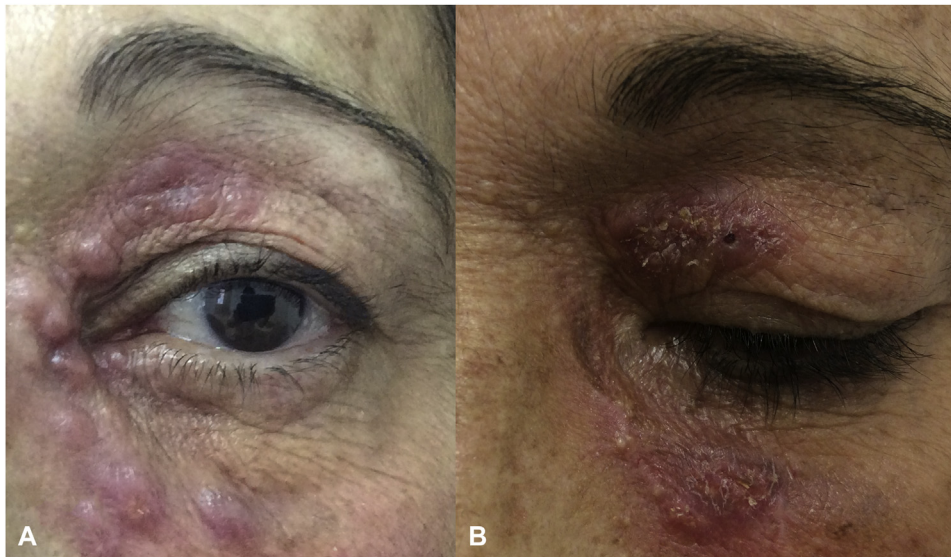


Fig 1. Case 1: A 65-year-old woman with eyelid involvement. **A**, Erythematous-infiltrated nodules. **B**, One month after first clinical appointment: crusts on the left upper eyelid and medial canthus.

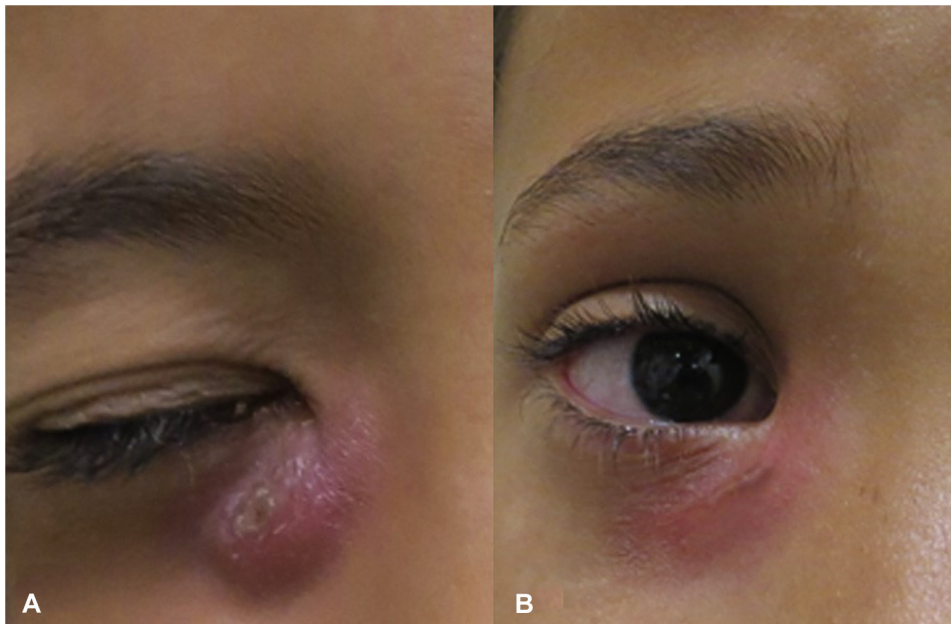


Fig 2. Case 2: A 7-year-old girl with dacryocystitis with a fistula on the inferior right eyelid. **A**, Acute phase with edema, erythema, and crust. **B**, Chronic phase, with scar and epiphora.

purulent exudate. No subcutaneous nodules were palpated (Fig 3). An incisional biopsy was performed, and *Sporothrix* species was isolated in culture. Histopathologic examination revealed dense granulomatous lymphomononuclear infiltrates (suppurative granuloma).

In all these patients, the mean time of onset was approximately 4 weeks, with report of previous contact with domestic cats. Upon enquiry, all patients confirmed that their cats had skin lesions.

Cases 1 and 3 were cured with oral itraconazole 100 mg twice daily for 3 months. In case 2, the lesion cleared after oral itraconazole 100 mg/day for 3 months, but a second course of the same treatment was required because of a culture-proven relapse 2 months later, for an additional 5 months to cure.

DISCUSSION

The ocular adnexa are the tissues and structures surrounding the eye, including the orbital soft tissue,

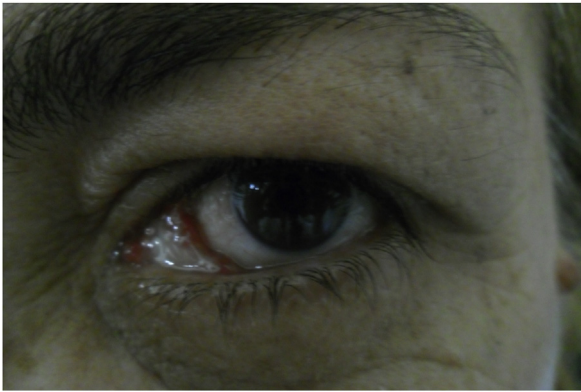


Fig 3. Case 3: A 47-year-old woman with “primary conjunctival sporotrichosis.” Clinical presentation at admission: membranous yellowish lesion, with a smooth and shiny surface involving the plica semilunaris and caruncle of the left eye, associated with conjunctival hyperemia and purulent exudate.

lacrimal system, conjunctiva, eyelids, and eyebrows. To date, reports of sporotrichosis in the ocular adnexa are limited, and no comparative studies have been published related to the above-mentioned structures.⁴

An increasing number of cases from endemic areas of sporotrichosis have been diagnosed, specifically involving the ocular adnexal structures.^{2,4,5} The clinical overlap between sporotrichosis and the more common diagnoses of this anatomic site may lead to treatment delays, increasing the risk of sequelae.⁶ Familiarizing health care providers with the clinical presentations, classifications according to site, differential diagnoses, and treatment outcomes can increase diagnostic accuracy. These differentiating features are summarized in Fig 4.

Sporotrichosis in ocular adnexa has been reported mainly in the palpebral region.^{4,5,7,8} According to Zhang et al,⁵ the fixed type of sporotrichosis is the most common presentation at this site, with clinical findings of erythematous, painless, and indurated papules or maculopapules with a necrotic center and thick crusts. These result from rubbing the eyelids and surrounding soft tissues with the nails, transmitting fungal spores that can originate in soil, vegetation, organic matter, or the skin of cats with *Sporothrix* species.^{7,9} According to Zhang et al,⁵ the disseminated and lymphocutaneous forms are less commonly seen at this location. Case 1 illustrates lymphocutaneous sporotrichosis from the left upper eyelid through the ipsilateral malar region.

Disseminated cutaneous sporotrichosis is rare and consists of multiple skin lesions without extracutaneous spread, resulting from hematogenous dissemination from a primary lesion or occurring due to

multiple inoculation sites.¹ It rarely affects the eyelids, and usually the skin lesions arise in close approximation, although it can occur in multiple sites, as previously described.⁵ This form usually affects immunosuppressed individuals.

Primary sporotrichosis affecting the conjunctiva, as in case 3, can be classified as extracutaneous; however, it is important to exclude any preceding periocular or nonperiocular cutaneous involvement.⁶ Although there are some published studies of primary conjunctivitis caused by *Sporothrix* species,^{6,10-12} there is still no clear consensus about the route of infection in this presentation. Three hypotheses can be considered: direct contact of the hand containing infectant propagules onto the eye, respiratory droplets from a cat deposited on the ocular mucosa, and systemic dissemination. The last hypothesis seems less probable because there should be intraocular involvement as well, although in many cases it cannot be completely excluded. For that reason, sporotrichosis should be defined as a mycosis of implantation, traumatic or not, and not necessarily a subcutaneous mycosis. In the cases presented here, no patient reported ocular trauma.

Reports of sporotrichosis with intraocular involvement are even rarer; it occurs in immunosuppressed patients, often with exuberant skin lesions and other sites of dissemination. Five types have been described: granulomatous uveitis, granulomatous retinitis, choroiditis, iridocyclitis, and endophthalmitis.^{6,13} Nevertheless, it should be noted that sporotrichosis of the ocular adnexa, especially cases not responding to standard treatments and with ocular symptoms such as reduced vision, ocular pain, and redness, should be referred to an ophthalmologist for early detection of possible intraocular involvement.¹³ In the cases of ocular adnexal sporotrichosis reported in the literature, ocular symptoms and signs are usually not observed, except for redness of the eye as in case 3 of the present series, or epiphora, reported in cases with acute dacryocystitis by *Sporothrix* species, as in case 2 of the present series.^{4,5,14} An increase in atypical forms of the disease is observed in the ongoing epidemic in Rio de Janeiro. It is believed that the high virulence of *Sporothrix brasiliensis* (the most prevalent species in the hyperendemic area of Rio de Janeiro) is the reason for the increase in atypical cases and atraumatic inoculation of the fungus, as in the 3 cases reported here.² Another clinically relevant species, *Sporothrix globosa*, is prevalent in China but seems to cause superficial and localized skin lesions. In vitro and in vivo studies have shown this species to be less virulent and less thermotolerant, which may explain the milder clinical presentations in patients infected

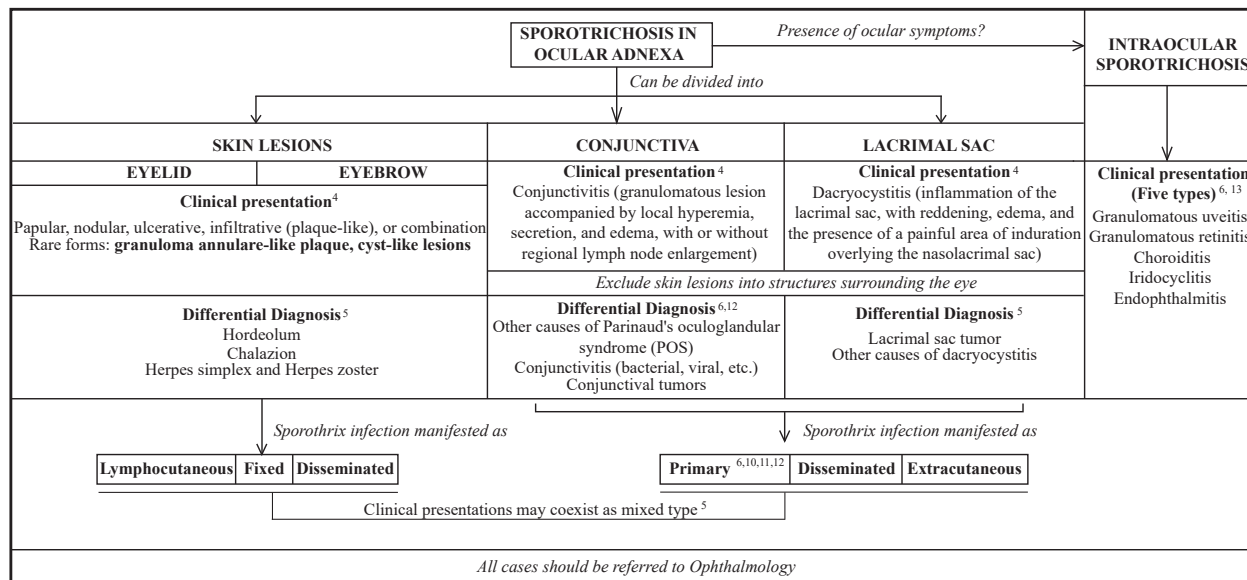


Fig 4. Classification and differentiation of ocular adnexal sporotrichosis.

by it. *Sporothrix schenckii* is more globally distributed and is considered of intermediate virulence.²

Unfortunately, Brazil has experienced a geographic expansion of zoonotic sporotrichosis due to *S brasiliensis* to different states and neighboring countries such as Argentina and Paraguay. In the state of Rio de Janeiro, where sporotrichosis is a notifiable disease, the main referral center, Oswaldo Cruz Foundation (FIOCRUZ), recorded approximately 5000 human cases during 1998-2015 and 5113 feline cases during 1998-2018. Considering the rest of the state, these numbers increase.¹⁵

To improve the treatment of patients with ocular adnexal sporotrichosis, the diagnosis must be confirmed by isolation of the fungus in culture, ideally by collecting scales or discharge from the affected structures. Biopsy can be helpful in suggesting the diagnosis; however, the yeast forms of the fungus are rarely visualized. Moreover, biopsy is an invasive procedure that may have complications.^{1,5}

Itraconazole has become the agent of choice for the treatment of several forms of sporotrichosis. Treatment of ocular adnexal sporotrichosis is similar to that of the cutaneous localized forms.^{6,16} The posology of this azole varies from 5 to 10 mg/kg/day for children and from 100 to 400 mg/day for adults, usually administered for a total of 3 to 6 months.^{9,16} It has been documented that local and invasive *S brasiliensis* disease responds well to this therapy, with shorter duration of the medication compared with sporotrichosis caused by *S schenckii*.² We did not identify the species of the agent; however, the cases reported here showed a good response to itraconazole in a median time of 3 months. Also,

considering the zoonotic hyperendemic scenario in Rio de Janeiro, where *S brasiliensis* is the main involved agent, it was probably the associated species in the cases described here. Physicians, especially infectious disease professionals, ophthalmologists, and dermatologists, should be aware of these forms of presentation, in order to make a timely and precise diagnosis, notably in patients from endemic countries. A thorough history is important, as the patient may not volunteer a relevant history, including trauma and exposure to cats.

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Conflicts of interest

None disclosed.

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