

Periocular Cutaneous Mucormycosis Caused by *Saksenaea erythrospora*

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Abstract

Keywords

- ▶ cutaneous mucormycosis
- ▶ dacryocystitis
- ▶ *Saksenaea erythrospora*
- ▶ liposomal amphotericin B

Saksenaea erythrospora is a rare pathogen in humans. Ten adult cases have been previously reported, eight manifested with cutaneous infection, and two presented with invasive rhinosinusitis infection. The authors present a 16-month-old boy with progressive painful mass at the right medial canthus and upper cheek that was unresponsive to broad-spectrum antibiotics. He underwent an anterior orbitotomy and biopsy. Histopathology revealed broad nonseptate sterile hyphae and grew *S. erythrospora*, which was confirmed by molecular techniques. The patient was treated with intravenous liposomal amphotericin B and oral itraconazole combined with aggressive surgical debridement. The patient made a complete recovery without long-term complications at 4 months of follow-up. Primary cutaneous mucormycosis caused by *S. erythrospora* may rarely involve the periocular region and mimic chronic dacryocystitis. We report the first case of pediatric periocular cutaneous mucormycosis caused by *S. erythrospora*.

Introduction

Saksenaea spp. is a monotypic genus in the order Mucorales capable of causing human mucormycosis infection.¹ There are three described species; *Saksenaea vasiformis*, *Saksenaea oblongispora*, and *Saksenaea erythrospora*.²

S. erythrospora is a new species, which has been recently reported to produce severe human infection. Most clinical manifestations are cutaneous and subcutaneous infection. In a review of the literature, there were 10 adult cases of human infection caused by *S. erythrospora* reported worldwide.^{3–8} We describe a case of pediatric primary cutaneous infection caused by *S. erythrospora* with subcutaneous spread involving the nasolacrimal, cheek, and nasal regions in an immunocompetent host.

Case Report

A 16-month-old boy was referred with a 3-week history of a progressive painful mass at the right medial canthus and upper

cheek. One week earlier, the patient had been admitted with a diagnosis of dacryocystitis at a rural hospital. He was given intravenous Augmentin 40 mg/kg/day and clindamycin 25 mg/kg/day. The mass continued to increase in size despite treatment and he developed restriction in downward gaze. Past medical history was unremarkable and there was no significant history of prior trauma except suspected occult skin trauma on his face while playing at a playground.

The patient was afebrile and appeared systemically well. On ophthalmic exam, the right eye revealed a normal visual acuity by preferential looking test. He had a hard 3 by 1 cm subcutaneous mass at right medial canthal region extending to the nose and cheek (► Fig. 1A). Down gaze was slightly limited with upward displacement of the right globe. There was no proptosis and the remainder of the ocular examination was unremarkable. His blood investigations for complete blood count and anti-HIV were within normal limit.

Magnetic resonance imaging (MRI) showed an enhancing infiltrative lesion involving the right nasolacrimal region,

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Fig. 1 (A) Photograph of the patient before treatment demonstrating subcutaneous mass at right medial canthal region extending to nose and cheek with eyelid edema and redness. (B) Four months after treatment with clinical improvement.

extraconal medial and inferior aspects of the right orbit, and the subcutaneous soft tissue of the right cheek and nose without sinus involvement (►Fig. 2A).

An anterior orbitotomy was performed via a subciliary incision with incisional biopsy. Intraoperative irrigation of the ipsilateral nasolacrimal system demonstrated patency without reflux, effectively ruling out the original presumptive diagnosis of dacryocystitis. Pale, firm nonviable tissue was found and sent for culture and pathological examination. Culture demonstrated broad nonseptate sterile hyphae. A piece of the Sabouraud dextrose agar with fluffy, white mycelial growth was then placed in yeast extract. After 1 week of incubation, long sporangiophores, flask-shaped sporangia were identified suggestive of *Saksenaea* spp. DNA extraction, amplification, and sequencing analysis identified *S. erythrospora*.

The patient was initially treated with 1.5 mg/kg/day of intravenous conventional amphotericin B and changed to 5 mg/kg/day of liposomal amphotericin B after 1 week due to rising serum creatinine. He also underwent additional surgical debridement. After 4 weeks of treatment, he was discharged and continued treatment with itraconazole oral suspension 10 mg/kg/day. At 1 month follow-up, he developed

recurrent right lower eyelid swelling and redness. He was readmitted and repeat MRI showed a residual enhancing lesion with no significant change compared with the radiographic appearance prior to treatment. He had a further surgical debridement and was again treated with intravenous liposomal amphotericin B for 33 days and followed by itraconazole oral suspension 13 mg/kg/day for further 2 weeks. At last follow-up, 4 months later, he showed complete clinical and radiographic resolution (►Figs. 1B and 2B).

Discussion

The genus *Saksenaea* was first reported by Saksena in 1953⁹ from forest soil in India. After the discovery of new species in *Saksenaea* spp. by Alvarez et al in 2010, there were three species overall within this genus. *Saksenaea vasiformis* was the first species in this genus reported to cause human infection in 1976¹⁰ and since then, approximately 40 cases have been published in the English-language literature.¹¹ *Saksenaea oblongispora* has not been reported to cause human infection.

Saksenaea erythrospora is increasingly being described as a cause of human infection. Up to now, there are 10 cases of *S. erythrospora* infection reported (►Table 1).^{3–8} However, it

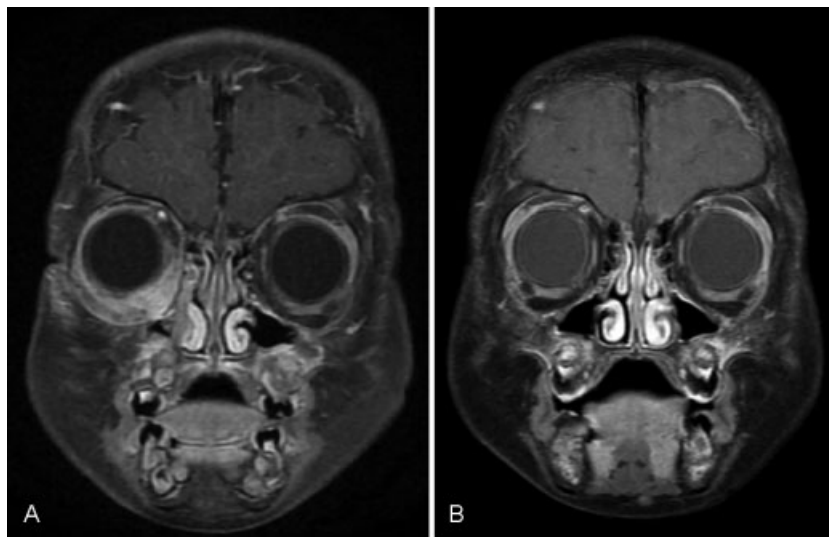


Fig. 2 (A) Coronal T1-weighted magnetic resonance imaging (MRI) before treatment shows enhancing infiltrative lesion involving the right nasolacrimal region and right orbit. (B) At 4 months of follow-up, MRI reveals complete resolution of the preexisting lesion.

Table 1 Published cases of *Saksenaea erythrospora* infection

Reference	Sex/age (y)	Cause of infection	Clinical manifestation	Immune status	Result of treatment
Hospenthal et al ³	Man/26	Combat trauma	Cutaneous and subcutaneous mucormycosis	Immunocompetent	Death
Chantharit et al ⁴	Man/56	Unknown	Invasive rhinosinusitis	Immunocompetent	Loss Follow-up
Relloso et al ⁵	Man/55	Sailing accident	Cutaneous and subcutaneous mucormycosis	Immunocompetent	Resolution
Tendolkar et al ⁶	Woman/44	History of recurrent sinusitis	Invasive rhinosinusitis	Immunocompetent	Resolution
Rodríguez et al ⁷	Woman/29	Breast augmentation procedure	Cutaneous and subcutaneous mucormycosis	Immunocompetent	Resolution
Chander et al ⁸	2 Men and 3 women/ 37,60,31,40,55	4 intramuscular injection, 1 medicated tape application	Necrotizing fasciitis	4 immunocompetent, 1 immunocompromised (DM)	4 resolution, 1 death (immunocompetent)
Present case	Boy, 1.4	Unknown (Suspected occult trauma)	Cutaneous and subcutaneous mucormycosis	Immunocompetent	Resolution

is likely that the actual number of clinical cases might be underestimated due to difficulties in the identification of the species in addition to cases not reported. To identify *S. erythrospora*, culture on different media including potato dextrose agar, Czapek-Dox agar, malt extract agar, and Sabouraud dextrose with incubation at 25 to 37°C is recommended. Typical microscopic features of long sporangio-phores, flask-shaped sporangia, and ellipsoid biconcave sporangiospores can be used to make the diagnosis. However, molecular identification based on DNA sequencing analysis of 3 loci (ITS, D1/D2, EF-1 α) is useful for definitive diagnosis.²

To the best of our knowledge, this is the first case report of pediatric periocular primary cutaneous and subcutaneous infection caused by *S. erythrospora*. In 2011, the first report of human infection caused by *S. erythrospora* occurred in a young man with invasive facial burns following blast trauma in Iraq.³ Since then there have been nine additional cases of *S. erythrospora* infection. All were adult patients and seven had either cutaneous/subcutaneous involvement or necrotizing fasciitis. Two patients had rhino-sinu-orbital diseases. All but one case was in immunocompetent individuals as in our case. From this point of view, it is possible that the immune status preferred by cutaneous and subcutaneous mucormycosis infected by this rare pathogen is immunocompetent status.

The mainstay of treatment should include adequate extensive surgical debridement along with appropriate antifungal regime and correction of any predisposing factors. Based on an in vitro susceptibility data, *Saksenaea* spp. had better response to posaconazole, itraconazole, and terbinafine (low minimum inhibitory concentrations: MICs) compared with echinocandins, voriconazole, and amphotericin B

(high MICs).^{2,3} These profiles were different from antifungal susceptibility testing for *Mucorales* spp. previously reported. In our case, the patient was treated with intravenous liposomal amphotericin B and followed by oral itraconazole combined with aggressive surgical debridement. The patient responded well without long-term complications. The majority of cases in the literature have shown complete resolution. Only two fatal cases have been reported; one was the patient from the war injury, his autopsy revealed main causes of death were from fungal and bacterial sepsis with acute myocardial infarction.³ The second was one of the immunocompetent Indian cases who died from extensive tissue damage and fungal spread.⁸

In conclusion, primary cutaneous mucormycosis caused by *S. erythrospora* is rare but has been increasingly reported over the last decade. These cases can occur in immunocompetent individuals and we report, to the best of our knowledge, the first pediatric case. In our case, involvement of the medial canthus was initially diagnosed as dacryocystitis. The majority of reported cases have responded well to a regime of antifungal treatment and debridement.

Conflict of Interest

The authors have no financial or conflict of interest to disclose.

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