

Pheohyphomycosis: A curious case of cyst

Sir,
Many fungi causing human disease may be melanized and are known as “dematiaceous fungi.” Pheoid fungi may cause clinical syndromes ranging from insidious cutaneous disease to life-threatening infections involving the central nervous system.¹ When these fungi cause subcutaneous infections, it is known as pheohyphomycosis. Melanin acts as a virulence

factor for the fungus as it has scavenging properties and protects it from phagocytosis by neutrophils owing to which the fungus can cause disease in the immunocompetent host as well.²

Exophiala spinifera is a rare cause of cutaneous infection manifesting as pheohyphomycosis or chromoblastomycosis mainly in tropical and subtropical regions of the world.

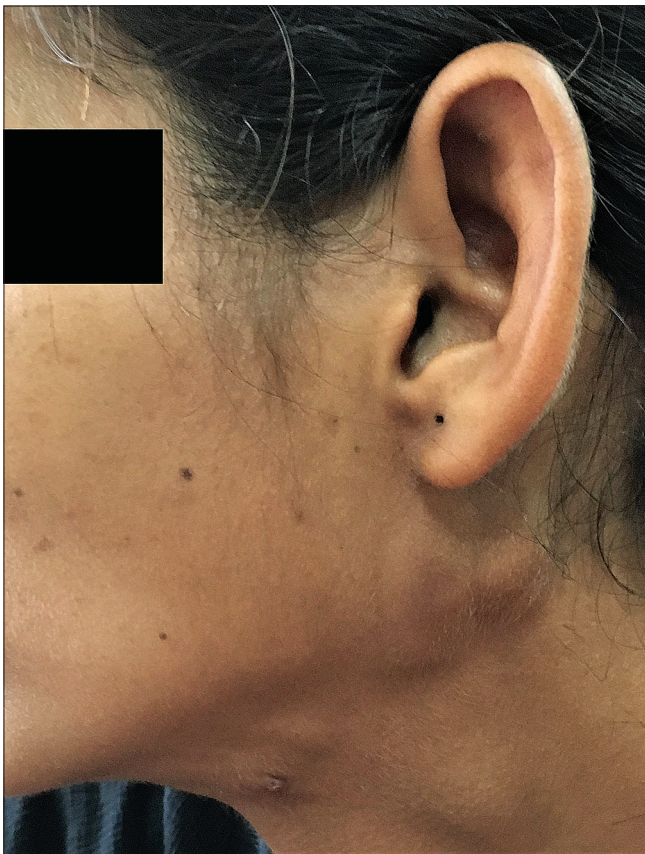


Figure 1a: Multiple subcutaneous cysts seen over the neck along the sternocleidomastoid



Figure 1b: A large subcutaneous cyst 10 cm × 8 cm is seen over the left scapula

There have been only a few case reports of the agent causing cutaneous phaeohyphomycosis from India.³⁻⁶ It is uncommon for the disease to be multifocal, particularly in an immunocompetent individual.⁷

A 15-year-old girl, resident of Uttarakhand presented to our outpatient clinic with a 5-month history of multiple, asymptomatic, skin-colored cysts over both sides of the neck along the sternocleidomastoid; simulating enlarged lymph nodes [Figure 1a]. Similar lesions had been observed two years ago which had resolved with atrophic scarring. The cysts were multiple but discrete, with diameter ranging from 2 to 4 cm. The overlying skin was normal, with no warmth or tenderness. The largest cyst present over the left scapula measured 10 × 8 cm in size [Figure 1b] and was non-tender, fluctuant and negative on transillumination. On the back there were multiple discrete to coalescent, oval, hypertrophic scars, some atrophic areas and some sites of depigmentation at the

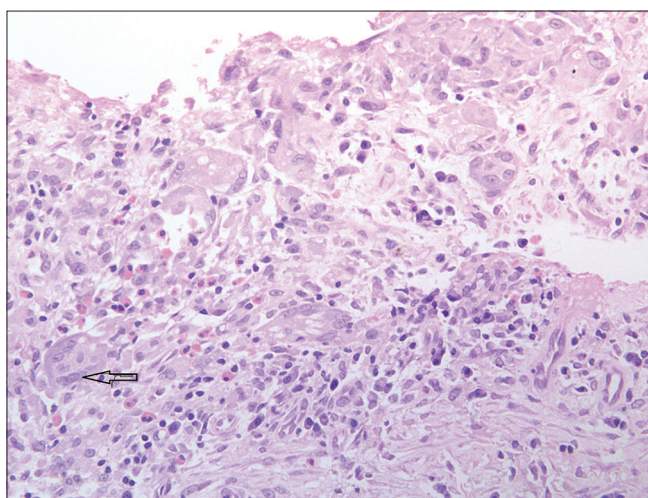


Figure 2: Epithelioid cells with admixed lymphocytes, eosinophils and multinucleated giant cells (arrow) (H and E, ×400)

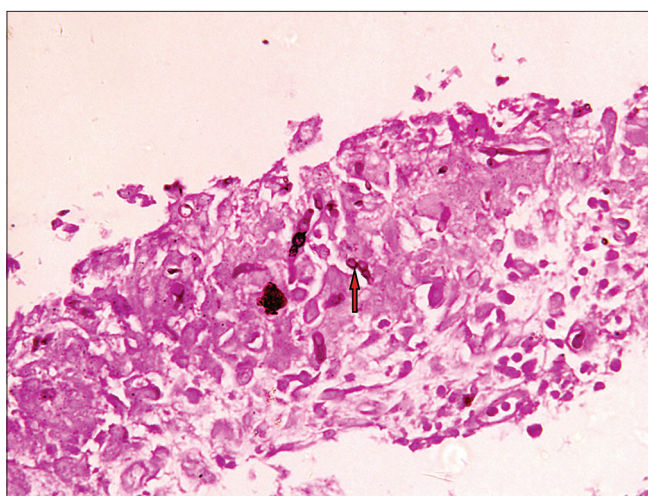


Figure 3a: Periodic acid-Schiff stain highlighting the septate hyphae (red arrow) in the inflammatory tissue (×600)

site of previously healed lesions. During the last episode, no confirmed diagnosis was made, but on a clinical suspicion of deep fungal infection, she was given oral itraconazole 200 mg per day for 6 months with significant improvement, after which the treatment was stopped. There were no associated comorbidities and the systemic examination was normal.

The long duration and asymptomatic nature of the lesions prompted us to consider differentials of mycobacterial or fungal infection.

Routine hematological investigations, chest X-ray and Mantoux were within normal limits. Serology for HIV 1 and 2 were negative. Fine-needle aspiration cytology smear from the cysts showed multiple fungal elements with acutely branching hyphae in a dense acute inflammatory background with foreign body giant cells.

Skin biopsy showed a granulomatous inflammation in the dermis. At the dermis-subcutis junction, there was an abscess cavity lined by granulomatous inflammatory cells, epithelioid cells admixed with eosinophils, lymphocytes and multinucleated giant cells with fungal hyphae [Figure 2]. These hyphae were better visualized on periodic acid-Schiff and silver methenamine [Figure 3a and b].

Pus aspirated from the cystic cavity along with tissue was sent to the mycology department for culture (Sabouraud dextrose agar; 28°C and 37°C). The 10% potassium hydroxide mount showed septate branching hyphae and Sabouraud dextrose agar grew yeast-like dark-pigmented colonies after 5–7 days of incubation both at 28°C and 37°C. Slide cultures on potato dextrose agar for the micromorphology revealed septate, brown, long, annellophores along with obovoidal annelloconidia. On the basis of macro- and micromorphology and polymerase chain reaction analysis the isolate was identified as *E. spinifera*. On the basis of

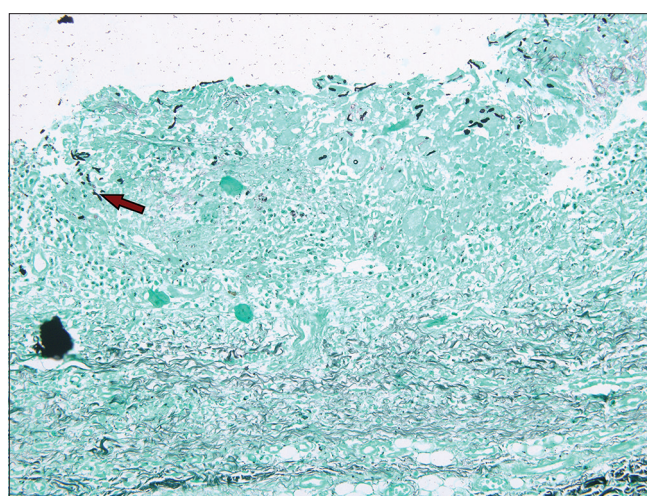


Figure 3b: Numerous fungal hyphae seen in the inflammatory tissue easily picked up as black structures (red arrow) on silver methenamine stain (silver methenamine, ×400)

Table 1: Case reports: Pheohyphomycosis due to *Exophiala spinifera* in India

Years	Age/sex	Lesions
Rajendran <i>et al.</i> (2003)	12/F	Multiple, disfiguring, verrucous, well-defined plaques encompassing lesions of varying sizes on face as well as chest, arms and thighs
Singal <i>et al.</i> (2008)	10/M	Multiple, verrucous, disseminated pheohyphomycotic lesions
Radhakrishnan <i>et al.</i> (2010)	20/F	Nonhealing ulcers over her face, body, arms and legs with unhealthy black granulation tissue and foul-smelling pus
Badali <i>et al.</i> (2012)	55/M	Nodules over hand and knee
Singh <i>et al.</i> (2012)	26/M	Painful papular lesions disfiguring the face

clinical, pathological and microbiological findings, we made a diagnosis of multifocal cystic pheohyphomycosis.

The patient was given itraconazole 100 mg twice a day the larger cyst was aspirated and a drain kept *in situ* for 3 days. The patient has received 8 months of itraconazole with marked improvement. We plan to continue itraconazole 200 mg a day for 18–24 months.

The clinical manifestations of pheohyphomycosis may be protean; cutaneous, subcutaneous, cystic or disseminated. The presence of melanin in the hyphal cell wall gives it the name “black mold.” *E. spinifera* may clinically present both as pheohyphomycosis as well as chromoblastomycosis. The distinguishing feature between the two is the presence of “Medlar” or copper penny bodies on histopathology in chromoblastomycosis.

Molecular techniques to detect species have only been in clinical practice recently, hence, on reviewing the literature, *E. spinifera* was identified as the pathogen causing cutaneous disease in only around 40 cases worldwide.² The first description was in 1954 in a young boy from India with chromoblastomycosis due to *E. spinifera*, since then there have only been five cases of cutaneous disease caused by *E. spinifera* from India.³⁻⁷ [Table 1] All the five descriptions include pheohyphomycotic morphology. Recently, Srinivas *et al.* have reported a 12-year-old immunocompetent boy from India diagnosed with chromoblastomycosis due to *E. spinifera* on the lower leg, who in a span of few months developed osteomyelitis and left hemiparesis.⁸

The disease in our case involved the neck and trunk which are unusual sites as the inoculation of the disease is often by injury over extremities. The disease presented in an immunocompetent young host with chronic, recurring course.

The extensive and recurrent disease with multiple cysts, predominantly truncal involvement in a healthy young girl is, thus, a rare and noteworthy presentation of these uncommon mycoses.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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