Congenital Candidal Onychomycosis
-A Case Report

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Onychomycosis represents 40% of all nail disorders. It is commonly observed in adults but rarely in children. Candida species, although frequently isolated in many occasions, are rarely proved to be the causative agent in the disease. Here we present a male infant who presented with congenital candidal onychomycosis since 1 week of age. He was born to a 33-year-old healthy woman (gravida 2 para 2) via normal vaginal delivery at 36th week of gestation. Before childbirth, the mother had a history of vaginal discharge, from which the bacterial culture showed yeast-like microorganism in moderate amount. On examination, the neonate had no evidence of mucosal and cutaneous candidiasis except yellowish discoloration and thickening of the fingernails and toenails with inflammation of the periungual area. The KOH preparation from nails revealed a large number of pseudohyphae. The fungal culture revealed the presence of Candida albicans. Histopathologically, the nail clipping showed numerous PAS-positive hyphal elements. In addition, ribosomal internal transcribed spacer (ITS) amplified by PCR from nail clippings hybridized only with Candida albicans-specific oligonucleotide probe immobilized on a DNA chip which contained an array of oligonucleotides of detection of yeasts and dermatophytes. The patient received topical sulconazole nitrate 1% solution for 2 months, which led to cure of infected fingernails. (Dermatol Sinica 25: 214-218, 2007)

Key words: Congenital candidal onychomycosis, Candida albicans
INTRODUCTION

Onychomycosis, a fungal infection of the nail, represents 40% of all nail disorders. Fungal nail infections are predominantly observed in adults but rarely in children. Trichophyton rubrum is the most common pathogen while Candida species are not common. Candidal onychomycosis often occurs in patients with chronic mucocutaneous candidiasis. Here we present a healthy male infant who presented with congenital candidal onychomycosis since 1 week of age.

CASE REPORT

A 6-week-old male infant was admitted to the pediatric ward due to young infant fever. We were consulted for yellowish discoloration with thickening of bilateral fingernails and toenails which had been noted since 1 week of age. The nail changes started from the left index finger, and extended to other nails gradually.

He was born to a 33-year-old healthy woman (gravida 2 para 2) via normal vaginal delivery at 36th week. Apgar scores were 9 and 10 at 1 and 5 minutes, respectively. The mother was HIV negative and had not used an intrauterine device. Before delivery, she had a history of vaginal discharge and from which bacterial culture showed yeast-like microorganisms in moderate amount.

On examination, the neonate had no evidence of mucosal and cutaneous candidiasis except yellowish discoloration and thickening of the fingernails with inflammation of the periungual area in all but little finger of both hands and one toe on left foot (Fig. 1A). A potassium hydroxide (KOH) preparation of affected nails revealed a large number of pseudohyphae within nail tissue (Fig. 2). A fungal culture on Sabouraud's dextrose agar tubes at 25°C showed growth of yeast-like colonies only. Germ tube test of the isolate was positive. Histopathologically, the cross section of the distal nail clipping revealed marked subungual hyperkeratosis with parakeratosis and multiple pustules (Fig. 3A). PAS stain revealed numerous hyaline hyphal

Fig. 1
(A) Yellowish to brownish discoloration and thickening of the fingernails with inflammation of the periungual area of the four fingers of each hands.
(B) Progressive disappearance of nail dystrophy after local treatment of sulconazole nitrate 1% solution for 2 months.
elements and some spores throughout the nail plates and subungual hyperkeratosis and a few hyphae were also noted focally within pustules (Fig. 3B). In addition, ribosomal internal transcribed spacer (ITS) amplified by PCR from nail clippings hybridized only with *Candida albicans*-specific oligonucleotide probe immobilized on a DNA chip (Fig. 4).

Topical sulconazole nitrate 1% solution was applied twice daily to the nail plate and the periungual regions of all affected nails. The nail dystrophy almost cleared in 2 months (Fig. 1B).

**DISCUSSION**

The prevalence of onychomycosis in children varies between 0.3% and 0.44%.[4] Yeasts are the source of approximately 5% of onychomycosis, and the majority of which is caused by *Candida albicans*.5 *Candida* species are abundantly present in the environment of neonates, but they affect mainly the skin and mucosa, whereas the nails are usually spared.6 The rarity has been attributed to faster linear nail growth with subsequent elimination of the.
fungus. In infants, the nail involvement with Candida albicans, Candida tropicalis, and Trichophyton rubrum has been reported. Congenital cutaneous candidiasis was first reported in 1960 by Sonnenschein et al. Infants with congenital cutaneous candidiasis typically have generalized eruption of erythematous macules, papules, and/or pustules that sometimes evolved to vesicles and bullae. Onychia and paronychia occurred occasionally, and rarely congenital candidiasis was limited to the nails. Clegg et al. reviewed 10 infants with congenital cutaneous candidiasis with nail dystrophy. Six of the 10 patients were born at 26 to 37 weeks of gestational age. Nail lesions were first noted at birth in one patient, but others were noted to develop nail changes at 2-6 weeks of age. In only one patient among them, the only manifestation of congenital cutaneous candidiasis was nail dystrophy.

Candidal onychomycosis may affect toenails and fingernails by invasion of the nail plate via the hyponychial epithelium. The presence of candidal vaginitis in the mother during pregnancy may be the cause of congenital candidal onychomycosis. Candidal vaginitis occurs in 20% to 25% of obstetric patients and usually resolves after the end of gestation without causing significant morbidity. Congenital cutaneous candidiasis appears to be acquired in uterus by the ascension of organisms from a infected vagina into the uterine cavity. However, the mechanism whereby this may occur is largely unknown. Recently, Chen et al. showed that the isolates of Candida albicans from the bloodstream and the oral cavity of the premature neonate with invasive candidiasis and the vagina of the mother with candidal vaginitis shared a common genotype. It provided direct evidence of the association of congenital candidiasis with candidal vaginitis in the mother. In our case, the mother complained of vaginal discharge before childbirth, and the bacterial culture showed yeast-like microorganism (without further identification then) in moderate amount, which presumably caused the infection.

In a preliminary study of a new method for diagnosing tinea unguium, the positive rates of direct KOH examination and fungal culture of clinically suspected tinea unguium nail specimens are 44% and 16% respectively. The reasons for such poor yields are unclear, although nonviability of fungal hyphae at the edge of the nail has been proposed as an explanation. Kurgansky et al. suggested that histological processing of nail plate clippings with routine PAS staining offers an easy and safe diagnostic tool when evaluating nail disorders in infants, however, it could not provide information about causative fungal species. Here, we demonstrate an improved method which could obtain the causative organism in 2 days. Previously, it has been proved that species identification of clinically important molds by oligonucleotide chip hybridization is reliable and can be used as an effective alternative to the conventional identification methods. In the present case, the PCR products of ITS amplified from nail clippings hybridized only with Candida albicans-specific oligonucleotide probe immobilized on a DNA chip containing an array of multiple oligonucleotides for detection of clinically important yeasts and dermatophytes.

Clinical experience on treating pediatric onychomycosis is scarce. The nail plates are thinner in children, which facilitates penetration of the drug. Several classes of topical antifungal agents are effective against Candida species, including imidazoles (clotrimazole, miconazole, econazole, ketoconazole, etc.), polyenes (nystatin, amphotericine B, etc.) and pyridine-ethanolamine salt (ciclopirox olamine). Ketoconazole, clotrimazole 1% solution, and ciclopirox olamine 8% have been used topically to treat congenital candidal onychomycosis. Topical antifungal agents rarely cause systemic adverse effects, as they are not systemically absorbed. Therefore, it may be prudent to consider topical therapy initially in very young children with a confirmed diagnosis of onychomycosis. In our case, we chose topical sulconazole nitrate 1% due to the convenience and the cost.

In conclusion, we report a rare case of con-
genital candidal onychomycosis, whose causative organism was well demonstrated by the pathologic study with PAS staining, fungal culture and PCR study. In addition, topical sulconazole 1% offered a safe and effective therapy.

REFERENCES