A Case of Congenital Cutaneous Candidiasis with Nail Involvement in A Premature Baby

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Congenital cutaneous candidiasis is a rare disease acquired by an ascending route, and liable to affect the offspring of pregnant women suffering from candidal vulvovaginitis. The skin lesions are present at birth, or shortly thereafter, and the most common cutaneous findings are diffuse erythema with superimposed papules, vesicles, pustules, often involving trunk and limbs.

A 27-day-old male was presented with generalized erythematous scaly patches, pustules since birth and a 17-day history of yellowish discoloration of the finger and toe nails. His mother suffered from vulvovaginitis at the 6th month of pregnancy and her pregnancy ended in premature labor at 33 weeks' gestation. Numerous pseudohyphae and spores were seen in the skin and nails by KOH microscopic examination. Candida albicans was identified by a culture on Sabouraud dextrose agar and API 20C Aux system.


Key Words : Congenital cutaneous candidiasis, Nail involvement, Premature baby

Neonatal candidal infection is manifested most often in the form of oral thrush and less often as lesions confined to the diaper area . The infection is presumed to be acquired by the infant during the passage through the mother's infected birth canal. But congenital cutaneous candidiasis is an entity distinct from these forms of candidal infection, detectable at birth or during the first 12 hours of life and represents an intrauterine infection caused by Candida albicans . We report a case of congenital cutaneous candidiasis with nail involvement in a premature baby which is thought to be very rare, and compare congenital candidiasis with neonatal candidiasis (Table 1).

REPORT OF A CASE

A 27-day-old male was presented with generalized erythematous papules, pustules, and scaly patches since birth and a 17-day history of hyperkeratosis, yellowish discoloration of the finger nails and toe nails.

The male infant was born of a 27-year-old primipara by normal vaginal delivery at the local obstetric hospital. During the 6th month of pregnancy, his mother suffered from vulvovaginitis, which was treated four times and the symptoms subsided. After that, her pregnancy was ended in premature labor at 33 week's gestation. At birth, erythematous maculopapules were found on the trunk. During the first few days of life, the maculopapules became vesicular, pustular, and then spread to the face, and extremities. On the 10th day of life, subungual yellowish discoloration of the proximal nail plate of the finger and toe nails were found. The skin lesions gradually dried and generalized desquamation occurred.
Table 1. Comparison between congenital and neonatal cutaneous candidiasis

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<thead>
<tr>
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<th>Congenital</th>
<th>Neonatal</th>
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<tr>
<td>Incidence</td>
<td>rare</td>
<td>relatively frequent</td>
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<tr>
<td>Onset period</td>
<td>at birth or during the 1st 12 hours of life</td>
<td>after the 1st week of life</td>
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<td>Pathogenesis</td>
<td>intrauterine infection</td>
<td>infection during passing through</td>
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<td></td>
<td></td>
<td>the infected vagina</td>
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<tr>
<td>Site</td>
<td>diffuse scattering on the trunk, neck, head,</td>
<td>oral cavity, diaper area</td>
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<tr>
<td></td>
<td>palm, sole, nail</td>
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Fig. 1. Erythema and scaling on trunk.

On physical examination at birth, his body weight was 2140 gm, the Apgar score was 8 at 1 minute, and erythematous maculopapules were found on the trunk. He suffered from respiratory distress, and had no palpable liver or spleen. At our hospital, there were generalized erythematous papules, pustules, scaly patches(Fig. 1, 2), desquamation and subungual yellowish discoloration of proximal nail plate of right thumb, 1st, 2nd finger nails, left thumb nail, right 1st, left 1st, and 3rd toe nails(Fig. 3). The oral mucous membrane was unaffected.

At birth, routine laboratory findings including LFT, TFT, Chest P-A, TORCH test were within normal limits or negative. But the WBC count was 52,000/µl, C-reactive protein 1.7 mg/dl, total bilirubin 2.7 mg/dl, indirect bilirubin 2.2 mg/dl, and arterial blood gas analysis showed hypoxemia(PO2 of 68.7 mmHg, PCO2 of 49.6 mmHg). These abnormal findings turned normal with time.

In the mycological study, the potassium hydroxide microscopic examinations of face, abdomen, lower extremity, finger and toe nails revealed numerous pseudohyphae & spores, and Candida albicans was identified by a culture on Sabouraud dextrose agar(Fig. 4) and API 20C Aux Strip system.
On the basis of our clinical findings, his present illness, and the mycological study of this premature baby, we made a diagnosis of congenital cutaneous candidiasis with nail involvement. The skin lesion improved by topical applications of 2% ketoconazole cream for two weeks, and the nail lesions resolved spontaneously without any treatment after 4 months.
DISCUSSION

Neonatal infection of *Candida* appears after the 1st week of life and is usually present as oral thrush, which may affect up to 5% of newborns, and less commonly lesions of diaper area. Vaginal candidiasis has been estimated as occurring in 20% to 25% of all pregnant women. Kozinn et al recovered *Candida* in 50% of the infants born to such mothers, and it is presumed that the infant became infected during the passage through the infected vagina. On the other hand, the congenital form of *Candida* infection is seen at birth or shortly thereafter and is very rare. Congenital candidiasis can involve the skin only, the viscera only, or the skin and viscera, and may be accompanied by infection of the placenta and cord. Cabaniss et al reported a case which was suspected to be congenital cutaneous candidal infection, and in 1960 Sonnenschein et al reported a typical case of congenital cutaneous candidiasis which revealed characteristic generalized cutaneous lesions. This is very rare, and as far as we know, about 60 cases of congenital cutaneous candidiasis have been reported in the literature.

The most common cutaneous findings of congenital cutaneous candidiasis are diffuse erythema with superimposed papules, vesicles, and pustules often involving the trunk and limbs. In most cases, the eruptions are noted at birth or within 12 hours, although the eruption appeared between 4th and 6th day of life in some cases. The lesions appear in the trunk, neck, head, and limbs. Palmar and plantar pustules are usually present and are a helpful diagnostic sign. Oral lesions are usually absent, and the diaper area is spared. The nail may also be affected, which is an important sign in distinguishing congenital candidiasis from neonatal infection. Arbegast et al and Lee et al reported cases of congenital candidiasis limited to the nail plates. As the eruption evolves, the erythema fades, the vesicopustules rupture, and desquamation becomes the most prominent feature. In our case, erythematous maculopapules were found on the trunk at birth but oral thrush was not present. During the first few days of life, the maculopapules became vesicular, pustular, and then spread to the face and extremities. On the 10th day of life, yellowish discoloration and hyperkeratosis of the finger and toe nails were found. The skin lesions gradually dried and generalized desquamation occurred. These skin lesions indicate a diagnosis of congenital cutaneous candidiasis rather than neonatal candidiasis. Whyte et al suggested that 0.5 to 2 mm, discrete rounded yellow papules in the affected umbilical cord and placenta were the pathognomonic change. In our case, because the patient was not born at our hospital, we could not examine the umbilical cord or find placenta lesions. Santos et al reported that three of their four cases had respiratory distress, which was observed in 20% of previous reported cases with congenital cutaneous candidiasis. Our case revealed mild respiratory distress, which recovered with time.

Infants with congenital candidial infection can have either a local or systemic disease. Local disease limited to the skin or the placenta and/or umbilical cord usually has a favorable outcome. But disseminated systemic disease, a condition that usually presents without skin lesions, can lead to intrauterine death or death in the immediate neonatal period. The risk factors associated with a high mortality rate are birth weight less than 1500 gm and respiratory distress. Clinical signs of sepsis also portend an ominous prognosis. Although our patient was premature, his body weight was 2140 gm at birth, and following his mild respiratory distress he then had a benign course.

The pathogenesis remains unclear. Ascending infection is the most likely mechanism of infection. In reviewing the pathogenesis of intrauterine infection, Flammin demonstrated that *Candida* can not pass the placenta barrier, and Benirschke and Raphael pointed out that fetal and placenta infection may occur from penetration of the intact membranes from the maternal vagina. Golz et al showed that *Candida albicans* can infect and penetrate through chick chorioallantoic membrane and kill the chick embryo. Furthermore, congenital candidiasis has been reported in an infant born by cesarean section without rupture of the membrane, which suggests that the organisms can penetrate intact membrane in humans. In vitro, *Candida albicans* can grow in the amniotic fluid, invade the fetal adnexa, placenta, or umbilical cord. The cutaneous candidal lesions appear 36 to 72 hours after infection of the skin, which indi-
icates that candidal infection in the congenital cutaneous candidiasis must have occurred within the intrauterine period before the labor. Whyte et al. reported 18 cases whose mother had candidal chorioamnionitis and funisitis infection before the labor, of which 5 cases had skin lesions. In their series, the mothers of 13 out of the 18 infants had a foreign body in situ in the genital tract and 17 infants (94.4%) were premature. They suggested that the above two factors might be related to pathogenesis. But Santos et al. reported that prematurity was found in 26.6% of cases, and prematurity can enhance the chances of the systemic spread of candida infection, either before or after birth.

The infection is limited to the skin in most instances, and has a benign course. In an uncomplicated case with cutaneous infection only, topical application of any of the antifungal agents (nystatin, clotrimazole, miconazole, econazole) for 1 to 2 weeks is usually effective and a nail lesion needs no treatment or only topical treatment. But in the following situations: (1) evidence of respiratory distress or other laboratory or clinical signs of sepsis in the immediate neonatal period; (2) birth weight of less than 1,500 gm; (3) treatment with broad-spectrum antibiotics; (4) extensive instrumentation during the delivery or invasive procedure in the neonatal period; (5) positive systemic cultures; (6) evidence of an altered immune response, systemic antifungal therapy with amphotericin B or 5-flucytosine is required. In our case, the skin lesion improved with a topical application of 2% ketoconazole cream, and the nail lesions resolved spontaneously without any treatment after 4 months.

REFERENCES


29. Flamm H: Die pranatalen infektionen des mensch-er. George Thieme, Stuttgart, Germany, 1959, pp70. Cited from ref. 1

