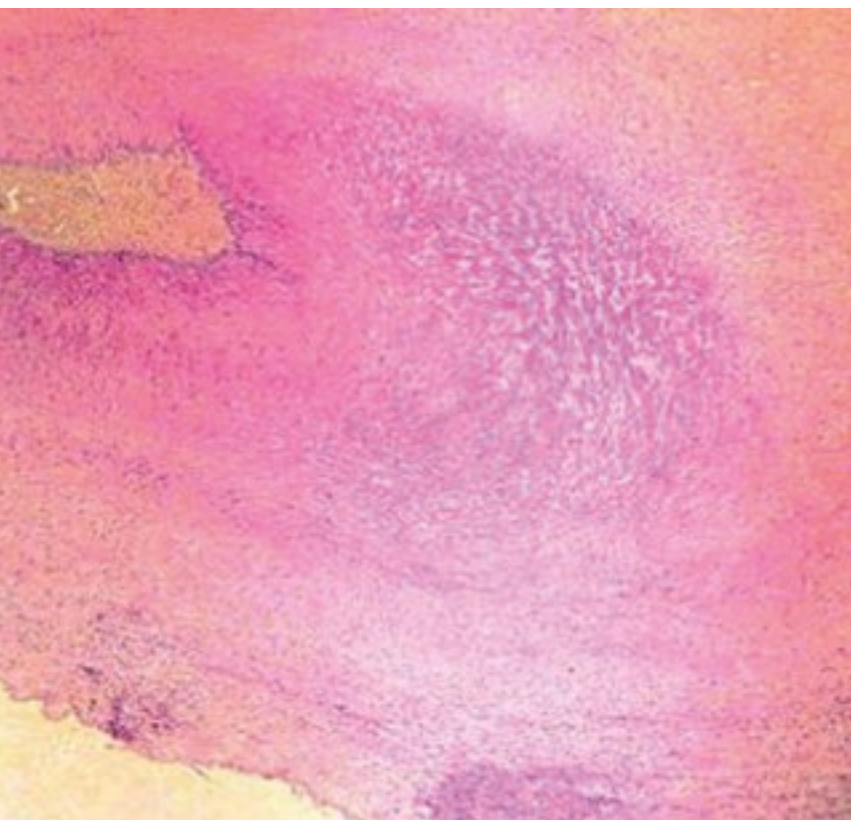


Congenital cutaneous
candidiasis (CCC):
a rare skin disorder of the
neonate

April 2003



Cutaneous congenital candidiasis (CCC) is a rare disease of the term or premature infant consisting of a typical generalized eruption (maculae, papules, or pustules) at birth, usually without other signs or symptoms. CCC is always secondary to *Candida chorioamnionitis*. Despite the high frequency of vulvo-vaginitis in pregnant women, the condition is very rare with at most some hundred cases described in the literature. However, as the condition is essentially benign and self-limited, underdiagnosis is likely. We present two cases of CCC, discuss the diagnostic features and proposed treatments.

A female infant with a birth weight of 3270 g (P 50-90) was delivered by Cesarean section at 38 weeks of gestation after an uneventful pregnancy except for vaginal candidiasis at 26 weeks of gestation treated locally with cotrimoxazole. Membranes ruptured seven hours before birth and amoxicillin/gentamycin was administered during labor for maternal fever. The presence of disseminated white granulomas was noted on the umbilical cord. Adaptation was perfect with Apgar scores of 9, 10 and 10 at 1, 5 and 10 minutes, respectively.

A red maculo-papular rash covering the whole body and sparing only the palms, soles and mucous membranes was noted at birth (Fig. 1, 2). Physical examination was otherwise normal except for mild tachypnea (60

to 80 breaths per minute) on day one with a normal chest x-ray. Total leukocyte count was 11.0 G/L with 52.5 % polymorphonuclear and 11.5% band forms.

Bacteriology of the placenta grew yeast and umbilical cord histology revealed the presence of mycotic abscesses (Fig. 3-5). Mannan antigens were positive and antibodies remained negative. A 7-day-course of oral fluconazole (5 mg/kg/d) was started on day two for presumed congenital cutaneous candidiasis. *Candida albicans* grew on the placental swabs and neonatal gastric aspirations. Blood cultures remained negative for bacteria and fungi. The rash became slightly squamous over the next days and disappeared completely by the end of the first week of life.



Fig. 1

Patient supported non-invasively on nasal prong CPAP.



Fig. 2

*Early appearance of skin rash (case 1):
general distribution*

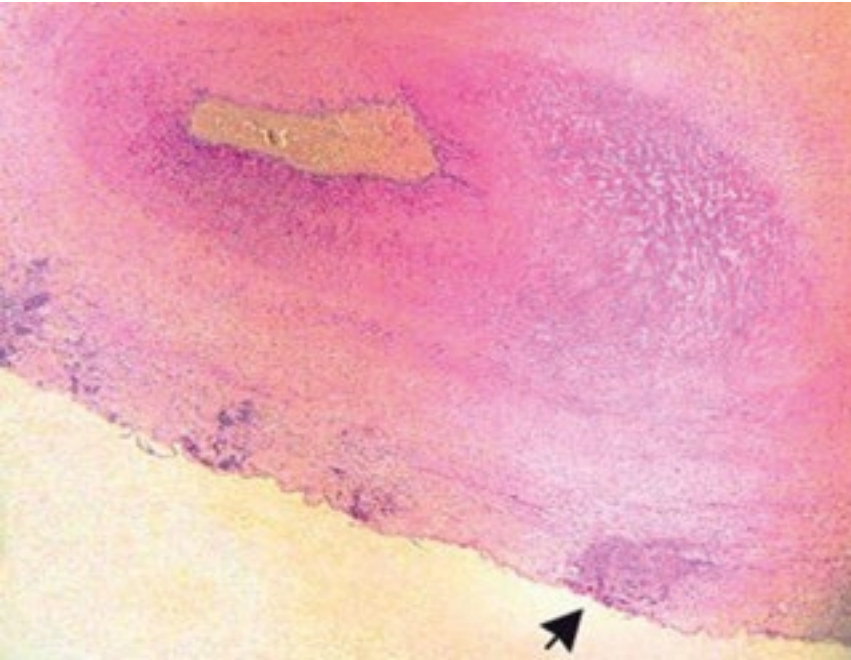


Fig. 3

*Early appearance of skin rash (case 1):
close-up view*

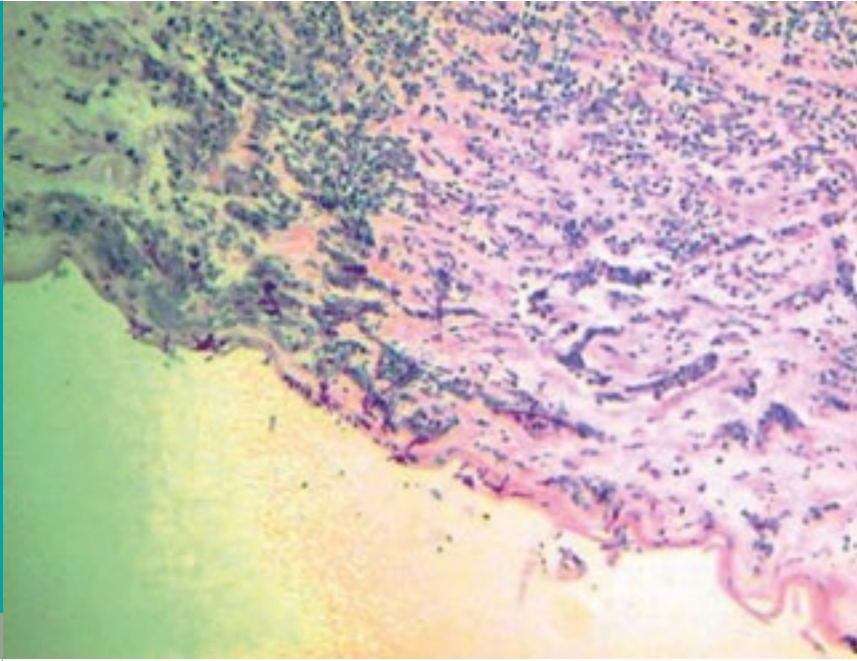


Fig. 4

Histology of umbilical cord (case 1): several candida granulomas (arrow) on umbilical surface (20x).

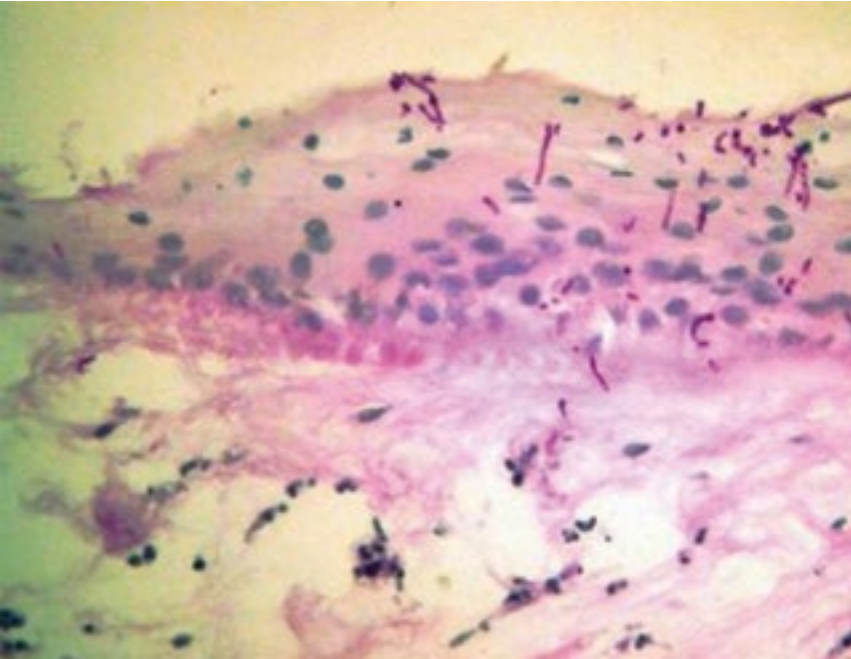


Fig. 5

Histology of umbilical cord (case 1): infiltration of stroma by mycelia (80x).

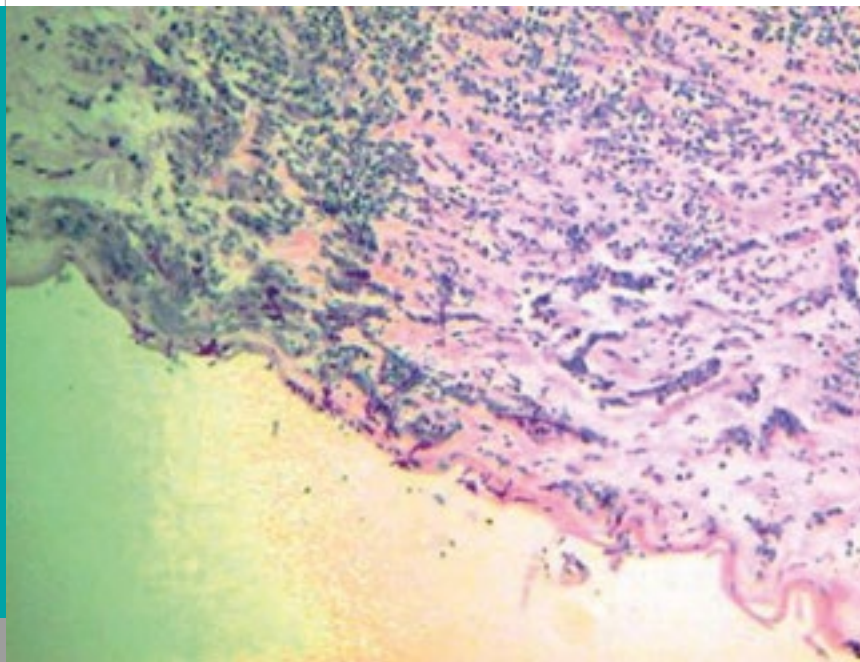


Fig. 6

Histology of placenta: amniotic surface with mycelial infiltration (80x).

A male infant with a birth weight of 2450 g (P 50-90) was born by normal vaginal delivery at 34 weeks of gestation after failed tocolysis. Membranes ruptured 10 minutes before delivery. Apgar scores were 6, 7 and 9 at 1, 5 and 10 minutes, respectively.

The child presented with a disseminated red maculopapular rash, predominant on the trunk and head. White granulomas were noted on the umbilical cord. Amoxicillin/gentamycin and acyclovir were started immediately despite an unremarkable physical examination with the exception of the rash.

Total leukocyte count was 21.5 G/L with 54% polymorphonuclear and 3% band form. Herpes simplex PCR was negative in cerebrospinal fluid and skin specimen.

Candida albicans grew on placental swabs and on neonatal skin swabs as well as in gastric aspirates and urine. Antibiotics and acyclovir were therefore stopped on day 4 based on the diagnosis of CCC and a 7-day-course of oral fluconazole was started. Blood cultures remained negative for bacteria and fungi. After a phase of desquamation (Fig. 6), the rash progressively disappeared over one week.



Fig. 6

Healing skin rash of case 2.

Cutaneous congenital candidiasis (CCC) always appears to be acquired in utero by ascending infection contrasting with neonatal candidiasis, acquired either at birth by an infected birth canal or postnatally. For infants weighing less than 1000 g, contraceptive intra-uterine devices and cervical cerclage have been described to increase the risk for CCC, whereas in neonates over 1000 g this association has not been shown. Premature infants under 1000 g also have the greatest risk of invasive disease due to their immature immune system and compromised muco-cutaneous barrier.

The initial finding may be a generalized or patchy erythema but typically the rash consists of generalized maculo-papular and/or pustulous eruptions with lesions 2 to 4 mm in diameter (Fig 1). CCC is reported more commonly in the full-term infant (1), where it usually has a self-limited and benign course, despite the obligatory presence of *Candida* species chorioamnionitis and funiculitis.

Although there is no proven benefit of any antifungal therapy, some authors recommend topical and oral antimycotics to reduce the risk of systemic spread. Indeed, in the VLBW infant, colonization is generally accepted to always precede invasive disease (1,2). We opted to treat our patients with the well tolerated oral antimycotic fluconazole, and would particularly recommend to do so in the VLBW infant.

Due to the relatively low sensitivity and long delay of blood cultures for yeasts, more recent tests focus on structural membranes of *Candida* by enzyme immunoassay of mannan antigens and antibodies. The recent generation detection tests are considered of moderate sensitivity but good specificity (4, 6) and may precede blood culture results. Positive antigen and negative antibody tests may suggest early systemic infection in our first case. Whether such a test may predict beginning systemic infection of the neonate with CCC remains to be proven as no follow-up serology was performed.

Differential diagnosis of CCC is extensive and should initially include *Listeria monocytogenes* infection, *Staphylococcal pustulosis*, impetigo, Herpes group infection, syphilis and epidermolysis bullosa. However, the diagnosis can rapidly be confirmed on microscopy by the presence of *Candida albicans* spores in skin scrapings, which, after a longer delay, will also result in positive cultures. *Candida chorioamnionitis* or funiculitis is an obligatory finding and placental swabs and pathology is essential. By definition, in CCC *Candida* is confined to the epithelium and gastrointestinal tract. Blood cultures, cerebrospinal fluid and urine remain sterile.

CCC is a rare disease of the neonate caused by *Candida* species acquired in utero. Chorioamnionitis is therefore mandatory and diagnosis requires microbiological or histological demonstration. The major risk factors are the presence of intrauterine devices during pregnancy and cervical cerclage. Diagnosis of CCC allows reassurance of the parents, as the disease is benign and exempt of long-term sequels. As the disease is self-limited and mostly benign, it may be more common than reported in the literature. Oral or systemic antifungal treatment is debated but should be considered particularly for the VLBW infant, at high risk of invasive disease.

We thank Dr. Sophia Taylor for histology images.

SUMMARY

ACKNOWLEDGEMENTS

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CONTACT

Swiss Society of Neonatology
www.neonet.ch
webmaster@neonet.ch