A full-term, 3,700-g male infant was born via vaginal delivery to a 26-year-old gravida 2 para 1001 Hispanic mother, who had good prenatal care. Prenatal laboratory evaluations documented that she was rapid plasma reagin-nonreactive; rubella-immune; HIV negative, hepatitis B surface antigen negative, Neisseria Gonorrhoeae negative; and Chlamydia trachomatis negative. There was no known maternal history of herpes simplex infection, candidal vaginitis, intrauterine device (IUD) usage, cervical cerclage, or amniocentesis. The mother tested positive for group B Streptococcus and received antibiotics before delivery. The mother had artificial rupture of membranes 2 hours before delivery, and there were no signs of chorioamnionitis.

On examination, the infant was vigorous, afebrile, with only mild grunting, although gas exchange was good. There were no signs of respiratory distress. On his skin were diffusely distributed, erythematous papules coalescing into plaques with overlying pinpoint vesiculopustules in face, neck, and trunk. These lesions were apparent at birth.

For diagnosis, see page 553.

Editor’s note: Each month, this department features a discussion of an unusual diagnosis in genetics, radiology, or dermatology. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome. Please e-mail pedann@slackinc.com.
papules coalescing into plaques with overlying pinpoint vesiculopustules on the face, neck, trunk, and extremities, including palms and soles (see Figure 1, page 553; Figure 2; and Figure 3). The back, diaper area, buttocks, and mucous membranes were spared. The umbilical cord was of normal appearance. There were no signs of hepatosplenomegaly. The remainder of the physical exam was normal.

Laboratory examination showed a total leukocyte count of 15,000/mm³, 66% neutrophils, 8% bands, 16% lymphocytes, 7% monocytes, and 3% eosinophils. Chest radiograph was normal, and his grunting resolved shortly after admission.

Metabolic profile, liver enzymes, and culture of blood, urine, and CSF were negative for bacteria, virus and fungi. A Wright’s stain result of the smear of the fluid from the vesicles lacked eosinophils. A potassium hydroxide preparation confirmed the diagnosis (see Figure 4).
The infant was treated empirically with ampicillin, gentamicin, and acyclovir for 48 hours. No topical or systemic antifungal therapy was given. The rash gradually resolved over several days, followed by diffuse superficial desquamation of the skin. The infant had no systemic signs of infection. He was discharged home from the hospital on the fourth day of life and had no recurrence of the rash. A potassium hydroxide preparation showed pseudohyphae and budding yeasts, which confirmed the diagnosis of congenital cutaneous candidiasis (CCC).

**DISCUSSION**

CCC is a rare condition that results from infection from *Candida* sp. acquired in utero. The infection is thought to occur by the ascension of organisms from candidal vulvovaginitis, which is present in 20% to 25% of pregnant women.1-3 Risk factors for CCC include a history of maternal candidal vulvovaginitis; intrauterine foreign body (such as an IUD or cervical cerclage); diagnostic amniocentesis; infant prematurity less than 27 weeks gestational age; and extremely low birth weight less than 1,000 g.4,5 There have been no reported associations between either systemic antibiotic or corticosteroid administration and the development of CCC.6 The preterm infant’s immature and compromised mucocutaneous barriers, as well as systemic host defenses, are thought to be predisposing factors.5,6

It is important to recognize that CCC is acquired in utero, whereas neonatal candidiasis is acquired during the passage through an infected birth canal.7

**CLINICAL MANIFESTATIONS**

Prenatal infection may range from skin eruption without systemic disease to severe systemic involvement. The infection is characterized by a generalized eruption of various morphologies representing different stages of evolution and includes erythematous macules, papules, vesicles, and pustules. The back, extensor surfaces of extremities and skin folds are usually most involved. Pustules are usually seen on the palms and soles. Nail dystrophy has also been documented. Thrush on the oral mucosa is not commonly seen. The eruption usually presents within the first 6 days of life, although most present at birth.7,8

**DIFFERENTIAL DIAGNOSIS**

CCC is diagnosed by presence of spores and pseudohyphae in skin scrapings and culture of the organism from lesions. The differential diagnosis of CCC includes neonatal candidiasis, which differs from CCC by appearing after the first week of life and manifesting as thrush or diaper dermatitis. Some other conditions that may resemble CCC include erythema toxicum neonatorum, transient neonatal pustular melanosis, miliaria, group B streptococcal infection, and herpes simplex/varicella infection. Transient neonatal pustular melanosis and miliaria crystallina present without any erythema. *Miliaria rubra* does not typically present at birth. Herpes simplex infection would present with more erosive disease and fever. *Listeria monocytogenes* may also present with a rash at birth and very characteristic white plaques in the umbilical cord (funiculitis) and placenta.

In almost all full-term infants, CCC tends to follow a self-limited, benign course. Skin lesions typically resolve within 1 to 2 weeks with desquamation. Neonates less than 27 weeks and less than 1,000 g at birth are at greatest risk for systemic infection and death. Neonates with burn-like lesions are at particular risk for systemic infection, with death occurring in 55% of such neonates weighing less than 1,000 g.7,9,10

**TREATMENT**

It is not known if medical intervention affects outcome, given the benign course of the disease. However, some authors, based on anecdotal experience, have recommended topical and/or systemic therapy. Infants with respiratory distress or signs of systemic infection, especially burn-like lesions, should be treated with systemic antifungal therapy. Amphotericin B has been documented to be the first line of therapy for at-risk infants.7,10
CONCLUSION

*Candida* sp. manifests in a variety of ways in humans, from a benign colonizing agent to a major pathogen. It is a well-recognized problem in the high-risk neonate that requires long-term central intravenous lines, so much so that prophylaxis with fluconazole is common practice in the neonatal intensive care unit for the extremely low-birth weight infant. However, we encountered an uncommon presentation of *Candida* sp. We believe it is important for the pediatric practitioner to consider the possibility of this diagnosis in an otherwise healthy newborn that presents with a rash at birth and to recognize the risk factors that would increase morbidity and mortality and that may require systemic treatment.

Most of these infants do not need antifungal treatment; however, those with respiratory distress or other systemic features should receive immediate anti-fungal treatment. Maternal history of intrauterine foreign device may be key to the diagnosis.

REFERENCES