Congenital Syphilis Presenting as Desquamative Dermatitis

Vernessa D. Wood, MD, and Sohail Rana, MD
Washington, DC

During the last 5 years the incidence of congenital syphilis has increased several-fold and reached epidemic proportions. This increase is directly related to a similar increase in cases of primary and secondary syphilis in women and has been linked to the use of "crack" cocaine. Factors responsible for the increase in reported cases of congenital syphilis include poor prenatal care, implementation of new surveillance case definition, failure to perform serological tests, treatment failures with benzathine penicillin, and maternal reinfection. Clinical manifestation of congenital syphilis are multisystemic but are often absent at birth. We report a case of congenital syphilis missed at birth and later characterized by prominent desquamative dermatitis affecting most of the skin surface.

Key words. Syphilis, congenital; dermatitis; prenatal care; neonatology.
J Fam Pract 1992; 35:327-329.

There has recently been a massive increase in the number of reported cases of congenital syphilis in this country. In 1990, more cases of this disease were reported than at any other time since the introduction of penicillin into syphilotherapy.[1] Clinical manifestations of congenital syphilis are protean and multisystemic but are often absent at birth. We describe a case of an infant with multisystemic manifestations of congenital syphilis, including a severe desquamative dermatosis resembling scalded skin syndrome. The epidemiology of congenital syphilis and current recommendations for treatment and follow-up are briefly reviewed.

Case Report

A 7-week-old male infant was brought to the emergency department with a 2-day history of severe diarrhea, progressive listlessness, and a decrease in appetite. His birthweight was 1630 g (3 lb 10 oz). During his 3-week stay in the newborn nursery, no ventilatory support or antibiotics had been administered. At the time of discharge, theophylline had been prescribed and a home apnea monitor was provided. The infant had been apparently healthy at discharge and weighed 2880 g (6 lb 7 oz).

On examination in the emergency department, the infant was found to be febrile, tachypneic, and tachycardiac. His vital signs were as follows: temperature 38.3°C (101°F), heart rate 144 beats per minute, and respiratory rate 56 breaths per minute. The height, weight, and head circumference were all well below the 5th percentile for age. He was ill appearing, lethargic, and severely dehydrated. The skin was mottled and waxy in consistency, with desquamation of the palms and soles. Significant hepatosplenomegaly was also present.

Laboratory data revealed thrombocytopenia (platelets 41 $\times$ 10^9/L), leukocytosis (white blood count [WBC] 61 $\times$ 10^9/L), and elevated liver enzymes (alanine transaminase [ALT] 122 U/L, alkaline phosphatase 315 U/L, lactate dehydrogenase [LDH] 4500 U/L). Urinalysis revealed numerous red blood cells and traces of glucose. There were no WBCs in the cerebrospinal fluid (CSF); the CSF protein and glucose levels were normal. Urine, blood, and CSF cultures were negative for bacterial pathogens.

The baby was hydrated and started on ampicillin and cefotaxime. The next day his hemoglobin dropped from 110 g/L to 77 g/L. Enzyme-linked immunosorbent assay (ELISA) for human immunodeficiency virus was negative. Serology was negative for toxoplasma (IgM <1:20) and cytomegalovirus (CMV <1:8). The rapid plasma reagin card test (RPR) was positive with a titer of 1:128 and the fluorescent treponemal antibody test ab-

Submitted, revised, April 22, 1992.

From the Department of Pediatrics, District of Columbia General Hospital, and Howard University Hospital, Howard University College of Medicine, Washington, DC. Requests for reprints should be addressed to Sohail Rana, MD, Department of Pediatrics, Howard University Hospital, 2041 Georgia Ave, NW, Washington, DC 20060.

ISSN 0094-3509

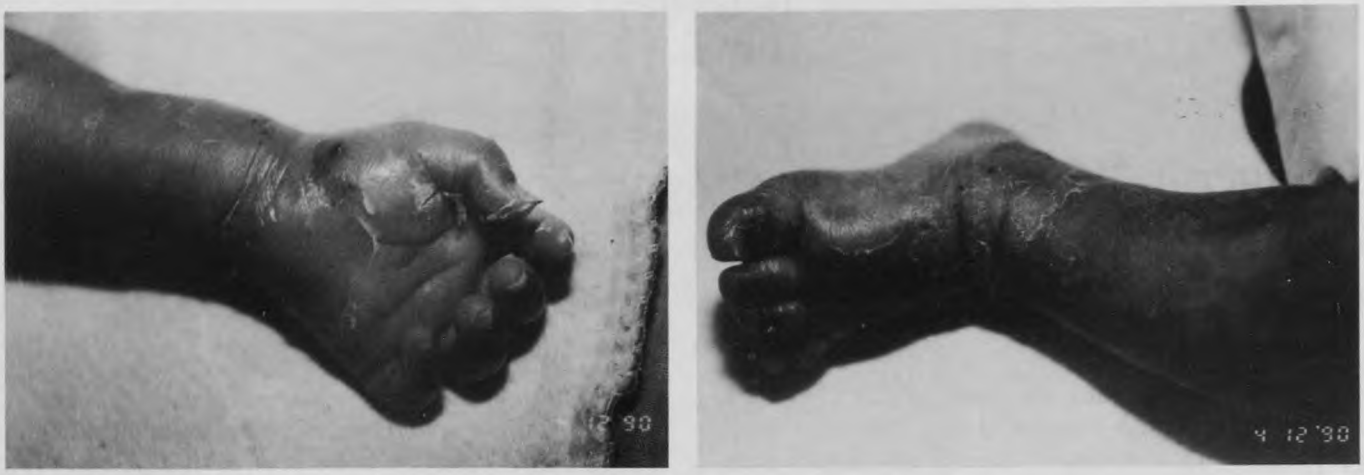

Figure 1. Desquamative dermatitis in an infant with congenital syphilis. The epidermal peeling was thick; the underlying surface was smooth, erythematous, and shiny.

sorbed with nonpallidum treponemes (FTA-ABS) was reactive. The CSF VDRL test was negative. Skeletal radiographs showed periosteal involvement of the long bones. A diagnosis of syphilis was made, and treatment with penicillin was begun.

The desquamation noted at admission progressed rapidly to involve most of the infant's skin surface. The epidermal peeling was thick, and the floor was erythematous, smooth, and shiny (Figure 1). No mucous membrane involvement was noted. Skin biopsy was not performed. By the 4th day of penicillin therapy, the skin lesions had resolved completely without scarring. It was subsequently discovered that the mother's VDRL test had been positive at the time of delivery, with a titer of 1:256, and she had been treated with antibiotics at that time. However, this information had not been transmitted to the nursery staff caring for the baby. Thus, no serological testing was performed on the baby during his stay in the nursery. No additional information regarding prenatal care could be obtained from the hospital where the baby was born.

Discussion

Infants with congenital syphilis often present with subtle and nonspecific symptoms and signs, such as failure to thrive, fever, rhinorrhea, hepatosplenomegaly, hematological findings (hemolytic anemia, thrombocytopenia), and skin rashes. This patient had multisystemic involvement. The skin rash, however, was the most prominent sign. Maculopapular exanthems, especially of the palms and the soles, are the most common cutaneous manifestation of congenital syphilis. Rarely do pemphigoid lesions or extensive epithelial sloughing develop, as seen in our patient. In this case, the exfoliation was a prominent finding, affecting all skin surfaces and resembling scalded skin syndrome.

In 1990, 2841 cases of congenital syphilis were reported to the Centers for Disease Control (CDC).[1] This represents a several-fold increase in the incidence of congenital syphilis from 1987, and parallels a similar increase in the number of reported cases of primary and secondary syphilis in women of childbearing age. The increase in the number of reported cases of congenital syphilis is due in part to the implementation of the new surveillance case definition in 1988.[1–5]

The mothers of almost half of the infants with congenital syphilis as reported to the CDC had received no prenatal care. The increase in cases of syphilis in women has been linked to the use of crack cocaine, in whom poor prenatal care has been documented. Other factors responsible for the increase in cases of congenital syphilis include failure to perform serological tests for syphilis, treatment failures with the use of benzathine penicillin G in neurosyphilis and during pregnancy, and maternal reinfection.[6,7]

Congenital syphilis is preventable in the majority of cases by detection and treatment of syphilis early in pregnancy. In all states, screening for syphilis is mandatory during the first trimester. Screening is also recommended during the third trimester and again at delivery for high-risk populations (substance abusers, women with poor prenatal care, those with other sexually transmitted diseases and women living in areas of high prevalence). The screening at delivery should be performed on maternal blood, since the cord blood may give either

false-positive or false-negative results.[8–11] All patients with syphilis should be encouraged to undergo HIV testing after appropriate pretest counseling.

Guidelines for evaluation of infants for congenital syphilis are outlined in detail in two excellent review articles.[1,10] All infants born to seropositive women (nontreponemal tests confirmed by a treponemal test) should be evaluated for congenital syphilis. In addition to the VDRL and FTA-ABS tests, complete physical evaluation for signs of congenital syphilis should be performed. Additional laboratory testing is required for infants who exhibit signs of congenital syphilis and those at high risk for the disease because of inadequate or no maternal treatment. Evaluation in these neonates must include the CSF VDRL test, protein and cell count determination, and radiographs of the long bones. Follow-up and treatment must be ensured. Neonates should not be discharged from the hospital until the serological status of the mother is known.

Optimal treatment for pregnant women consists of a completed course of penicillin G. For penicillin-allergic pregnant patients, desensitization followed by treatment with penicillin G is recommended.[9] Nontreponemal serological tests (VDRL or RPR) should be repeated monthly. Treatment failures with benzathine penicillin do occur. Most of the failures have been reported when therapy was started in the last trimester.[8–12] Procaine penicillin daily or penicillin G given intravenously for maternal syphilis may be required in the last trimester to treat syphilis in the fetus. This is especially important in HIV-seropositive pregnant women in whom treatment failures with benzathine penicillin are more common. Nonpenicillin regimens are not considered adequate at any stage of gestation, and many failures have been reported with the use of erythromycin.[8–11]

Recommendations for evaluation and treatment of congenital syphilis have recently been modified by the CDC.[8] All infants born to mothers with untreated syphilis should be treated with crystalline penicillin G at a dose of 50,000 units/kg every 8 to 12 hours for 10 to 14 days. This recommendation is based on reports of several cases of treatment failure with the use of benzathine penicillin. The rate of failure with benzathine penicillin G has been reported to be 3%; however, this may be an overestimate.[1]

Those infants who have normal findings on physical examination, normal laboratory evaluation (including CSF analysis and long-bone radiographs), and whose mothers are HIV-seronegative and who have received adequate treatment for syphilis, are at low risk of congenital syphilis. No treatment is necessary unless a close follow-up cannot be assured. These infants can be treated with a single dose of benzathine penicillin G, 50,000 units/kg administered intramuscularly. Infants whose mothers received antepartum treatment inadequate for the fetus (erythromycin or treatment late in pregnancy), even if no evidence of congenital syphilis is present (physical and laboratory evaluation is negative), may also be treated with benzathine penicillin G.[1]

It is essential that all infants born to mothers with a positive serological test for syphilis be followed closely. All treated and untreated infants should be seen at 1, 2, 4, 6, and 12 months of age. Nontreponemal antibody titers (VDRL or RPR) should progressively decrease and become undetectable by 6 months of age. In infants with CSF abnormalities, the CSF VDRL, cell count, and protein quantitation should be repeated every 6 months until they are normal.

Conclusions

Congenital syphilis has reached epidemic proportions in the United States. Clinicians must maintain a high index of suspicion and be prepared to evaluate and treat congenital syphilis despite the best prevention programs.

References

1. Zenker PN, Berman SM. Congenital syphilis: trends and recommendations for evaluation and management. Pediatr Infect Dis J 1991; 10:516–22.
2. Rolfs RT, Nakashima AK. Epidemiology of primary and secondary syphilis in the United States, 1981–89. JAMA 1990; 264: 1432–7.
3. Centers for Disease Control. Congenital syphilis–New York City, 1986–1988. MMWR 1989; 38:825–9.
4. Dorfman DH, Glaser JH. Congenital syphilis presenting in infants after the newborn period. N Engl J Med 1990; 323:1299–302.
5. McIntosh K. Congenital syphilis—breaking through the safety net. N Engl J Med 1990; 323:1339–41.
6. Greenberg MSZ, Singh T, Htoo M, Shultz S. The association between congenital syphilis and cocaine/crack use in New York City: a case control study. Am J Public Health 1991; 81:1316–8.
7. Rolfs RT, Goldberg M, Sharrar RG. Risk factors for syphilis: cocaine use and prostitution. Am J Public Health 1990; 80:853–7.
8. Centers for Disease Control. Guidelines for the prevention and control of congenital syphilis. MMWR 1988; 37(S-1):1–13.
9. Centers for Disease Control. Sexually transmitted disease guidelines. MMWR 1989; 38(S-8):1–43.
10. Ikeda MK, Jensen HB. Evaluation and treatment of congenital syphilis. J Pediatr 1990; 117:843–52.
11. Hook EW III. Treatment of syphilis: current recommendations, alternatives, and continuing problems. Rev Infect Dis 1989; 11: S1511–7.
12. Rawstron SA, Bromberg K. Failure of recommended maternal therapy to prevent congenital syphilis. Sex Transm Dis 1991; 18:102–6.