Congenital syphilis: The continuing scourge

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Abstract

Congenital syphilis is a severe, disabling infection that occurs due to the transmission of Treponema pallidum across the placenta during pregnancy or from contact with an infectious genital lesion during delivery. However, its early diagnosis is often difficult because more than half of the affected infants are asymptomatic, and the signs in symptomatic infants may be subtle and nonspecific. Although its incidence is declining, this long-forgotten disease continues to affect pregnant women, resulting in considerable perinatal morbidity and mortality. We hereby report a case of a 2-month-old infant with early congenital syphilis presenting with joint swellings and Parrot's pseudoparalysis, a comparative rarity in the present scenario. The report also stresses upon the importance of implementing the Centres for Disease Control and Prevention recommendation that all the pregnant women should be screened for syphilis in the first antenatal visit in the first trimester and again in late pregnancy.

Key words: Congenital syphilis, Parrot's pseudoparalysis, Treponema pallidum, venereal disease research laboratory

INTRODUCTION

Congenital syphilis is acquired by an infant from an infected mother by transplacental transmission of Treponema pallidum during pregnancy or possibly at birth from contact with maternal lesions. In the recent years, due to improved medical care, congenital syphilis has become a comparative rarity. We are reporting here a case of early congenital syphilis presenting with pseudoparalysis in order to emphasize that congenital syphilis still exists in the 21st century and global antenatal screening is mandatory to prevent this serious, yet largely preventable disease. Primary skeletal involvement in congenital syphilis is rare. Originally described by Parrot in 1871, this pseudoparalysis represents decreased movement of the extremities secondary to painful syphilitic periostitis.

CASE REPORT

A 2-month-old full-term female infant, born by elective lower section caesarean section, second by birth order, presented with complaints of restricted movements of her upper extremities along with swelling of the wrist joints and knee joints and mild fever since 20 days. She had an unremarkable neonatal course with no history of trauma, bleeding, seizures or altered sensorium. Examination revealed a swelling with no warmth or erythema of the bilateral wrist and knee joints along with marked paucity of spontaneous movements [Figure 1a and b]. The infant cried on passive movement of the joints. There was mild pallor and hepatosplenomegaly, with no icterus, lymphadenopathy, snuffles, feeding difficulty, rash, bulging anterior fontanel or cranial nerve palsy. The mother had an uneventful pregnancy with negative VDRL test and enzyme-linked immunosorbent assay (ELISA) for HIV at 20 weeks of gestation. Her
examination was noncontributory and her previous child was healthy. Based on the clinical presentation, a differential diagnosis of pseudoparalysis secondary to septic arthritis, scurvy or congenital syphilis was considered. The child was administered intravenous Ceftriaxone empirically along with multivitamins, but showed only mild improvement in her condition.

Laboratory evaluation revealed normocytic, normochromic anemia with hemoglobin of 5.7 g/dL, raised ESR (157 mm/h), raised C-reactive protein (132 mg/L), elevated lactate dehydrogenase levels (1607 U/L) and raised alkaline phosphatase (1472 U/L). Other blood cell counts, serum chemistry studies, coagulation studies, HIV ELISA and X-ray chest were within normal limits. Blood culture showed no growth. Ultrasonography of the abdomen revealed hepatosplenomegaly. Radiographs revealed metaphyseal erosions of the distal ends of the ulna and tibia along with periosteal reaction [Figure 2b]. The proximal tibial metaphyses revealed loss of density on the medial aspect along with periosteal reaction, suggestive of osteochondritis (Wimberger’s sign). Distal metaphyses of the humerus also showed periosteal reaction [Figure 2a].

VDRL tests were positive in the baby (1:128), mother (1:32) and father (1:32). Cerebrospinal fluid (CSF) VDRL test of the child was positive (1:4). CSF analysis showed elevated total protein levels (64 mg/dL) and lymphocytosis (5/mm³). The Treponema pallidum hemagglutination (TPHA) test was positive in the baby (1:5120), mother (1:5120) and father.

Based on the clinical manifestations along with pseudoparalysis, typical radiological findings of osteochondritis, positive VDRL and TPHA tests, a diagnosis of early congenital syphilis was made. The infant was managed with intravenous aqueous Crystalline Penicillin G 50,000 units/kg every 4 hours a day for a total of 10 days. The condition of the child started improving after 5-6 days of infusion. Both the parents were diagnosed as latent syphilis of unknown duration and were administered Benzathine Penicillin G 7.2 million units total, administered as three doses of 2.4 million units IM each at 1-week intervals. Repeat VDRL titers of the child 3 months after penicillin infusion showed a four-fold decrease in repeat radiographs of the child showed completed resolution of erosions and periosteal reaction [Figure 3a and b].

DISCUSSION

Congenital syphilis is a rare and serious disease that, although preventable, continues to be a major health care problem. Approximately 66% of infants infected with congenital syphilis are asymptomatic at the time of birth and are identified only by routine prenatal screening. Clinical signs appear in approximately two-thirds of affected infants from the 3rd to 8th weeks of life, and in most cases by 3 months of age.²

Skeletal manifestations of congenital syphilis usually include metaphyseitis, osteitis and periostitis. These
changes are often present at birth and involve the metaphyseal region of long bones. Polyostotic, bilateral, symmetrical involvement is commonly seen. In 1871, Parrot described several children in whom pain in the extremities secondary to syphilitic involvement of bone resulted in a lack of movement. This condition has come to be known as pseudoparalysis of Parrot. Radiographic changes were present in approximately 95% of infants who had overt clinical signs of disease in the series reported by Rosen and Solomon. Because most other clinical findings in congenital syphilis are less specific, the Centers for Disease Control and Prevention (CDC) recommend that radiographs of the long bones be included in the evaluation of all infants who are suspected of having congenital syphilis. In particular, focal erosion of the medial aspect of the proximal tibial metaphysis occurs commonly and is known as the Wimberger sign. Periosteal thickening presents as multiple, irregular layers of periosteal new bone formation. A remarkable feature of the osseous involvement in congenital syphilis is that treatment often results in complete healing with normal growth and an absence of radiographic changes.

In the case presented here, the negative VDRL results of the mother at 20 weeks of gestation misled toward an alternate diagnosis. This suggests that in a newborn presenting with joint swellings, one should have a high index of suspicion of syphilis even if the mother is VDRL negative.

CDC recommends serologic VDRL testing of pregnant women during the first prenatal visit and additional serologic testing at 28 weeks of gestation and soon after delivery in communities in which there is a high risk of congenital syphilis. Going by this recommendation, had VDRL testing been performed in this mother in late pregnancy, she could have been diagnosed and treated, and the congenital syphilis in the newborn could have been prevented. Our case highlights how varied presentations of congenital syphilis can be, and the consequent need for doctors to be aware of them. Also, vigilant screening prenatally, at delivery and an adequate follow-up are critical to reduce the incidence of congenital syphilis.

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Source of Support: Nil. Conflict of Interest: None declared.