Early congenital syphilis with isolated bone involvement: a case report

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Congenital syphilis is the oldest recognized congenital infection and still represents a serious healthcare problem in the 21st century. It is important to be fully informed regarding the early diagnosis and treatment of congenital syphilis to prevent its devastating complications leading to death. In this manuscript, we report a newborn infant with unusual clinical findings of congenital syphilis such as a non-fluctuant mass surrounding the left calf. She did not have any additional system involvement such as hepatic or skin involvement or lymph nodes. To our best knowledge, there are only a few case reports presented with isolated bone involvement. This case demonstrates that congenital syphilis should be considered in neonates with bone fractures, lytic bone lesions and periostitis.

Key words: congenital syphilis, bone involvement, neonate, intrauterine infections.

Congenital syphilis, caused by *Treponema pallidum*, has been recognized since the 15th century. Syphilis as a congenital infection is a worldwide public health problem especially in developing countries[1]. The World Health Organization (WHO) estimates that maternal syphilis leads to 460,000 abortions and stillbirths and 270,000 live born infants with congenital syphilis yearly[2]. At least two-thirds of all fetuses of mothers with syphilis in the infectious period are affected[3]. Most of the affected infants are asymptomatic at birth, but two-thirds develop symptoms in 3-8 weeks[4]. As it continues to be a serious public health problem in the 21st century, it is important to recognize all clinical features of congenital syphilis and begin early treatment. We report a newborn infant presenting only with bulging of the left calf who was diagnosed as an early congenital syphilis case.

Case Report

A 27-day-old female infant was admitted to our emergency room because of a bulging of her left calf. From the history obtained from the parents, the bulging was present at birth and gradually enlarged in two weeks and became painful at palpation. She was continuously crying. There was no remarkable family history except the mother's history of vulvae warts and a multi-partner sexual life. On physical examination, the infant's vital signs were in normal range and her weight was 3260 g. The only positive finding was non-fluctuant mass on the left calf (6x5 cm in diameter). Her physical examination revealed no hepatosplenomegaly. There was no skin eruption or palpable lymph node. The eye examination and hearing test were normal. Laboratory studies revealed anemia (Hb 9.1 g/dl) and leukocytosis (18,000/mm^3) with a normal platelet count. The sedimentation rate was 75 mm/hour. An ultrasonography was performed on the left calf and revealed a hypo-hyperechoic soft tissue mass, surrounding the left fibula. X-ray films showed deep soft tissue swelling, periostitis and bilateral lytic lesions of the proximal and distal fibula and tibia. Periosteal reaction and osteolytic lesions were also observed in the inner aspects of the right proximal femur, right distal radius-ulna and right proximal humerus, and metaphyseal demineralization was detected in all long bones in skeletal survey (Figs. 1, 2). The left calf magnetic resonance imaging showed wide periosteal reaction and hyperintensity on both

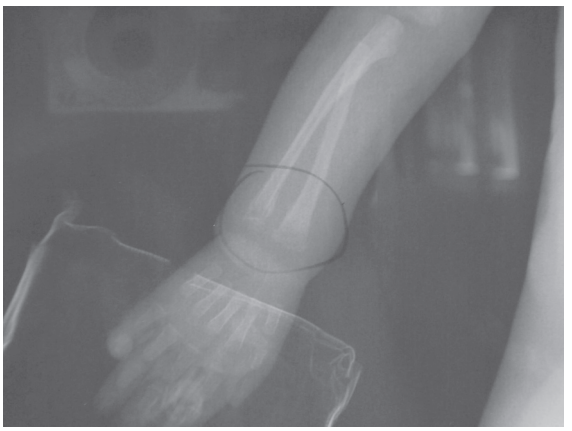

Fig. 1. Periosteal reaction and metaphyseal demineralization of the radius and ulna.

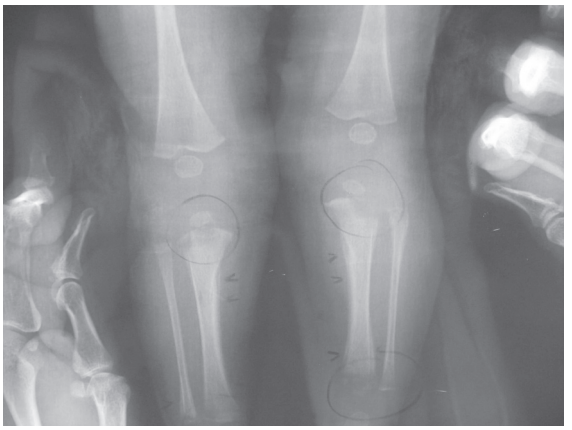

Fig. 2. Osteolytic lesions of the proximal tibial metaphysis.

proximal and distal metaphysis of the fibula and tibia. The Venereal Disease Research Laboratory (VDRL) test was reactive with 1:32 dilutions and *T. pallidum* hemagglutination (TPHA) test was reactive with 1:640 dilutions. VDRL tests for syphilis in both parents were reactive with 1:32 dilutions and TPHA with 1:1280 dilutions. A course of intravenous penicillin G was given to the patient for 10 days, with a resolution of the swelling within three days. The mother and the father were also treated with benzathine penicillin G. We planned to re-evaluate our patient for non-treponemal test, with repeat testing every two-three months until they became non-reactive or diminished four-fold.

Discussion

After the successful treatment of syphilis cases with penicillin in 1943, the disease incidence showed dramatic decline in the mid 1950s. However, several articles have been published recently reporting an increase in syphilis incidence, in the late 1980s in the United States and in the late 1990s in the United Kingdom[1,4,5]. In 2001, WHO estimated that there were approximately 12 million new cases of syphilis in adults globally, with increased prevalence noted in South and Southeast Asia and sub-Saharan Africa[6].

Congenital syphilis occurs when *T. pallidum* crosses the placenta from mother to fetus or during birth by contact with an infectious lesion[7]. Congenital syphilis may appear in two forms. The earliest form begins in the first two years of life and is characterized by cutaneous findings similar to those of acquired secondary syphilis[8]. Lesions are most commonly macular and papular, or purely papular, at first bright violaceous red, and then fading to a copper color[9].

Bone involvement occurs in 60-80% of all untreated early congenital syphilis cases and radiographic abnormalities may be noted in 20% of infants with asymptomatic infection[10]. Bone lesions commonly affect the tibia and other long bones of the body, and are usually multiple and symmetric[7]. The lesions can be classified as osteochondritis, osteomyelitis and osteoperiostitis[9]. The bone involvement can be very painful, causing an infant to refuse to move the extremity, and this finding is diagnosed as pseudoparalysis of Parrot[11].

Bulging of an extremity and tenderness in a newborn infant should be evaluated immediately. The differential diagnosis includes fractures, cellulitis, soft tissue tumors, osteomyelitis, vascular abnormalities and other intrauterine infections.

Other findings in early congenital syphilis include rhinitis, hepatosplenomegaly, lymphadenopathy, and failure to thrive. Anemia, leukocytosis, thrombocytopenia, hypoproteinemia, hypoalbuminemia, hyperbilirubinemia, and elevated liver enzyme levels can also occur[12]. The late presentation of congenital syphilis, generally occurring after two years of age, is characterized by lesions of the bones (perisynovitis, gummas, tooth malformations, saddle nose), cornea (interstitial keratitis), and central nervous system (tabes dorsalis, seizures, generalized paresis)[9]. The laboratory methods for diagnosis of early syphilis are dark-field examination and direct fluorescent antibody tests from lesion

or tissues[11]. However, a rapid diagnosis is possible with the use of serologic tests: the non-treponemal tests (VDRL, rapid plasma reagin) and the treponemal tests (TP-PA = Treponema pallidum particle agglutination, FTA-ABS = fluorescent treponemal absorption)[11].

For definitive diagnosis, the Centers for Disease Control (CDC) recommends identification of syphilis in the mother; lack of evidence of adequate maternal treatment; presence of clinical, laboratory or radiological evidence of syphilis in the infant; and comparison of maternal and infant non-treponemal serologic titers using the same test and preferably the same laboratory, as shown in our patient[13].

Diagnosis of early congenital syphilis is difficult because more than half of the infants are asymptomatic, and signs in symptomatic infants may be subtle and nonspecific[8].

The case reported in this article is one of the early congenital syphilis cases, with no skin lesion, hepatosplenomegaly, lymphadenopathy or failure to thrive, etc. Bone involvement and mild anemia were the two main findings leading to diagnosis. We found two cases reported in the literature which presented with only bone involvement, similar to this case[8,14]. Regarding the increase in the incidence of syphilis, all pediatricians should be aware of silent findings of early congenital syphilis. Bone fractures and lytic bone lesions in neonates should be considered as caused by possible intrauterine infections, especially syphilis[15]. The CDC recommends serologic VDRL testing of pregnant women during the first prenatal visit and additional serologic testing and a sexual history at 28 weeks of gestation and soon after delivery in communities in which there is high risk of congenital syphilis[13]. Congenital syphilis is a totally preventable disease; however, it continues to be a serious healthcare problem in the 21st century, especially in developing countries[16,17]. Regarding WHO reports[6], carrying out syphilis screening tests and preventive strategies should be considered again in some countries.

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