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Case Report

Skeletal manifestations of congenital syphilis: Rare but clinically relevant☆,☆☆,★

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ABSTRACT

Congenital syphilis occurs as a result of maternal transmission of *treponema pallidum* in utero. This condition is mainly diagnosed by treponemal and non–treponemal serologic tests. However, both maternal nontreponemal and treponemal IgG antibodies can be transferred through the placenta to the fetus, thus complicating its interpretation soon after birth. We report a case of a neonate with congenital syphilis whose mother became infected after the first trimester of pregnancy. We report how skeletal radiographs expedite the clinical decision-making process and direct further management of neonates. This case also highlights the need for repeated syphilis screening in the latter part of pregnancy.

Introduction

Congenital syphilis is a multisystemic condition caused by transplacental *treponema pallidum* infection. The clinical, biochemical, and radiographic findings of congenital syphilis are due to the inflammatory response induced by the spirochete in various organs [1]. Multiple studies have shown that vertical transmission to the fetus correlates with the stage of maternal syphilis and that the highest transmission rates, up to 60%, are seen in early maternal syphilis [1].

Early congenital syphilis refers to the manifestation of disease within the first 2 years of life [1]. Skeletal changes in early congenital syphilis include osteitis (0.7%), metaphysitis (24%), and periosteal reaction (34%) [2]. These changes are typically symmetrical in the involved long bones in keeping with its systemic etiology [1,2]. Recognition of these imaging findings can help pediatric teams diagnose early congenital syphilis and initiate antimicrobial treatment.

The prevalence of syphilis among pregnant women in Malaysia is low and was reported as 0.04% in 2016. National congenital syphilis rates have been on the decline from 6.0

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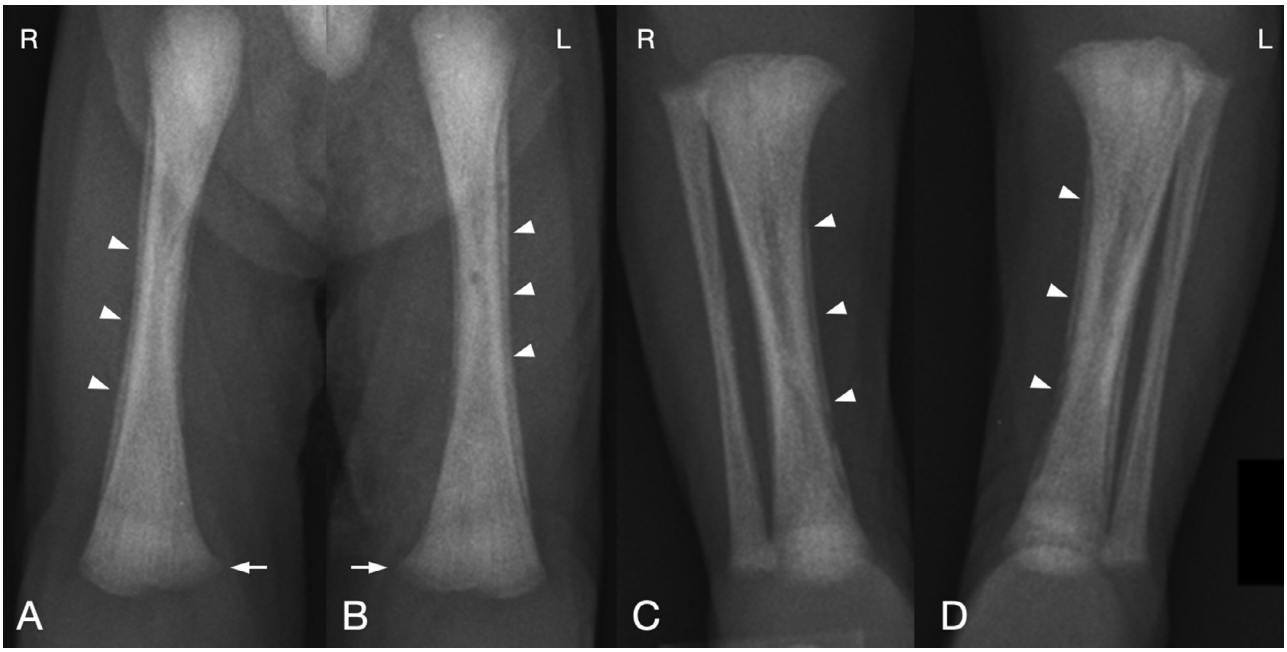

Fig. 1 – Composite radiograph of bilateral femora (A, B), and tibiae and fibulae (C, D). There are symmetrical periosteal reactions in both femora and tibiae (arrowheads). Subtle lucent bands seen at the distal femora (white arrows). Fine linear opacities are present at the distal metaphyseal regions giving rise to a "celery stalk" appearance.

per 100,000 live births in 2012 to 5.0 and 4.0 per 100,000 live births in 2015 and 2016 respectively [3]. In October 2018, the World Health Organization (WHO) reported that Malaysia had eradicated maternal-to-child transmission of syphilis [3]. Yet, sporadic cases do infrequently occur.

Case report

A premature infant was born to a mother who had untreated syphilis during pregnancy. Maternal screening for syphilis was negative during the booking (first) antenatal visit at 8 weeks period of gestation (POG). During subsequent antenatal follow-up visits, serial fetal ultrasound scans demonstrated symmetric intrauterine growth retardation (IUGR). In the third trimester, the mother was admitted for dexamethasone injection, and planned for elective delivery at 37 weeks POG. However, the mother presented with contractions and light meconium-stained liquor at 36 weeks. The fetus was delivered via emergency cesarean section for fetal distress.

The baby boy was vigorous at birth with normal Apgar scores and a weight of 1.42kg. On examination, no hepatosplenomegaly or bruises were noted. The rest of the physical examination was unremarkable. Routine screening for neonatal sepsis was carried out because of prematurity. The boy's rapid plasma reagin (RPR) was reactive with a titer of 1:32 and his Treponema pallidum particle agglutination (TPPA) test was also reactive. His full blood count revealed polycythemia and thrombocytopenia (hemoglobin of 20.6g/dL and platelet count of 74.5×10^3 /μL). Cerebral spinal fluid (CSF) analysis showed low glucose (2.19 mmol//L, reference range 2.22-3.89) and elevated protein (0.57g/L, reference range 0.15-0.45). The CSF culture was negative. Post-delivery, the mother's serum RPR (1:32) and TPPA were positive. The father did not receive treatment at our center and was not contactable by our outpatient medical team.

Long bone radiographs showed symmetrical periostitis involving the diaphysis of bilateral femora, tibiae, and fibulae (Fig. 1). Fine linear opacities were seen extending from the diaphysis to the metaphysis of both distal femora giving rise to a "celery stalk" appearance. Subtle lucent bands were present at the distal femora. Wimberger sign was not observed in this patient. No pathologic fracture was seen. Auditory testing was normal.

Based on the serologic, clinical, and radiological findings, the diagnosis of presumed congenital syphilis was established. Our patient was treated with intravenous penicillin and was subsequently discharged after achieving adequate weight gain. He was scheduled for a repeat RPR titer assessment 3 months after delivery.

Discussion

Congenital syphilis occurs as a result of maternal transmission of *treponema pallidum* in utero. This condition is mainly diagnosed by treponemal and non–treponemal serologic tests. However, both maternal treponemal and non-treponemal IgG antibodies can be transferred through the placenta to the fetus, thus complicating its interpretation [1,4]. As such, RPR and TPPA can show false positive results in uninfected infants of seropositive mothers [1,5]. In our case,

the radiographic bone changes were evidence that presumed vertical transmission has occurred, mandating immediate treatment.

Skeletal manifestations of congenital syphilis occur in 60%-80% of infants with clinical signs of congenital syphilis and in 20% of infants who appear clinically normal [2]. Long bone radiographs are useful in demonstrating skeletal manifestation of early congenital syphilis. Neonatal radiographs showing periosteal reaction, diffuse or localized osteitis, and metaphysitis fulfill the criteria for presumed congenital syphilis [2,6]. In our case, metaphysitis manifested as fine linear metaphyseal opacities giving rise to a celery stalk appearance.

Prompt treatment of congenital syphilis is important in order to avoid later complications that usually manifest after 2 years of life. The most specific sign for late congenital syphilis is Hutchinson's triad which consists of Hutchinson's teeth, eighth cranial nerve deafness, and interstitial keratitis [1]. These complications are a result of persistent inflammation or scars caused by *treponema pallidum* infection in various organs [1].

The incidence of congenital syphilis is low in Malaysia and other developed countries due to comprehensive national antenatal screening programs. Expectant mothers infected with syphilis are provided with appropriate antibiotics to treat the disease and prevent further morbidity. In our case, we postulate that the mother was infected after the initial screening at 8 weeks POG. As repeat screening is not routinely performed, the mother was untreated throughout her pregnancy. Some authors have called for screening in the third trimester or before delivery to address the possibility of treponemal infection during the latter part of pregnancy as demonstrated in our case [4,7].

Repeat RPR testing is used to assess the treatment response in infants who have completed the standard antibiotic regime. Retreatment or repeat CSF study is indicated if there is an absence of fourfold titer reduction over 12-18 months or if titers increase [8].

Conclusion

Long bone radiographs are useful for establishing the diagnosis of congenital syphilis in neonates of seropositive mothers. Familiarity with the radiographic findings of early congenital syphilis can assist clinicians in making appropriate treatment decisions, especially in equivocal cases. As demonstrated in this case, repeat screening for syphilis in the latter part of pregnancy is a prudent measure to avoid vertical transmission of syphilis.

Ethics approval

Ethics approval is not required.

Patient consent

Written informed consent was obtained from the patient's next of kin for the publication of this case report.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2021.09.004.

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