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Case Report

Congenital Syphilis : Still a Reality in Bangladesh

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Abstract

Congenital syphilis is caused by Treponema Pallidum. The fetus can acquire it trans-placentally during maternal spirochetemia, or contact with genital lesion during the intrapartum period. The prevention and treatment of congenital syphilis are very much cost-effective, but if it remains undiagnosed and untreated, can result in a deadly outcome. It can affect multiple organs and systems.

In this present case, skeletal systems were predominantly involved. The clinical presentation was the failure to gain weight, painful mobilization of the right arm, and swelling of the proximal and distal end of this arm. Erythematous, maculopapular scaly skin eruption was also noticed in the scalp and scrotum. Radiological examination showed metaphyseal irregularity, and periosteal reaction at the right humerus, radius, and ulna. Nontreponemal antibody test (VDRL) and a treponemal antibody test (TPHA) both were reactive.

Early congenital syphilis may have a subtle clinical presentation or may remain asymptomatic. Therefore physicians should aware of the clinical presentation of congenital syphilis; at the same time, detailed clinical history can play an important role to suspect congenital syphilis.

Promotion of pregnant women for antenatal care and screening for syphilis in the first trimester are the keys to eliminating congenital syphilis.

Key words: Antenatal care; Congenital syphilis; Skeletal involvement; Syphilis screening.

INTRODUCTION

Congenital Syphilis continues to be a serious health public problem worldwide especially in developing country despite the availability of preventive strategies like antenatal screening of syphilis at the first trimester of gestation.[1] According to World Health Organization (WHO) every year, 12 million people are infected with syphilis globally.[2] The incidence of congenital syphilis per 100.000 live births decreased from 539 in 2012 to 473 in 2016 as reported on the global Sexually Transmitted Infections (STI) surveillance in 2018[2]. Despite the global decrease in its incidence; congenital syphilis remains a significant problem in both developing and developed countries.[3] To the best of our knowledge, there is no recent reported case and no published data regarding the prevalence of congenital syphilis in Bangladesh. The prevention and treatment of congenital syphilis are cost-effective and very cheap, but if it remains undiagnosed and untreated can result in a deadly outcome.[4]

Congenital syphilis is caused by Treponema pallidum subspecies pallidum, which can be transmitted transplacentally to the fetus following maternal spirochetemia or intrapartum by contact with maternal genital lesions. 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.[5]

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Congenital syphilis can involve multiple organs and systems. It is classified as early congenital syphilis when the clinical manifestations occur below 2 years of age, and as late congenital syphilis when the signs and symptoms appear after the 2nd year.[6] The commonest manifestation of early congenital syphilis mostly cutaneous findings like macular and papular, or purely popular rash which are usually at first have bright violaceous red color and then fade to a copper color[6]. In addition to these common skin manifestations, there are maybe acral skin desquamation, vesiculobullous lesion (Pemphigus Syphiliticus) mucous patches, petechiae, erythema multiforme like targetoid lesion. Perioral, perinasal, perianal fissure, and condylomata lata could be the other skin lesion in congenital syphilis.[6] 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.[7] Bone lesions commonly affect the tibia and other long bones of the body and are usually multiple and symmetric. The lesions can be classified as osteochondritis, osteomyelitis, and osteoperiostitis.[8] The bone involvement can be very painful, causing an infant to refuse to move the extremity, and this finding is diagnosed as pseudoparalysis of the Parrot.[9] 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 in early congenital syphilis.[8]

The late presentation of congenital syphilis, generally occurr after two years of age, and 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).[8]

Here we report a case of an infant with congenital syphilis. Who was referred to Outpatient Door (OPD) of Child Neurology Unit of Chattogram Maa - Shishu O General Hospital (CMSOGH) with the provisional diagnosis of Erb痴 palsy of the right arm?

We gave all effort to focus on early diagnosis and treatment of congenital syphilis; we also tried to shed light on the importance of maternal screening and awareness.

CASE PRESENTATION

A 3 months old male infant was delivered at home at term with a birth weight of 2.5 kg. There was no history of regular antenatal check-ups and the mother neither screened for Syphilis nor received any treatment for syphilis. Mother was a housewife and father was the driver of CNG by profession. She had an uncomplicated pregnancy. The baby cried immediately after birth and his post-natal period was uneventful. He was exclusively breastfeeding. Few weeks after birth, mother noticed there is less movement and painful mobilization of the right arm. At one month baby was noted to have poor weight gain. With this complaint, she visited the consultant physician and the baby was sent to the OPD Child Neurology Unit of CMSOGH with the provisional diagnosis of Erb's palsy for further evaluation.

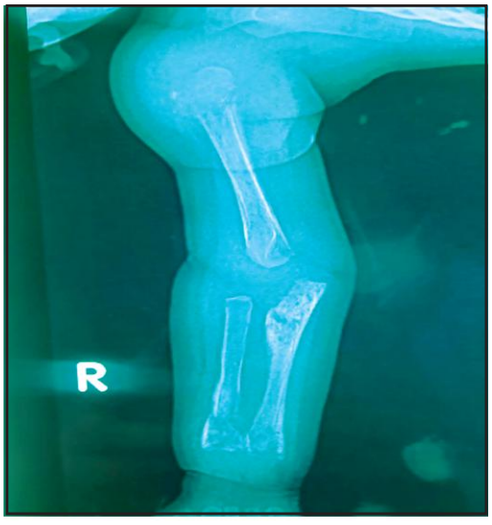

Figure 1 Metaphyseal Irregularity with periosteal reaction at Humerus, Radius and ulna

Figure 2 Erythematous maculopapular scaly lesion over the occipital region of scalp

Figure 3 Clinical photograph of the baby after receiving treatment

On physical examination, the baby was ill-looking, irritable, and emaciated. His weight is 3.6 Kg; Length is 52 Cm; weight for age Z score -3.4 SD, weight for length Z score – 0.8 SD, length for age Z score – 4 SD (According to CDC growth chart 0 – 2 years boy).

There was swelling at the proximal and distal parts of the right upper limb. Lower limbs and left arm revealed normal. The right arm was appeared to be flaccid, hypotonic, and fallen apart. Excessive crying was noticed during the movement of this limb. On skin survey, the macular erythematous scaly eruption was observed in the occipital region of the scalp and scrotum. There was no hepato-splenomegaly and ophthalmological evaluation showed no evidence of chorioretinitis, uveitis, and glaucoma.

The initial laboratory reports showed anemia (Haemoglobin 9.0 g/dl), Thrombocytosis (4,70,000/cm), ESR- 60 mm1st hour, Total count 9,300/cm, Neutrophils 36%, Lymphocytes 56%. CRP- 69.3mg/l. Radiological examination of the right upper limb showed metaphyseal irregularity with periosteal reaction at humerus, radius, and ulna. Non-Treponemal antibody test like the Veneral Disease Research Laboratory test (VDRL) was reactive at a dilution of 1:16. Treponemal antibody tests like Treponema pallidum haemagglutination assay also showed positive at a dilution of 1:320. CSF examination revealed no abnormalities, CSF VDRL was non-reactive. There was no evidence of any meningeal enhancement or hydrocephalus on a CT scan of the brain. Hearing assessment could not be evaluated. The liver function test was normal. The baby was treated with injection crystalline Penicillin G (50000 unit/kg/dose every 6 hourly) for 10 days, after excluding penicillin hypersensitivity by skin prick test. After the ten days course of treatment dramatic improvement was noticed. He became playful, interested to surroundings, gained weight; swelling was disappeared, and able to move his limbs spontaneously. He was planned for monthly doing VDRL test to monitor the titer and discharge. Both parents were reactive to VDRL. They were referred to OPD (Out Patient Door) of Skin&VD for treatment and further evaluation.

DISCUSSION

The case reported in this article is one of the early congenital syphilis that predominantly involved the skeletal system. Bone involvement occurs in 60-80% of all untreated early congenital syphilis cases and radiographic abnormalities may be noted in20% of infants with asymptomatic infection[7]. Bone lesions commonly affect the tibia and other long bones of the body and are usually multiple and symmetric. It is almost consistent with this reporting case, though it was a unilateral involvement, and bony lesion localized at the right upper limb. Radiological examination showed there was a metaphyseal irregularity and periosteal reaction at humerus, radius, and ulna. Clinically the baby presented with painful mobilization of the right upper arm and it appeared to be flaccid, hypotonic. These clinical findings were initially consistent with Erb痴 palsy and make us think about this condition. Nonetheless, the typical posture in Erb's palsy like waiter tips hand, extension at the elbow joint, flexion at the wrist joint, and extension at the metacarpophalangeal joint were absent in this case. Moreover, the swelling at the proximal and distal part of this arm leads us to do the radiological examination.

In contrast, Armangil D et al reported a case, of twenty-seven days old infant females with congenital syphilis, had symmetrical involvements of all four limbs.[10] Who clinically presented with swelling of the left calf muscle, radiological examination showed a bilateral lytic lesion at the tibia and fibula. Periosteal reaction and osteolytic lesion were also observed in the inner aspect of the right proximal femur, the distal end of radius-ulna (right), and the right proximal humerus. Metaphyseal demineralization was present in four limbs.

In our case, erythematous macular scaly skin eruption was found, which was localized at the scrotum and scalp (Occipital region). On the contrary, Kim H.Y et al reported in their case with congenital syphilis, widespread, generalized skin eruption characterized by erythematous targetoid scaly macules, papules, pustules, and desquamation at the hand and foot.[11] Radiological examination of both lower limbs revealed diaphyseal periostitis.

Onesimo R et al found in their case report along with hepato-splenomegaly, copper-red macula popular lesion on the face and hand with erythema, desquamation, and scaling. This was a two-month-old girl with congenital syphilis.[12]

A case report by Subedi S et al reported a two years old male toddler, who presented with condylomata lata(foul-smelling, asymptomatic, erythematous cerebriform plaque) over the perianal region.[13] He didn't have any other systemic involvement.

According to the case report by Khafaja S et al, a five months old infant was having a history of facial and perioral maculopapular rash, since two months of age-associated with poor weight gain.[14] Further evaluation showed moderate bilateral periosteal reaction along with femoral and tibial diaphysis with no metaphyseal abnormalities.

So, according to described case scenario, cutaneous and skeletal involvements were the predominant clinical presentation in congenital syphilis. It is similar to our case as well. Failure to thrive was another common finding had present in most of the cases.

The most effective method to reduce congenital syphilis is to reduce the rates of primary, secondary, and latent syphilis in women of reproductive age. Prenatal syphilis screening is recommended by the American College of Obstetricians and Gynecologists and the American Academy of Pediatrics at the first prenatal visit and again at 32–36 weeks, if the woman is at risk for syphilis.[15] In our country in Bangladesh screening for syphilis in pregnant women is a part of antenatal care in the first trimester. To the best of my knowledge, there is no recent data regarding coverage of antenatal care and screening for syphilis in pregnant women of our country. It was reported that

there is only 68% of pregnant women attending antenatal clinics in developing countries. The average time of first attendance for antenatal care is five to six months.[16]

World Health Organization (WHO) launched the Elimination of Congenital Syphilis Program (ECS) intending to reduce the incidence of congenital syphilis by 0.5 per 1000 live birth and with the target screening for pregnant women for syphilis more than 90% by 2020.[17]

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. At the same time, congenital syphilis is a preventable and treatable disease.[18] Physicians should be vigilant and aware of the diver's clinical symptoms and signs.

The key elements for the elimination of congenital syphilis are providing Antenatal Care (ANC) to pregnant women, screening pregnant women for syphilis, treating seropositive women and their partners, and treating newborn infants of seropositive women[17].

CONCLUSION

To creating awareness about congenital syphilis in the community, promoting pregnant women for seeking ANC are important things to be considered for the eradication of congenital syphilis.

Finally, mobilization of communities including healthy volunteers, traditional birth attendants to refer a pregnant woman for antenatal care and screening for syphilis can play an important role to eliminate congenital syphilis.

DISCLOSURE

All the authors declared no competing interest.

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